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CONTENTS

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Editorial

Reliable Publications - A challenge for the scientific community

Dr G .R. Francis

Today we live in an era of where knowledge grows at an exponential rate far surpassing any other period in the history of humanity. The internet and the ease of global communication is the driving force behind this phenomenon.

However, associated with this growth spurt, we are also facing an unprecedented growth of false claims, and pseudo knowledge. Although this is not a new trend, the sheer volume of false information is overwhelming. With the rush to publish, and the institutional requirements for voluminous publications, the time-tested methods of filtering unwanted claims and disseminating only facts established through rigorous experiments and the proper scientific method are being slowly tossed away. For centuries scientific journals played a crucial role in this vetting process and only disseminated reliable knowledge.

In today's context with the rise of predatory journals this check and balance system, is being removed. and currently the scientific community is faced with the unenvious task of sorting through a mound of rubbish data when searching for true facts and knowledge.

Predatory journals are usually journals which publish open access articles by extracting a fee from the author. The commercial aim of these entities overshadows its scientific purpose. To make the most profit many unsuitable articles and even false claims are published under the term of a scientific publication.

So, the question arises "What can the scientific community do about this problem?" Analyzing the issue, we will realize that it is virtually impossible to monitor and regulate each and every journal published in the worldwide web. The scientific community must accept the fact that these journals are here to stay. An aspect to investigate is "Why do authors publish in these predatory journals?" One of the answers to this is the fact that these journals have developed remarkably easy methods of submission. Usually, submission can be done by a click of a button. Time constrained authors pressurized to make some publication or another for the purpose of employment, renumeration and promotions, very often go against their conscience and submit their works to these journals, because it is easy to submit, and they are fully assured that it will definitely be published.

To counter this problem the scientific community must using multiple strategies. One such strategy is to simplify the submission process. The Batticaloa Medical Journal, identifying this requirement has created a web-based portal for easy submission. Articles can be submitted to our journal by just a click of a button. Another strategy we are employing is not to place a limitation of the number of articles published in each issue. Our main criterion for acceptance of an article is its scientific credibility. If an article carries scientific credibility and it can be understood by the scientific reader, then we feel that it is worth publishing.

Our review process mainly focusses of the accuracy, reliability and methodology of the scientific process employed in writing the article. Our hope is that our journal will provide authors with an alternative option for publication so that they are provided with an opportunity to avoid predatory journals and publish their work with minimal hassle.

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Original research Papers

Experience of students from online mode of assignment in Medical Education during covid - 19

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Abstract:

Assignment is a learning tool for undergraduates to engage with. Peace Medicine is an Inter professional course conducted by Faculty of Health Care Sciences of Eastern University, Sri Lanka. This paper explores the experience gained in this tool as online mode with objectives of its acceptability, application and challenges by the students as group activity in the distance mode during covid era. As part of the teaching-learning activity of a module delivered via online mode, MBBS and BSc Nursing students were grouped, given the task of preparing a report on the roles and responsibilities of different categories of health workers in state health care facilities. The groups were instructed to make communications among themselves via an online platform, visit the nearby health facility, obtain information via direct observation, communicate with staff and submit the report. The experience student groups gained were gathered via a google questionnaire. Twelve groups assigned submitted reports. Six groups responded to the questionnaire. All groups mentioned that they could not visit the health facility and meet health workers due to covid restriction. The information for the report was obtained mostly from the internet sources. All groups responded positively for teamwork and agreed upon the final version of the report via the online communication mode. They also responded positively on MBBS and Nursing undergraduates working as a team, adult learning and self-learning. This exercise explored the difficulty we may face in clinical teaching-learning and the positive outlook on teamwork among different categories of professionals at undergraduate level.

Keywords:
COVID, inter professional education, online distance learning, Peace medicine

Background

Assignment may be a way of assessing the subject material which can either be submitted online or as a written document. Teachers can request to do it as an individual or as a group assignment. It has many benefits such as it reinforces the concepts learned, improves student's performance and motivates the students. Teachers also get feedback from the students about the subject that they facilitated.

Giving assignments to learners is one of the best practices in teaching and learning. The assignment is sufficient if it reaches the goal of achieving it (1).

Peace medicine is a compulsory co-course conducted by the Dept. of Medical Education and Research, FHCS since the first batch of students in 2006. This course is aimed at producing health care professionals' serving the need of Peace building through health care. This is designed as an inter-professional course where MBBS and BSc Nursing students study together. This approach proves their team work and collaborative skills which will help them in the workplace to achieve the health outcomes like quality of care to the patients.

When the assignment is given as a group between two professional courses, there will be an additional advantage of gaining soft skills such as team work, interpersonal and communication skills. A study also proves this experience as to achieve communication, confidence and to achieve knowledge, skills, attitude and professional behavior (15).

Covid pandemic has created more changes in higher education as well where online education has become a new normal method and the inter professional education is also facilitated through this platform. But the staff is facing challenges in making it more engaging and supportive to the students (3).

There was a study which was done in Biochemistry, that also proves that the open book assignment and tutorials' help to strengthen the concepts of the subject and keep the students to have the attention during the class especially the low achieving students (5). Our aim is to motivate the students in whatever way they prefer to learn.

Nowadays there is a high demand for healthcare professions and the educators are facing more challenges to overcome that need of the society. Medical students also adapt to the needs as they are more connected to each other and have the ability to interconnect, collaborate with others and solve problems (10). Knowles stated that adult learners when they mature, they develop their own concepts with the experience they gained and their problem oriented learning skills improve (16). Learning helps the students as mature adult learners to learn what is necessary and to apply at the workplace.

Objectives

Objective of this study was to explore the experience gained in online mode of assignment during covid era.

Method:

Twelve groups were assigned to submit the assignment considering the course. Both MBBS and BSc Nursing students were mixed together in a group. The assignment that we considered in this course is to 'Recognize the roles and duties of different categories of health care workers and list out the roles and duties by visiting them in the hospital, observe their work, talk to them, understand their work and then make the list.' Each group was given a task to prepare a report on the roles and duties of healthcare professionals that each group had chosen to do.

As we are aware, during COVID pandemic, there was a sudden change in the delivery of education via online mode universally (4), in 2020 the modules on Peace medicine were conducted on ODL (Online Distance Learning) via zoom. The assignment was also given in the online mode to the students to visit the health care facility, meet health care professionals and prepare the report for a period of 1 month. A questionnaire was prepared to gather information on their experiences in conducting this assignment. Responses were given to the open ended questions and analyzed using thematic analysis method.

Result and discussion:

Student groups submitted the report of assignments on time. However, they reported the challenges faced due to the restriction imposed due to COVID 19.

Out of twelve groups six groups responded to the questionnaire. All groups mentioned that they could not visit the health facility and meet health workers due to covid restriction. The information for the report was obtained mostly from the internet sources. The identified themes were teamwork, adult learning and self-learning.

Did students do better on teamwork?

All groups responded positively for teamwork and agreed upon the final version of the report via the online mode. They responded positively for both MBBS and BSc Nursing undergraduates working as a team. They tried their best to relate their previous experience with the current knowledge that they gained to do this assignment.

A study which was done by Borges and colleagues says that medical students are more connected nowadays and they enjoy team work, collaborative learning using social media for learning (13). Students stated that "It was a great opportunity to work with different people from different backgrounds and talents [Gp 1]". The same was stated again "We have got a very good experience through the team work [Gp 2]."

Another group also agreed that having an inter professional team benefitted them. We could see the statement as 'We work together and share our opinions with each and others. Finally, we have agreed on the most suitable opinion. I think it was a great chance for us to communicate with others. There was a good understanding between the medicine and the nursing students in doing the assignment as a group [Gp 3]." The statements of students on team work reinforces the benefit of team work which will help them throughout their training and career at workplace.

Did students perform well as adult learners?

Students also showed enthusiasm working together in online mode, students stated that 'We were curious to know about the nature of work done by health care workers [Gp 4].'' There was another finding that it was a great opportunity to have lifelong learning. As adult learners they plan what they learn and have the control over it (7). It also confirms that the students are having skills to develop lifelong learning skills. Adult learners are target oriented in learning (7).

Did students get encouraged to do self-study?

We could perceive that through a statement as 'We could search and get more knowledge about the medical professionals [Gp 5].' It was good to know that they think of them as healthcare professionals which will help them when they work together in future.

A group acknowledged that 'We shared our ideas and thought to find information in various areas. We gather information from books, internet etc [Gp 6]''
A study also states that self-study is a more active form of learning where the students get engaged more and they put more mental effort during discussions (2).

Above mentioned capabilities of students help in working as a team while doing an assignment.

There is more evidence that team skills training and collaborative learning skills augments future practice (14). Our ultimate aim is to produce better health care professionals for the society and every opportunity we give them should promote such abilities. Student motivation has been proved to give academic success as well as better patient outcomes during practice (8). This is said by Thierry Pelaccia & Rolland Viau in another study as higher the motivation of students' greater the quality of learning and performance (6). Educators should encourage students to self-directed learning and in choosing appropriate learning resources which can help them to do the learning to fit their learning styles (9). As the world is changing to a more student centred way of learning, as educators we need to guide the students to achieve their educational goals.

Conclusion:

Inter professional education is becoming a vital component during training in health professions education. COVID crisis has become very challenging for academics and students worldwide when moving to virtual teaching and learning. Students found a way to complete the assignment successfully though it was challenging.

This exercise explored the difficulties faced in teaching-learning especially during covid era and the positive outlook on teamwork among categories of students of different professionals at undergraduate level.

A study which was done in Pharmacy education also stated that students benefit more from team based learning, when they discuss as a team they are actively connected and they share their thoughts and ideas with each other (11).

From our study and the experience we could say that inter professional education trains the students for future practice and a study also states that when there is increased communication and collaboration among inter professional teams can lead to a better care (12).

From this study we could conclude that the students identified online best practices in teaching and learning such as team work especially inter professional team work, self-learning, skill development like creative skills and lifelong learning practices as adults. This approach will lead to a holistic way of practicing their profession in future.

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Original research Papers

Risk factors of common waterborne diseases in Jaffna district

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Abstract:

Objective: To determine the risk factors of common waterborne diseases in Jaffna district.

Methodology: We conducted acase control study from March to September 2015 with 205 cases and 205 controls using an interviewer administered questionnaire by systematic random sampling method. All cases with the final diagnosis as dysentery or enteric fever were considered as cases and controls were selected from the same area of the patient/case. Ethical clearance was obtained from the ethical review committee of Faculty of Medicine, Jaffna. Simple descriptive statistics, with OR and LR were used to describe the results.

Results: Mean age of the respondents was 8.98 years (SD= 13.035). Persons with age above 10 years had more risk for getting enteric fever or dysentery (OR=1.848, p=0.017). Persons with improved water source (p=0.002), safe drinking water (p=0.011), habit of hand washing with soap before meal (p=0.007), children with the past history of same disease (p=0.02), having good knowledge of typhoid transmission (p=0.006) had less risk for enteric fever or dysentery.

Conclusion: age, water source, drinking water, hand washing with soap before meal, past history of food/water borne disease, knowledge of typhoid transmission were the variables had statistically significant association with common waterborne diseases. Unsafe water source, no hand washing with soap before meal, no past history of food/water borne disease and poor knowledge of typhoid transmission had shown significant association in multivariate analysis.

Keywords:
waterborne diseases, Enteric fever, Dysentery, risk factor, Jaffna district.

Introduction

Incidence of some infectious diseases is on the rise in developing countries. According to World health statistics 2012, age-standardized mortality rates for Communicable diseases were 14 per 100000 population in Austria and 1060 per 100 000 population in Central African Republic (WHO 2012). People suffer from diseases due to contaminated food or water. It was estimated that each year food borne diseases causes approximately 76 million illnesses, 325000 hospitalizations, 5000 deaths in the USA and 2 366 000 cases, 21 138 hospitalizations, 718 deaths in England and Wales(2).

An infectious disease spread through the water and cause illness by ingesting the agent with water is called water borne disease(3). In Jaffna enteric fever and dysentery are the common water borne diseases (4). Diagnosis of enteric fever: The causative organisms can be isolated from blood early in the disease and from urine and faeces after the first week. Blood culture is the diagnostic mainstay of typhoid fever. Serological tests based on agglutination antibodies (SAT) are of little diagnostic value because of the limited sensitivity and specificity. However, the demonstration of a fourfold rise in antibody titre is confirmatory of salmonella infection. But because of uncontrolled antibiotic usage by both doctors and patients themselves (over the counter), none of the above is fully appropriate in Jaffna. So, the ultimate diagnosis is done by treating physician considering all clinical features and above lab results.

Diagnosis of Bacillary dysentery: Clinical feature is mainly supportive for the diagnoses. Culture (growing the bacteria in the laboratory) of freshly obtained diarrhea fluid is the only way to be certain of the diagnosis. But even this is not always positive, especially if the patient is already on antibiotics (5).

According to the Epidemiology Unit, Sri Lanka, in 2014, out of 4795 reported dysentery cases1067 were from Jaffna (22.3%) and out of 1032 reported enteric fever cases 321 were from Jaffna (31.1%). In Jaffna, among 1678 reported cases of common notifiable diseases 925 were (55.1%) food and waterborne (dysentery, enteric fever and food poisoning) diseases in 2011 and they were 3958&1428 respectively in 2014(6). These data were generated from inward patients of government hospitals mainly. Relatively high number of patients with the diseases may be managed in private hospitals and OPDs of government hospitals. Same time there was a gap between the data derived from indoor morbidity and mortality report (IMMR) and reported through notifications(7).

Immunization is a short term solution for the control of food and water borne infections. Factors affecting the food and water safety must be studied and sustainable interventions should be planned according to the findings. Though Sri Lanka has a system to monitor both food and water quality through local government authorities and health department we could not maintain a better food and water quality in all parts of the island due to issues in the respective places. Though there are enough literature about risk factors for water/food borne diseases(8) [5, 6,8], we need to study what are the factors predominantly play role in the transmission of disease in our areas. Then only it's easy to plan effective interventions to minimize the incident of the diseases in our areas.

Our aim/objective was to determine the risk factors/ associated factors of common waterborne diseases (enteric fever and dysentery) among patients admitted to 5 major hospitals in Jaffna district.

Objective

To determine the risk factors of common waterborne diseases (enteric fever and dysentery) in Jaffna District.

Methods:

Design and setting

We conducted prospective case control study from March to September 2015 in all four base hospitals, teaching hospital and households (persons) in Jaffna district.

Sample

We recruited 410 (368(184+184)+ 10% non-response allowance) samples which was calculated by "WinPepi" software with 5% significant level, 80% power, 0.66 exposure(9) (using unsafe water for drinking) in cases and Odds ratio 2 by systematic random sampling method.

All cases with the final diagnosis (by the consultant physician or paediatrician as this is the most suitable method for the context-see introduction, p.3) as dysentery or enteric fever were considered as cases irrespective of any age and controls were selected from the same area of the patient/case. A person without any faeco-oral disease within past 6 months (confirmed by detailed history about fever and or diarrhoea) was taken as control. First patient from the ward discharge/diagnosis book was taken and every third case was taken; one out 5 neighbouring house was selected by lottery method and the 1st person above 6 month of age according to the English alphabetical order was selected from the house as control. Persons who stay temporarily and those cannot give the information due to physical or mental status were excluded.

Measures

Data was collected using interviewer-administered questionnaire with variables of sex, age, education level, occupation, water source, drinking water, outside eating habit (within one month), household food safety, safe toilet , hand washing with soap before meal, hand washing with soap after defecation, past history of food/water borne disease, refuse in the environment, knowledge of typhoid agent, knowledge of dysentery agent, knowledge of Typhoid Transmission and knowledge of dysentery transmission

The informed written consent was obtained from the participants; the privacy was maintained while obtaining data by gathering at separate place individually. If the patient's age was less than 18 years, consent and assent took from parents or guardian.

Pre test

Pretest was done before the actual study period from same setting.

Ethical considerations

Ethical clearance was obtained from the Ethical review committee of Faculty of Medicine, University of Jaffna.

Analysis

We analyzed 410 completed questionnaires using SPSS 21. Simple descriptive statistics, Odds ratio & multivariate analysis were used to describe the results.

Results

Background data

The median age of the subjects (403) was 5 years and mean was 8.98 years (SD=13.035). Fifty nine percentages of the subjects were male (Fig. 1). Most of the subjects were non-school going aged children (Fig.2).

Figure 1: Distribution of subjects according to sex

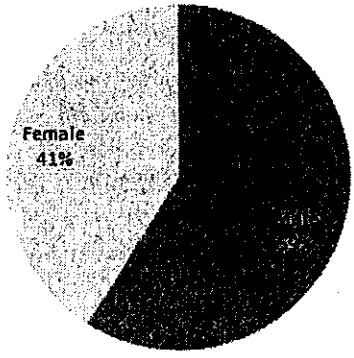
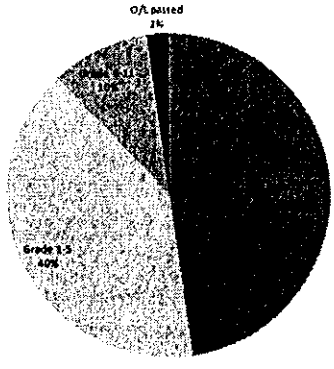


Figure 2: Distribution of subjects according to the education



Risk factors for enteric fever and dysentery

Out of the studied variables following variables had statistically significant association with enteric fever or dysentery (Table 1). Compared to the children aged between 6 months to 10 years, children aged above 10 years had 1.85 times(95% CI=1.117-3.060) more risk for getting enteric fever or dysentery. Compared to persons who used non- improved water source, persons who used improved water source had 0.54 times (95% CI=0.36-0.80) less risk for getting enteric fever or dysentery. Compared to persons who used un- safe water, persons who used safe water had 0.60 times (95% CI=0.40-0.89) less risk for getting enteric fever or dysentery. Compared to persons who practiced poor habit of hand washing before meal, persons who practiced the habit of hand washing with soap before meal had 0.57 times (95% CI=0.37-0.86) less risk for getting enteric fever or dysentery. Compared to children who did not had past history of food/water-borne disease, children with past history of same disease had 0.53 times (95% CI=0.31-0.91) less risk for getting enteric fever or dysentery. Compared to the respondents with poor knowledge about typhoid transmission, those with good knowledge of typhoid transmission had 0.22 times (95% CI=0.07-0.64) less risk for getting enteric fever or dysentery. Other variables (sex, education level, occupation, outside eating habit (within one month), household food safety, safe toilet, hand washing with soap after defecation, refuse in the environment, knowledge of typhoid, knowledge of dysentery agent, knowledge of dysentery transmission) did not show statistically significant association with enteric fever or dysentery.

Unsafe water source, no hand washing with soap before meal, no past history of food/water borne disease and poor knowledge of typhoid transmission had shown significant association in multivariate analysis.

Table 2: Multivariate analysis of Factors associated with enteric fever / Dysentery

Exposure	Beta	SE	OR	95% CI Lower limit	Upper limit	P value
Unsafe Water source	0.59	0.23	1.80	1.14	2.84	0.011
No Hand washing with soap before meal	0.73	0.26	2.07	1.25	3.41	0.005
No Past history of food/water borne disease	0.78	0.34	2.18	1.12	4.24	0.021
Poor Knowledge of Typhoid Transmission	1.21	0.58	3.36	1.09	10.44	0.036

Model Chi square- 30.14, df- 4 and p value -<0.001.
Model Nagelkerke's R square- 0.12

Discussion

Age, water source, drinking water, hand washing with soap before meal, past history of food/water borne disease, knowledge of typhoid transmission were the variables had statistically significant

association with common waterborne diseases (enteric fever and dysentery). Unsafe water source, no hand washing with soap before meal, no past history of food/water borne disease and poor knowledge of typhoid transmission had shown significant association in multivariate analysis.

The principal investigator worked as regional epidemiologist in Jaffna district. During that period he learned that waterborne diseases were occurring in certain households in a same local areas where majority of the houses were not affected. So to elicit the real difference in risk factors between them, community controls near the patients' houses were selected.

Because of an outbreak of dysentery among children during the study period most of the study participants were non-school going aged children (Fig.2).

These findings are in consistency with other investigated outbreaks of typhoid in Bangladesh where contaminated drinking water was found as risk factors. Environmental condition around open well supported the possibilities of its contamination. Faecal contamination seen in water samples provided additional evidence for the source of infection(10). Same findings were observed in Kenya also. But the age group at risk was below 10 years in their study as ours (11). Children 2–4 years old had the highest incidence in an Indian study also (12). In contrast to our study, a Canadian study investigated the demographic determinants of acute gastro-intestinal illness (AGI) stated females were significantly more likely to have AGI than males (13). In an Australian study, women between the ages of 25 and 64 years both with and without AGI were compared; the study found that 18% of women with AGI had at least one child less than five years of age in their household, compared to 5% of women without AGI. Thus, it is possible that females are at an increased risk due to the presence of and interaction with young children within the household (14).

Another study in Bangladesh revealed that young children, persons who consumed un-boiled water and area with poor drainage were at higher risk of developing typhoid (15). Same findings were observed in China and in our study too (16).

Persons with safe drinking water and hand washing with soap before meal had less risk for getting enteric fever or dysentery as significant in our study. These results are broadly consistent with a findings of a systematic review. It expressed several water, sanitation and hygiene interventions were associated with lower risk of diarrhoeal morbidity. Point-of-use filter interventions with safe storage reduced diarrhoea risk by 61% (RR = 0.39; 95% CI: 0.32, 0.48); piped water to premises of higher quality and continuous availability by 75% and 36% (RR = 0.25 (0.09, 0.67) and 0.64 (0.42, 0.98)), respectively compared to a baseline of unimproved drinking water; sanitation interventions by 25% (RR = 0.75 (0.63, 0.88)) with evidence for greater reductions when high sanitation coverage is reached; and interventions promoting hand washing with soap by 30% (RR = 0.70 (0.64, 0.77)) vs. no intervention (17). But a survey conducted in Mozambique revealed that although there is no conclusive evidence of the additive effects of the water supply, sanitation and hygiene promotion on diarrhoea, it seems reasonable to design comprehensive programmes that take into account the joint improvement of these three factors (18).

Our study resulted persons with good knowledge of typhoid transmission had less risk for getting enteric fever or dysentery. Nearly same finding observed in a study that effective locally-informed education programs have the potential for clarifying misconceptions, improving practical knowledge, and instigating behavioral changes, which in turn may reduce diarrhea-related mortality along a more sustainable long-term platform than what has been undertaken to date in the Niger River Basin of Mali (19).

A study demonstrated that highly localized clustering of typhoid fever during an epidemic in an urban African setting suggested the targeted intervention (20). It is necessary to understand how food becomes unsafe to eat and what proactive measures can be taken to keep food safe to prevent foodborne illness.

Conclusions and Recommendations

Persons with age above 10 years had more risk for getting enteric fever or dysentery. So it's recommended to do more awareness programs to secondary school children in both government and private education institutions. Persons with improved water source had less risk for getting enteric fever or dysentery. So the government or non-government organizations need to take measures to improve the water sources as this is a known problem in Jaffna district. Persons with safe

drinking water had less risk for getting enteric fever or dysentery. So the people must be educated to use safe drinking water. Persons with the habit of hand washing with soap before meal had less risk for getting enteric fever or dysentery. Hence it's essential to promote hand washing with soap before meal in both personal and public settings. Children with the past history of same disease had less risk for getting enteric fever or dysentery. It shows that the children with the past history of the disease had some opportunity to improve their context may be due to the visits of PHI or PHM to follow the notification. It indirectly tells that general public health education should be improved. Persons with good knowledge of typhoid transmission had less risk for getting enteric fever or dysentery. This is also indirectly tells that general public health education need to be improved. Unsafe water source, no hand washing with soap before meal, no past history of food/water borne disease and poor knowledge of typhoid transmission had shown significant association in multivariate analysis (table2). So, in long-term, safe water supply, proper hand washing practice and improvement of knowledge about waterborne diseases need to be ensured by the relevant authorities for healthy future.

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Conflict of interest

The authors report no conflict of interest

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Original research Papers

Leprosy case detection in Jaffna district: A searching strategy in low endemic area

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Abstract:

Background and objective: Large scale health education or awareness program and active case detection are the most effective activities used to detect new leprosy cases. The objective of this study is to describe a modified method of new case detection of leprosy in Jaffna district.

Methods: This was a descriptive study with retrospective and prospective data collection. This was done through house to house visits/community based mobile clinics and dermatology clinic at the teaching hospital Jaffna which is the only hospital providing diagnosis and treatment of leprosy in the district.

Results: Overall 55.9% of 7792 target population was examined or screened. MOH Chavakacheri had the highest and MOH Velanai had the lowest target population. Out of 4354 persons examined, 16.4% had some skin lesions and 2.0% had suspected (as leprosy) skin lesions in the district. Out of 88 suspected persons (as leprosy) with skin lesions, 50% visited to dermatology clinic in the district. New cases were detected in MOH Chankanai (02), MOH Sandilipay(01), MOH Kayts (01)and MOH Kopay(01) among screened population.

Conclusion: Around 16% of screened, had some skin diseases and around 2.0% had suspected (as leprosy) skin lesions and 5 new leprosy cases (6.4/10000) were detected.

Keywords:
Leprosy, Jaffna District, new case detection.

Introduction

Leprosy cases are detected mainly on the basis of three cardinal signs of leprosy: hypo-pigmented or erythematous skin patch with loss of sensation, definite enlargement of peripheral nerve/nerves and diminishing of nerve function, finding acid fast bacilli (AFBs) in a slit skin smear or the biopsy [1]. Large scale health education or awareness program and active case detection are the most effective activities used to detect new cases. And the control strategies depend on early case detection and treatment with multidrug therapy (MDT). In early years followed by the introduction of MDT, leprosy elimination campaigns (LECs) were very effective in new case detection in high endemic countries. But in last 10 years the reduction is not significant [2] over the years globally. But in long run, LECs were highly resource consuming, especially where the public health infrastructure and community participation are poor. Therefore, the promotion of self-reporting is an easy and cost-effective way of case detection. As large scale active case-finding campaigns are expensive it is important to identify the suspected cases by active case finding with the help of available resources like field staff or community volunteers with adequate basic training. Then the suspected persons can be referred and followed to see the compliance of referrals. In Sri Lanka, the new case detection has been fluctuating between around 8/100000 and around 12/100000 for the last 20 years. For the last 10 years it is fluctuating around 10/100000.This may be due to change in the program from vertical to integration of services with general health care provisions in 2001 with the following key objectives [3],

- MDT made available at all curative health centres, private and public.

- Health care providers ready and willing to diagnose and manage leprosy cases.
- Community awareness of the early signs of leprosy and where to go for treatment.
- An operational system of record keeping and monitoring.

However, with time, the case detection programs have been centred on consultant dermatologists and the workforce towards the field was reduced or none in some parts of the country. So, more cases are detected in resource rich provinces. As western province enjoys with more resources, it detected the highest percentage. Non-specialist medical officers were reluctant to diagnose and manage leprosy cases due to increased role of dermatologists. But, there are some districts which detect large number of cases in the absence of dermatologists [4](1) (Wijesinghe and Ranjan Wijesinghe, 2013). The cases were detected by trained MO Dermatology. This in turn with other factors, made Sri Lanka also to experience vast variations between districts in new case detection, from 1.7/100000, the lowest in Nuwara Eliya and 28.4/100000, the highest in Batticaloa [5]. At the same time some non-government organizations (NGOs) try to fill the gap by helping case detection at field level with different methodologies like assisting mobile skin clinics, house to house search for skin diseases and contact examinations, etc.

The new case detection rate was 9.3/100000 in Jaffna which is slightly lower than the national average and higher than global and regional averages. Again, within the district, there were variations between MOH areas. It shows that with time, cases have been detected in new MOH areas and within MOH areas detected from new PHI areas. The objective of this study is to describe a modified method of new case detection of leprosy in Jaffna district.

Methods

This was a descriptive study with retrospective and prospective data collection. This was done through house to house visits (HHV)/community based mobile clinics (CMC) and dermatology clinic at the teaching hospital Jaffna (THJ) which is the only hospital providing diagnosis and treatment of leprosy in the district. HHVs/CMCs were held in suitable locations in the communities from where (all parts of district including the islands) patients had been reported within the past 5 years and in communities which are considered to have 'pockets' by health workers/related NGOs.

Initially a list containing details of patients reported within the past 5 years was prepared by extracting information from the district leprosy register (this is a standard register provided by ALC, Colombo to each district) from RDHS office. The quality of data was ensured by receiving the list of notified patients from the Infection control nursing officer, THJ, receiving the notified patients' information from all MOH areas and cross checking the individual patients' forms kept at the RDHS office, Jaffna. Then it was categorized into MOH areas in order to identify the localities by PHI areas and the residential address of the patients. These information were shared with the MOH 2 localities from each MOH area except Kopay, Chavakachcheri and Chankanai (where 3) were selected because of endemicity (Chankanai) and vast geographical area (Kopay and Chavakachcheri) with the consensus of them to conduct the HHVs/CMCs

Initial data from district leprosy register were collected retrospectively and prospective data were collected during HHVs/CMCs, followed them in the dermatology clinic, THJ and finally from district leprosy register.

The research was started with the discussions with RDHS and MOH staffs on September 2012. As the principal investigator was the Regional Epidemiologist, he has started to talk more about leprosy during public health review meetings and monthly conferences. The data collection period was from 07.07.2015 to 06. 07.2016.

All residents (have lived in the district for more than 6 months) of Jaffna District were the study population.

Initially a community mapping was done to identify locations for the HHVs/CMCs. More such locations were selected near to the already identified 'pockets' and the 'pockets' of contacts of the patients.

There was no formal sample size calculation as this was based on purposive sampling. In each MOH area, two localities were selected on the basis of high number of reported leprosy cases within past 5 years (from 2010-2014) and socio economic background of the localities. Altogether 27 HHVs/ CMCs were conducted and on average 161 people were screened in a location. In each locality, all the houses were visited according to the map given by the area PHI. If one area could not be covered in the same day, it was visited the following day to cover the all houses. The number of houses in locality

varied from 22 to 151.Therefore, this component has screened 4354 persons and 88 suspects were referred and followed with the help of MOH team.

Data record sheet and data extraction sheet were used to collect information from HHVs/CMCs and district leprosy register respectively.

Initially a one day workshop was conducted to all field staff (MOOH, PHII and PHMM) in the district to update their knowledge and skill of leprosy case detection and the methods of planned case detection and follow up. Following that, health education programs were planned in the divisional level/MOH level in all MOH areas.

Two pre-intern doctors and 2 health volunteers were trained by the PI and pre-intern doctors were given additional training in the dermatology clinic, THJ. With the help of the MOH, PHI and PHM of relevant area, HHVs/CMCs were conducted in all the above areas after adequate publicity about the program in respective villages. Initially tentative schedules were prepared in a planning workshop at RDHS office with the participation of Supervising PHI and PHI of the selected areas. Proposed scheduled dates were shared with the field staff in advance through Medical officers of health. MOH and staff conveyed the message about the HHVs/CMCs in clinics and domiciliary visits. Information was placed on the public notice boards of the Community Centre/ public library one week before the respective area program. In some Medical officer of health areas a public announcement with loudspeakers were made on the day preceding the program.

The PHI of the selected locality prepared a map of the locality. Three teams headed by the trained pre-intern doctors and the PI visited the localities in the weekends with the help of area PHI, PHM and Grama niladhari. The same day morning a brief introduction was given to the teams by the PI with a power point presentation prepared in Tamil and using leaflets. After the introduction the teams visited the houses according to the road maps. After a brief talk with the residents they look for any skin lesions/deformities with their consent. If they found any suspected lesions, the particular person was asked to visit the pre-intern doctor/PI in a selected place (a preschool/community centre) in the locality for further evaluation and advice/ management. If the persons did not come to see the doctor, the doctor visited the respective house at the end of the program. The inconclusive/ probable leprosy suspected persons were advised to visit the dermatology clinic TH, Jaffna or to the monthly satellite clinic in BH, Pointpedro or DH, Chankanai. A template (data record sheet) was prepared in the program and shared with the MOH and the Kaveri Kala Manram (KKM-a local NGO work with leprosy affected people and The Leprosy Mission) in advance and used by the team.

Ethical considerations

Ethical clearance was obtained from the Ethical review committee of Faculty of Medicine, University of Jaffna.

Analysis

In the community (active case detection) survey, individual tables of the house visits/mobile clinics form were summarized by columns. The examined and suspected persons were summarized as percentages according to MOH areas.

Results

This section described the results of the house to house visits and mobile clinics in all 12 MOH areas. Table 1 describes the distribution of target population of the screening programmes according to the selected localities in MOH area in the aspect of visited houses and total residents in those areas. MOH Chavakacheri had the highest and MOH Velanai had the lowest target population (Table 1).

Table 1: Distribution of target population of the screening programmes according to MOH area in Jaffna district

MOH area	Number of screening programs	Localities of screening programs	Number of houses visited	Total members in family
Chankanai	03	Pandaveddai	69	336
		Kaddupulam	61	335
		Thikkirai(Chulipuram centre)	51	308
Sandilipay	02	Navali north	45	200
		Sahayapuram	42	248
Jaffna	02	Navanthurai, sea street	71	324
		Aaseervatham Rd, Nallur	69	272
Kopay	03	Urumpirai east	22	132
		Kalaimathy,Puttur west	54	405
		Urumpirai north	30	112
Nallur	02	Kondavil east	87	378
		Kondavil north	199	704
Point-Pedro	02	Suppermadam	95	440
		Punithanakar,Katkovalam	44	203
Karaveddy	02	Karaveddy west	47	267
		Valavanthoddam	36	150
Thellipalai	02	Konapulam	120	559
		Kuddiyapulam,Vasavilan	47	178
Chavakachcheri	03	Varani north	72	356
		Iyattalai	47	171
		Navatkulli	151	646
Kayts	02	Kalapoomi	52	233
		Piddiyellai	34	163
Velanai	02	Ward No.1	32	160
		Ward No.6	22	110
Uduvil	02	Thavady south	59	280
		Earlai west	30	122
Total	27		1688	7792

Coverage of the screening programmes

Table 2 describes the coverage of screening programme according to MOH areas. Overall 55.9% of 7792 target population was examined or screened. All the family members who were there during the visit of the team (irrespective of sex and age) were examined by the team members. MOH Sandilipay had the highest coverage examined and MOH Jaffna had the lowest coverage examined of the target population.

Table 2: Coverage of the screening programmes according to MOH area in Jaffna district

MOH area	Total members in family	Screened members	
		Number	Percentage
Chankanai	979	518	52.9
Sandilipay	448	316	70.5
Jaffna	596	257	43.1
Kopay	649	357	55.0
Nallur	1082	657	60.7
Point-Pedro	643	366	56.9
Karaveddy	417	226	54.2
Thellipalai	737	374	50.8
Chavakachcheri	1173	666	56.8
Kayts	396	220	55.6
Velanai	270	157	58.2
Uduvil	402	240	59.7
Total	7792	4354	55.9

Prevalence of skin lesions and suspected lesions

Table 3 describes the Prevalence of skin lesions and suspected lesions among screened population according to MOH areas. Out of 4354persons examined, 16.4% had some skin lesions and2.0% had suspected (as leprosy) skin lesions in the district. All these skin lesions were confirmed by pre-intern doctor or principal investigator. MOH Chankanai had the highest prevalence of skin lesions (24.1%) and suspects (4.1%) in examined population and MOH Karaveddy had the lowest prevalence of skin lesions (8.4%) in examined population. MOH Uduvil had no persons with suspected skin lesions.

Table 3: Distribution of persons with skin lesions and suspected lesions according to MOH area in Jaffna district

MOH area	Total number screened	Persons with skin lesions		Persons with suspected lesions	
		No.	%	No.	%
Chankanai	518	125	24.1	21	4.1
Sandilipay	316	67	21.2	11	3.5
Jaffna	257	25	9.7	04	1.6
Kopay	357	79	22.1	05	1.4
Nallur	657	78	11.9	14	2.1
Point-Pedro	366	65	17.8	06	1.6
Karaveddy	226	19	8.4	04	1.8
Thellipalai	374	79	21.1	05	1.3
Chavakachcheri	666	82	12.3	13	2.0
Kayts	220	37	16.8	04	1.8
Velanai	157	19	12.1	01	0.6
Uduvil	240	40	16.7	00	0.0
Total	4354	715	16.4	88	2.0

Follow up of persons with suspected lesions and new case detection

Table 4 describes the follow up of persons with suspected lesions and detected new leprosy cases among screened population according to MOH areas. Out of 88 suspected persons (as leprosy) with skin lesions 50% visited to dermatology clinic in the district. MOH Chankanai had the highest follow up and MOH Velanai had the lowest follow up rate of persons with suspected lesions. New cases were detected in MOH Chankanai (02), MOH Sandilipay(01), MOH Kayts (01)and MOH Kopay(01)among screened population. This is 6.4/10000 of target population (assume no cases among absentees) or 11.5/10000 of screened population.

Table 4: Distribution of persons with suspected lesions and follow up according to MOH area in Jaffna district

MOH area	Number of Persons with suspected lesions	Persons Visited to dermatology clinic		Number of detected New leprosy cases
		Number	Percentage	
Chankanai	21	12	57.1	02
Sandilipay	11	05	45.5	01
Jaffna	04	01	25.0	
Kopay	05	03	60.0	01
Nallur	14	06	42.9	
Point-Pedro	06	03	50	
Karaveddy	04	02	50	
Thellipalai	05	03	60	
Chavakachcheri	13	07	53.8	
Kayts	04	02	50	01
Velanai	01	00	0.0	
Uduvil	00	-	-	
Total	88	44	50	05

Discussion

The study has screened 56% of the population (7792) of selected localities and found that around 16% of them having some skin diseases and around 2.0% had suspected (as leprosy) skin lesions in the district. Out of them 5 new leprosy cases (rate of 11.5/10000 of screened population) were detected.

Though the community surveys were conducted in the weekends, only around 56% of the members of the visited family were screened. Most of the other members were absent even in the weekends as majority of the households had at least one person with daily paid works mainly in the agricultural fields, who may be the breadwinner of the family. In addition to that a few persons were doing some complicated household works when the team had visited their homes. Although the team members visited the same houses on the return of the day some people could not be examined that instance also. This may be due to lack of interest of people in some selected localities because of the repeated programs conducted by health staff or NGO in this way or may be a reflection of some level of stigma. But adequate precautions were taken in training of data collectors to make a comfortable conversation during the house to house visits to encourage the household members in the screening programs by role play and showing video clips of effective communication sourced from health education officer of RDHS office, Jaffna. The PI has involved fully in the training program. He shared the experiences and methods in communication from his professional experience, especially from the IYCF (Infant and Young Child Feeding) package, in which the PI was a key trainer in the Northern Province and from the training on Risk Benefit communication of immunization, in which the PI was a trainer of the district team as a Regional Epidemiologist. In most instances MOOH arranged an announcement previous day of the program with the help of public addressing systems in addition to previous information through the field staff.

To increase the coverage measures were taken in addition to the initial awareness to DS office staff and health staff, by giving a brief instructions to the team with the help of street map drawn by the area PHI and maintain at least 2 females including one female pre-intern doctor and one field staff or health volunteer of that area. To maintain the data quality, a small PowerPoint presentation prepared in Tamil language with pictures of common skin conditions, was done by the PI to the team members in each locality. To motivate the performance of team members, necessary provisions were made and a file cover containing data record sheets, details of the locality/map and mobile numbers of the PI and area PHI were given. For the whole day, the PI was available to clarify and attend the doubts or issues of the team members. Additional precautions were made when the programs held in areas where stigma issues were high according to MOH by considering the MOH ideas and advised the team members to use the terms carefully without upset the residents. They were educated by informing the advantage of screening in general than specifically looking for skin conditions. The pre-intern doctors used to start the talk with the general wellbeing of the household members and checking available medical records of other illnesses and CHDRs of the children. At the end of the each day program, the PI and the pre-intern doctor who coordinating the activities cross checked the data sheets and clarifications attended.

Only about 17% of screened population had some type of skin disease at the time of the examination. Though it was not a prevalence study with a representative sample of Jaffna district, the percentage of people with any type of skin disease was relatively low compared to a study done in Piliyandala, Sri Lanka, where the prevalence was around 48% [6].However, the present study recorded only treatable skin diseases or included only certain age groups, which was the first community survey of skin disease in Sri Lanka. Likewise in an Egyptian study reported prevalence of 87% [7]. The vast differences may be due to the study period where the Egyptian study was done in 1994-1996 and the Sri Lankan was done in 1997. The other reason may be the study setting and the seasons prevailed.

Another study done in Egypt reported a prevalence of 42 % among the children [8]. A prevalence study done in mountainous part of India shown 45% prevalence [9]. But in Gujrat, 10-20% of patients seeking medical care were with skin diseases in general practice [10]. A study done in Taiwan revealed nearly 18% of school children affected by common skin diseases [11]. But in Sri Lankan school children the prevalence of skin disease was around 14 per 1000 during the routine school medical inspection [12]. In these evidences, the prevalence reported in the present community screening was relatively a good valid measurement.

Among the 88 persons with suspected lesions, only 44 visited for the confirmation at dermatology clinic, Teaching Hospital, Jaffna by the follow up visits of

team members of data collection, staff of KKM and field health staff. Five patients were confirmed as leprosy and a few were requested further follow up for confirmation. As only 50% suspected patients visited for the follow up and some yet to visit, there is a possibility of more cases to be detected and equal chances for missing them. As the data was shared with the authorities, the outcome depends on the effectiveness of further follow up. Those areas with less follow up coverage were recommended for the routine mobile skin clinic of the health department, in which one of the consultant dermatologist used to visit. In addition to that, discussions were initiated with the MOOH for a sustainable program once in a month during weekends.

The five newly detected leprosy patients could be expressed as a rate of 6.4/10,000 of examined population or 11.5/10,000 of screened population. In either situation the rate is above the said elimination level of 1/10,000 population according to the WHO [13]. And the rate is well above the new case detection rate of 0.6/10,000 population of the district for 2015 [14] and of 1.04/10,000 population of Sri Lanka for 2014 [15]. Similarly a survey done in India reported the detection of more cases as a rate (4.4/10000) above the elimination level than the usual reports from government [16]. These results clearly demonstrate the ability of this method to detect more cases than the routine mobile skin clinics. But, in the previously mentioned study by Perera et al., leprosy was diligently looked for, and they found only one patient out of 1806 people, who also was under hospital treatment.

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Conflict of Interests

There are no conflicts of interest

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Original research Papers

Role of Sentinel Lymph Node Biopsy in Early Stages of Carcinoma of the Breast.

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Abstract:

Background

Sentinel node biopsy is the standard of care for the management of axilla in the early breast cancer, however it is not widely practiced in Sri Lanka. It minimizes the morbidity associated with axillary node dissection

Method

Retrospective analysis of the clinical records of 30 women who underwent SLNB in Onco-Surgical Unit, Teaching Hospital- Batticaloa during the period from May-2018 to December 2021.

Results

In this study 23 (76.7%) patients had invasive carcinoma (IBC) & 7 (23.3%) had ductal carcinoma in-situ. Out of the invasive carcinoma patients only 7 (30.43%) patients had positive sentinel nodes & underwent subsequent axillary clearance. Out of the axillary clearance patients (&) only 4 patients (57.14%) had residual diseases in the axilla. All the DCIS were node negative.

In this study sentinel node biopsy has avoided unnecessary axillary clearance in 23 (76.7%) patients and thereby prevented associated morbidities.

Conclusion

Majority of early breast cancer patients became sentinel node negative in this study. By employing this technique, we have avoided unnecessary axillary clearance & associated morbidities in (82.6%) of patients with invasive breast carcinoma.

Keywords:
Carcinoma of the Breast, Sentinel Lymph Node Biopsy, Axillary Nodal Dissection.

Introduction

Carcinoma of the breast is a major health burden in 21st century. Management of breast and axilla is ideal. Axillary lymph node management provide the prognostic information and help to eliminate the diseases from the axilla.

The immunologically vigilant node which drain the primary site of cancer is called Sentinel Lymph Node (SLN). SLNB has become the gold standard staging tool (Basnayake, Jayarajah and Seneviratne, 2018) for the management of axilla in early cancer minimizing the morbidity related to ALND (Iancu et al., 2021). The American Society of Clinical Oncology (ASCO) also recommend the SLNB in early stages of carcinoma of the breast (Giuliano et al., 2017).

Mapping of lymphatics with the combination of radioisotope and blue dye is ideal (Ahmed, Purushotham and Douek, 2014). The accuracy of detection rate is above 95 % in SLNB and the axillary recurrence after SLNB is less than 1 percentage in combined mapping (Jayatunge, 2012).

However, there are so many limitations to use radioisotopes such as storage, handling and waste disposal and therefore it is practically not feasible to practice in developing countries. Hence, mapping with blue dye (isosulfan/ methylene blue) alone is most commonly practiced. The sensitivity and specify of this technique is moreover similar to combine mapping (Mariani et al., 2001) and it is being practiced at Teaching Hospital-Batticaloa as well. The SLN identification accuracy was 100 % with combined mapping versus 98 % in blue dye alone and the false negative rate was zero in combined method while it was 5 % in blue dye alone (Hung et al., 2005).

Materials and Method

This retrospective descriptive single centered study was carried out in the Onco-Surgical Unit Teaching Hospital-Batticaloa which is a tertiary care center with fairly well equipped Onco- Surgical health care facilities.

Data of 30 women with early (T1, T2) tumor with clinically and radiologically negative nodes underwent SLNB during the period from May-2018 to December 2021 were retrospectively analyzed.

The 3-5 ml of methylene blue dye technique is practiced in this unit for the mapping of lymphatics and the dye is injected around the sub areolar plexus. Following that, 3-5 minutes of brief massaging is given to the breast to dilate the lymphatics. A separate axillary incision is made and SLNs are harvested with adjacent enlarged nodes aiming to include 4 nodes. No drains are inserted. Average time consumption for the SLNB in this unit is 25 minutes. Patients are discharged on the following day after the SLNB and it is cost effective when compare to ALND in which the patients need inward management for average 3 days due to drain.

Results

A total of 30 patients had been underwent SLNB. Mean age of the population was 53.7 years ranging from 27 to 72 years.

Seven (23.3%) were Ductal Carcinoma in Situ (DCIS) and rest (23= 76.7%) of them were invasive Carcinoma.

All the cases with DCIS were node negative. Among the 23 cases with Invasive Carcinoma Seven cases (23.3%) had positive nodes.

All seven node positive cases underwent subsequent ALND and four of them found (57.14 %) to have residual disease in the axilla.

SLNB has prevented unnecessary ALND and associated morbidities in 23 (76.7%) patients in this study.

Discussion

Carcinoma of the breast is a number one leading cause of cancer related morbidity and mortality among women.

Commonest breast cancer is Invasive Ductal Carcinoma (IDC) which accounts about 80 percentages of all the breast cancers while the second common is Ductal Carcinoma in Situ (DCIS) which accounts about 14 percentages.

Management of breast carcinoma comprises management of breast and axilla. The SLNB has significantly lessens the morbidities including lymphedema, shoulder joint stiffness, seroma, damages of nerves in the axillary region and upper limb numbness associated with conventional axillary lymph node clearance (Basnayake, Jayarajah and Seneviratne, 2018).

The accurate identification of sentinel lymph node is crucial in the early stages of carcinoma of the breast for the individualized optimal management of axilla which directly reflects the prognosis (Chatterjee, Serniak and Czerniecki,2015).

Intraoperative identification and histological confirmation is the gold standard for the identification of SLN. This study also reveals the vital role of SLNB in the prevention of unnecessary ALND and associated morbidities in the carcinoma of breast.

Conclusions

Majority of early breast cancer patients became sentinel node negative in this study. By employing this technique, we have avoided unnecessary axillary clearance & associated morbidities in (82.6%) of patients with invasive breast carcinoma.

Recommendation

Sentinel lymph node biopsy in early breast cancer should be widely adapted as it prevents unnecessary axillary clearance.

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Original research Papers

ROAD TRAFFIC INJURY WITH LIMB BONE FRACTURES AND SOCIO DEMOGRAPHIC STATUS IN EASTERN PROVINCE, SRI LANKA

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Abstract:

Introduction: In Sri Lanka, traumatic injuries with Road Traffic Accidents (RTAs) as a major component, is the leading cause of hospital admission and occupies the 11th rank in hospital mortality

Objective: To describe the socio demographic characteristics related to the road traffic accidents among patients with limb bone fracture following RTAs admitted to secondary and tertiary care hospitals in Batticaloa and Kalmunai RDHS area.

Methods: Hospital based descriptive cross-sectional study was conducted among 480 patients with limb bone fractures. Sample size was calculated using standard formula (1) and the proportionate sampling method was used. interviewer administered questionnaire was used (response rate 96%).

Results: Among the participants, 88.5% were males and young aged, and most were self-employed and married. Most of the accidents occurred in the evenings, mainly involving motorbikes, and followed by bicycle. Mostly injuring the drivers/riders. More than half of the RTAs occurred in carpet roads and the commonest type of RTA was a vehicle colliding with another vehicle. Lower limb bone fractures were slightly higher than of the upper limb. Only 6% received first aid before reaching the hospitals, no ambulance was used to transport to the hospital. During the hospital stay, majority had favorable attitudes towards the information provided by the health staff.

Conclusions. Majority of the vehicles involved in the RTA were motorcycles. Most of the injured road users were drivers / riders and lower limb fracture. No Ambulance services were not used to transport to hospital. Majority of patients were satisfied with health staff information and treatment towards fracture.

Keywords:
Road Traffic injury (RTA), Lower Limb, bone, fractures

Introduction:

Road Traffic injuries (RTIs) have emerged as a major global public health problem of this century and are now recognized as veritable neglected pandemic (2). The RTIs take the lives of nearly 1.3 million people per annum, and disable millions more (3). RTAs occur because of complex interactions between the road user, environment and vehicle. Several risk factors operate on these three components before, during, and after an accident resulting in a road traffic injury or death. Around 90% of deaths and injuries due to RTIs take place in low-income and middle-income countries. The South Asia region has a one fifth share in these fatalities (3). In South Asia, motorized traffic has been increasing at a rapid pace, typically over 10% annually in major urban areas. Pedestrians, cyclists, and motorcycle riders are most vulnerable and account for majority of traffic deaths in low- and middle-income countries. Also, vehicle population as well as the numbers of accidents has been steadily increasing with growing concern about its social and economic implications (4). RTAs have increased by 249% from 1977 to 2004 and RTI related fatalities increased by 160%. The RTI was ranked as the number one reason for admissions to government hospitals in 18 of the 23 districts of

the country and in rest of five other districts, it was always among the top five (5).

Limb injuries are identified as one of the major injuries. Lower extremity is the most frequently injured body region, accounting for 32.8% of all injuries, causing long-term impairment and disability, resulting in enormous social and economic impact (6). A recent study by Sampth and Seneviratne at National Hospital of Sri Lanka, found that limb fracture injuries are the commonest among RTAs victims with majorities are fractures (7).

Methodology:

This is a hospital based descriptive cross-sectional study conducted during the period 2015 – 2016. The study commenced on October 1st, 2015 and ended on 26th of February 2016. As study population, patients who were admitted with limb bone fractures following RTA were selected in a study setting of secondary and tertiary care hospitals in Batticaloa and Kalmunai RDHS areas, in Sri Lanka. Residing in the study area for more than 6 months & Aged above 18 years were taken as inclusion criteria and following were excluded, Re-admission for the same injury, Brain or spinal cord damage or vertebral bone fractures, and victims diagnosed with major psychiatric disorders due to reliability of the data.

Sample size was calculated by using the formula (1) to estimate a population proportion with specified absolute precision. Expected prevalence was taken as 50% and the sample was calculated as 384. Considering response rate would be 80%, the sample size was increased by 20% to account for non-respondents. The final calculated sample size was 460.

Proportionate sampling technique was used to select the samples. Interviewer Administrated Questionnaire (IAQ) was designed to collect information on patient identification, demographic and socio-economic details, type of RTA, circumstances and details of RTAs, safety practices related to RTAs, hospital care, and details from Medico-Legal Examination Form (MLEF) and Bed Head Ticket (BHT).

Results:

Among the participants 88.5% (n= 425) were males with median 31years, (IQR 21-46years). Of the participants 69.2% (n=332) were Tamils. Majority were Hindu 60.8% (n=292,). Of the participants had highest educational level up to GCE Ordinary Level. Out of the total victims 42.1% (n=202) were self-employed while 30.2% (n=155) were unemployed.

Table 1: Selected socio-demographic characteristics of the participants (N=480)

Characteristic	Number (N)	Percentage (%)
Ethnicity		
Tamil	332	69.2
Muslim	148	30.8
Religion		
Hindu	292	60.8
Islam	148	30.8
Catholic/Christian	40	8.3
Education level		
Never	23	4.8
Up to Grade 5	68	14.2
Up to O/L	234	48.8
Up to A/L	86	17.9
Occupation		
Self-employed	202	42.1
Paid work	133	27.7
Unemployed	155	30.2

Table 2: Distribution of day, time and type of vehicle in accident (n=480)

Characteristic	Number (N)	Percentage (%)
Day of the week		
Weekend	133	27.7
Week days	347	72.3
Time of the day		
Morning	170	35.4
After noon	88	18.4
Evening	180	37.5
Type of vehicle		
Motorcycle	346	72.1
Bicycle	69	14.4
Three-wheeler	48	10.0
Other vehicles	17	3.5

It was found 72.3% of RTAs occurred in week days. Evening was the peak time for RTA (n=180, 37.5%) and the morning was also almost the same as the evening 170 (35.4%). Motorcycle (n=346, 72.1%) was the most commonly involved vehicle (Table 2).

Table 3: Distribution of Type of Road-Users, Road, and RTA (N= 480)

Characteristic	Number (N)	Percentage (%)
Type of road user		
Driver/riders	253	52.7
Cyclist	67	14.0
Pedestrian	55	11.5
Passenger	105	22.9
Type of accident		
Vehicle colliding with vehicle	241	50.2
Vehicle colliding with pedestrian/animal	58	12.1
Pedestrian colliding with vehicle	56	11.7
Falling from vehicle	40	8.3
Rolling over vehicle	05	1.0
Other	80	16.7

Table3 Summarizes the distribution of type of road users, type of road and the type of accident.

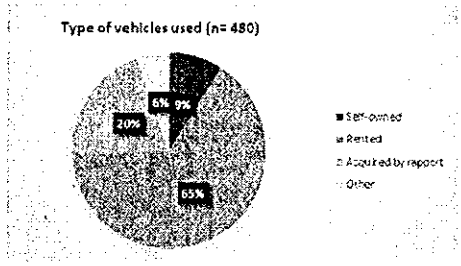
Drivers (n=253, 52.7%) were the commonest type of road users to be presented with limb fracture. Out of the victims of RTAs, the number of passengers (n=105, 22.9%) was nearly two times compared with the pedestrians (n=55, 11.5%). Nearly a half of the RTAs (n=241, 50.2%) occurred due to a vehicle colliding with another vehicle. More than 23% of the accidents were due to either a vehicle colliding with a pedestrian/animal or a pedestrian colliding with a vehicle (Table 3).

Table 4: Presence of selected safety practices (n=480)

Characteristic	Number (N)	Percentage (%)
Having a license		
Yes	218	45.4
No	35	7.3
Wearing a helmet		
Yes	247	51.5
No	63	13.1
Wearing the seatbelt		
Yes	2	0.4
No	8	1.7
Received first aid		
Yes	29	6.0
No	395	82.3
Could not remember	56	11.7

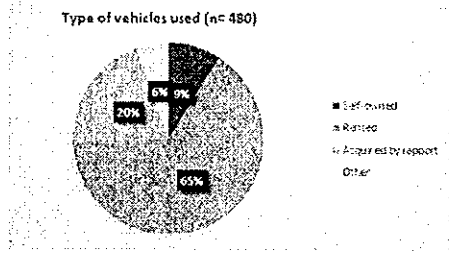
Of the respondents 45.4% (n= 221) were drivers with a valid driving license. Only half of the (51.5%; n= 247) riders of motor bicycles had worn a safety helmet. Drivers (n=253, 52.7%) were the commonest type of road users. Out of the victims of RTAs, the number of passengers (n=105, 22.9%) was nearly two times compared with the pedestrians (n=55, 11.5%). Of the accidents 56.9% took place on the carpet road (n= 273), followed by Tar roads (n=149, 31%). Out of the four-wheeler occupants, seat belt was worn by 0.4% (n=2) at the time of the accident (Table 4).

Fig1: Distribution of ownership of vehicle used to transport the patient to the hospital



The rented vehicles, (n=314, 65.4%) were mainly used to transport the fractured participants to the hospital following the accident. One fifth (n=96) were transported on vehicles provided on humanitarian grounds. Not a single ambulance was used to transport the patient to the hospital.

Fig 2: shows influence of alcohol during admission following RTA to the hospital.



Out of 480 participants 8.5% (n= 41) were documented as under the influence of alcohol at the time of hospital admission.

Table 5: Satisfaction of the participants on in-ward care received (n=452).

Category	Level of satisfaction				
	Very good N (%)	Good N (%)	Fair N (%)	Poor N (%)	Very poor N (%)
Conveying information by health staff	150 (33.2)	218 (48.2)	61 (13.5)	6 (1.3)	17 (3.8)
Treatment provided by the institution	140 (31.0)	188 (41.6)	73 (16.2)	38 (8.4)	13 (2.9)

Nearly 80% of the participants (n=368) were having a favorable level of satisfaction (very good or good) on conveying information by health staff and Nearly 73% (n=328) were having a favorable level of satisfaction on treatment provided by the institution.

Discussion:

Injuries due to RTAs are preventable. Although road traffic accidents are a major cause of death and disability worldwide and several risk factors have been identified for their occurrence, only15% of countries have comprehensive laws related to the five major risk factors: excessive speed, driving under the influence of alcohol, mandatory use of helmets by motorcyclists, usage of seat belts and safety seats for children (8). According to United Nation, the sixth target of Sustainable Development Goal 3 (Third goal) is to reduce the global deaths and the injury due to RTAs by half at the end of 2020 . In Sri Lanka, according to Annual Health Bulletin (2013) traumatic injuries (with RTAs as a major part) are the highest leading hospital admission and 11th rank in mortality (9). In developed countries, a standard set of inter¬ventions have contributed to significant reductions in the incidence of RTIs. These include the enforcement of legislation to control speed and alcohol consumption, mandating the use of seat belts and helmets, and the safer design and use of roads and vehicles.

There are studies from Sri Lanka (10-12) and several international studies (13-15) have described that, Limb bones were the most common type of injuries following RTAs. Trauma from traffic accidents was a main cause of disability in people of working age, especially among men. A study by Fitzharris and others revealed that disabilities due to injuries in RTAs cause medical, social, and economic problems and make a considerable influence on the quality of life (16).

The highest average was 35.3 yrs and men were involved in more than 70% of cases (17). According to the present study, majority (88.5%) them were males. Many of the respondents were married in civil status. This is like the findings of local studies of road traffic accident (7,10,12).

According to a study on characteristics of three-wheeler crashes by Amarasingha, most of the accidents have taken place during weekends than weekdays (18). At the same time the same author described on her study the characteristics of motorcycle crashes and motorbike risk. There she described that, motor bike accidents are more common in weekdays than weekends (18). In the current study, it was revealed that 27.7% of the accident responsibility for limb bone fractures occurred on weekends and majority (72.3%) in week days.

Sampath and Senaviratne study at National Hospital of Sri Lanka on direct road traffic injuries also found that more RTI took place during morning (22%) and evening (27%) and Dharmaratne study in Kandy also found some similar findings (7,10).

The majority type of a road user at the time of the accident was the drivers (52.7%). The next commonest categories were; then passengers (22.9%), cyclists (16%) and pedestrians (11.5%). Jeepara and Pirashanth also found more or less comparable results with drivers (45%), passengers (42%) and pedestrians (13%) being the different users in the descending order of the incidence (11). Another similar result has been described by Sampath and Senaviratne in which the motorbike drivers (44%), pedestrian (15%) and passengers (15%) were the commonest road users involved in RTIs (7).

When the type of accident is concerned, almost half (50.2%) of the participants stated that they met the accident because of a vehicle colliding with another vehicle. In another 12.5 % of the participants, the type was a "vehicle colliding with pedestrian/ animal" and a "pedestrian colliding with a vehicle" (11.7%). Due to the abundance of carpet roads and the study area is a mainly agricultural in occupation, there is a higher risk for the animals and pedestrians to get involved in RTIs. According to Tahera and others in a study in Bangladesh (19), it was revealed that hitting on pedestrian is the commonest type of accident (45%) followed by a vehicle colliding with another vehicle (29.7%). A possible explanation for this difference between the global and present study would be that though they studied all road traffic injuries, the current study was only to limb bone fracture.

Out of total 480 road traffic accident limb bone fractures 72.1% accidents reported motorcycle involvement, 14.4% due to bicycle involvement and 10.0% due to three-wheelers. Study by Jeepara and Pirashanth (11), in Batticaloa Teaching Hospital found that 71% of the total accidents of their study involved motor bikes. In another study by Weerawardane, in Anuradapura (20), it was found that the descending order of vehicle involvement

was motor bike (65%), push bicycle (11%) and then three-wheelers (11%). The present study and those results are comparable. This may be due to that in Anuradhapura, except ethnicity all other socio-economic characteristics are similar to the present study settings (21). According to department of transport and civil aviation publication in 2016, the distribution of vehicle involvement in RTIs, motor bikes are the commonest vehicle involved in 2015 (n=10147) and three-wheeler is the second commonest vehicle (n=6871). Meanwhile in a study by Phillipo in the African region, it was also found that motorbike (58.8%) was responsible for most of the road traffic accident (22).

Wearing a helmet is a law in Sri Lanka. Out of those involved in motorcycle accidents 79.7%, were wearing helmets. According to Jeepara and Pirashanth, 21% of participants had not worn helmet at the time of the accident and 35% were not wearing on a regular basis (11). The result of the current study was almost similar to that study done in Batticaloa. In another study by Sampath and Senaviratne (7), it has been stated that among motorcycle riders and pillion riders 93.8% and 80% were wearing helmets at the time of the accident respectively.

In Sri Lanka wearing seat belt in front seats of vehicle is mandatory from 2012 according to Motor Traffic Act (2011). In the current study 10 participants were traveling in four-wheeler motorized vehicles and only two of them (20%) had worn it. In a study by Fernando and others in the Central Province of Sri Lanka, it was found that 64% of drivers or front passengers did not wear seat belt and only 14% of drivers regularly wore the seat belt (23). According to WHO, in Argentina only 26% of front seated passengers in Buenos Aires wear seat belts. Higher percentage of not wearing seat belt (80%) of the present study points towards the inadequacies of implementation of the existing rules (24).

Receiving first aid before reaching the hospital is one of the key factors deciding the progression of the severity of the injury. The current study found that, only few (6%) received any sort of first aid before reaching the hospital. This finding is in concordance with the injury surveillance report of Sri Lanka, in which it has been mentioned that only 4.4% of injured victims had received first aid (25). The figure is mentioned as 5.1% in the study by Sampath and Senaviratne (7). It suggests the need to improve the pre-hospital care services in Sri Lanka urgently.

Following an accident there are many modes of transport to transfer the victims. The current study has found that, none of them were transferred via an ambulance. Meanwhile Sampath and Senaviratne (7), found that only 0.7% of the participants had been transferred in an ambulance. This result is also consistent with the findings of National Trauma Surveillance System in which it was mentioned that only 1% of patients were transferred in an ambulance (25). According to Sesser and others, the time factor is very crucial to raise the probability of survival among RTA victims (26). Therefore, many of them die or the injury is made worse by delays in transport and by incorrect postures while transferring. These preventable negative consequences can be minimised if a proper pre-hospital care system is available.

Conclusions:

Young self – employed males, most as the bread winner for their family with a monthly income between Rs. 20,000 to Rs 40,000 were the majority and most of the accident occurred during the time period of 6am to 12 noon and 2pm to 6pm of the weekdays. The main type of the road users were drivers/riders, the main vehicle involved in the RTAs was motorbike, majority of the RTAs occurred in carpet roads and the accident was a vehicle colliding with another vehicle. Lower limb bone fractures were slightly higher than upper limb bone fractures. The safety practices related to road traffic accident, almost half of the participants were having driving licenses and worn helmets and only one in twentieth have received first aid just after the injury. Ambulance services were not used to transport the victim to hospital. Majority of patients were satisfied with health staff information and treatment towards fracture.

Recommendations:

Post-accidental pre-hospital care services must be given adequate emphasis. A comprehensive plan must be laid which covers all aspects like training of the personnel for provision of first aid and having a proper patient transport system etc.

The monitoring mechanisms of the competency of drivers must be strengthened. Mechanisms like re-validation of the driving license as well as point-deduction systems in the of RTAs must be considered.

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Review articles

Request for an Inquest and Medical Officers – A Brief update

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Abstract:

Introduction: All around the world, when an individual dies, it is mandatory to register that particular death in the death registry. Sri Lanka also no exception needs to be recorded all the deaths. The identity of the person, cause of death, and circumstance are the prerequisites for death registration. Extensive investigations of the death are paramount to fill the above requirements[1]. The inquest is the procedure for investigating sudden deaths in Sri Lanka, a fact-finding exercise rather than a fault-finding inquiry[2]. It is framed on the model of the English coroner system; it is different from what presently exists in England. The office of a "Coroner" does not exist in our country. Instead, we have an Inquirer into sudden deaths (ISD). Such inquiry or an "inquest" is carried out either by a magistrate or an inquirer. ISD is appointed by the Secretary of the Ministry of Justice[3].

Laws and Regulations on Inquest
Statutes

1. Present Inquest system in our country is based on the Code of Criminal Procedure (CCP) Act No-15 of 1979. Statutory provisions about inquest procedure, the cases that have to be undergone inquiry, duties, and powers of the ISD are elaborated in Chapters II, III, and XI and sections 369 – 373 of the CCP[4].
2. Penal Code of Sri Lanka 1883 and amendments (1993, 1995. 2006)

Special circulars

Special circulars have been issued by relevant authorities from time to time to improve death investigation.

Circular NO- 04/2007 and No- 03/2004 of Ministry of Justice regarding the appointment, payments, disciplinary control of ISDs[5].

Circular NO- 03/2008 of the Ministry of Justice regarding inquests on maternal deaths, deaths from accidents, suicides, deaths from firearm injuries, and deaths where the cause of death is unknown[5].

Circular NO- 01/2010 of the Ministry of Public Administration and Home affairs points out the payments of the ISD[6].

DGHS also issued a letter dated on12-01-2011 stating about mandatory inquest and autopsy for maternal deaths[7].

Circular No. 01-25/2011 by Ministry of Health regarding conducting Post- Mortem Examination on Maternal Deaths[7].

Circular No- 01-25/2012 by Ministry of Heath regarding guidelines of pediatric autopsy following immunization-related deaths[8].

Government Gazette notification (23/09/2011)
This Gazette notification states recruitment and qualifications of ISD[9].

Relevant judgments by a court of law

Seneviratne vs. Attorney General (71 NLR 429), known as Dodampe Mudalali's case, Justice Tennakoon pointed out A magistrate or an inquirer has no power to pronounce a verdict at the conclusion of an inquiry, and the law does not permit the Supreme Court to quash the findings of an inquirer or magistrate[3].

Indication to request for an Inquest

According to the CCP and other statutes, the following deaths must be requested for an Inquest.

1. All natural deaths where the cause of death is not known.
2. Natural deaths with known cause of death, but any suspicion arises.
3. All the deaths on admission at POD irrespective of the circumstances of the death.
4. Accidents include road traffic accidents, industrial accidents, domestic accidents, aviation accidents, naval accidents, and locomotive accidents.
5. Deaths due to suicidal acts.
6. Deaths due to violence (Homicide).
7. All deaths in custody include inmates of prisons, police custody, lawful and unlawful detention, patients in the mental hospital, leprosy hospital, TB hospital, elder's homes, children's homes, certified schools, etc.
8. Death due to medical, surgical, or anesthetic procedures immediately afterward.
9. Death following administration of blood, blood products, or a drug.
10. Death due to animal bites, rabies, or tetanus.
11. Maternal deaths
12. Vaccine-related deaths.

Duty of medical officers about Inquest procedure

Medical officers should request an Inquest if aa death occurs in one of the categories mentioned in the above list. The medical officer should inform the police to arrange an inquest. At the same time, authorization from the head of the institution should be obtained. It is of paramount importance to document every step in the BHT. A death that has occurred within 24 hours after admission to the hospital need not go for an inquest provided the cause of death can be determined[2].

The medical officer should not fill out the death declaration form when an inquest is requested. If they fill out the death declaration form, it will create confusion among ward staff, and sometimes they may release the body that has to be subjected to an inquest. A death declaration form should be filled out if the medical cause of death is known and the death is natural. A probable cause of death can be given even an inquest has been requested in natural deaths. Still, in unnatural deaths, medical officers refrained from providing the likely cause of death.

Under subsection (5) of section 370 of CCP of Sri Lanka have clearly stated the powers of the ISDs. ISD/Magistrate has empowered with summoned any person for the inquest. Medical officers also can be called by the ISD/Magistrate for inquest procedures. Medical officers must comply with such summon, and failing to do so would amount to contempt of courts[4].

Deaths that can be missed to request for an inquest

In medical wards, poisoning-related deaths, trauma patients take over for medical treatment from the surgical wards, and trauma patients get readmitted for a medical condition that the medical officers can easily overlook. Patients stay in the ward for a prolonged period in surgical wards, such as burn patients, head injury, spinal injury, etc. Patients retransfer from another hospital for conservative management also can be missed.

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Review articles

Radioactive iodine (131I) - A Therapeutic Summary

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The artificial production of new radioactive elements including iodine isotopes was a breakthrough in 1934 and the use of 131I began in the 1940s for medical purposes. Saul Hertz was the first physician who used radioactive iodine in January, 1941 with the purpose to treat diffuse toxic goiter patients. Samuel Seidlin was first who used radioactive iodine in March, 1943 to treat patient with differentiated thyroid cancer.

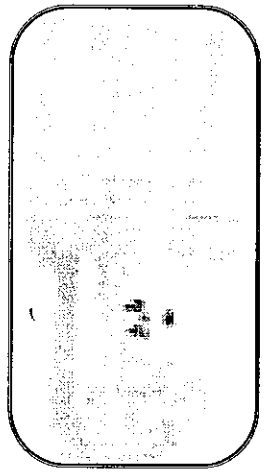
Physical property and mechanism of action

^{131}I is produced by the fission of uranium or by the neutron bombardment of stable tellurium in a nuclear reactor. The 131I isotope has a half-life of 8 days and this is an ideal property in therapeutic setting. The longer half-life is therapeutically beneficial as it stays in thyrocytes for a longer period. It emits both β and γ radiation. Therapeutic effects of the isotope are obtained by the emission of β radiation and γ radiation is utilized for imaging purposes. The pathological length of β radiation is approximately 1-2 mm and this destroys the thyroid follicular cells without damaging the nearby structures.

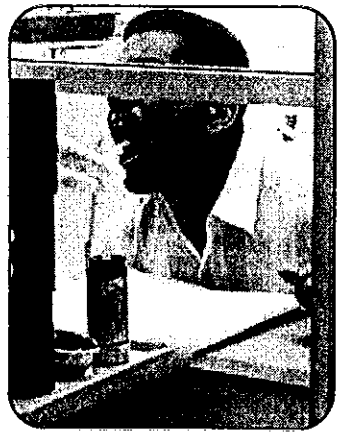
Radioactive iodine resembles the dietary iodide uptake in the thyroid gland and taken up by the sodium-iodide symporter mechanism. The absorbed 131I will stimulate the formation of free radicals, which destroys the DNA leading to cell death. High activities of radioactive iodine used in therapy is adequate enough to cause cell death and leading to a reduction in thyroid volume, function.

^{131}I preparation and routes of administration

The radioactive iodine preparation is available in liquid form as oral solution or injectable form and gelatin capsule. There is no pharmacological or therapeutic difference between these preparations. Radioactive iodine is routinely given orally for patients who can swallow without any difficulties and in whom the vomiting is not a problem. (Figure1) Intra venous route is reserved for patients who have vomiting as a problem.



[a]



[b]

Figure 1 [a]: 131I gelatin capsule, [b]: 131I oral solution [Picture courtesy: Nuclear Medicine Unit, Faculty of Medicine, University of Peradeniya]

Indications and Contraindications to 131I therapy

131I therapy is indicated in Graves' disease, Toxic Multi Nodular Goiter and Solitary Toxic Nodule. Common indications among these patients include; failure or intolerance or contraindication to anti thyroid drugs, patients with recurrent hyperthyroidism, thyrocardiac syndrome and for patients unfit or refusal to surgery. 131I is used as a theranostic agent in the management of differentiated thyroid cancer (DTC). (Figure 2)

The absolute contraindication to 131I therapy is pregnancy and pregnancy should be excluded before planning the 131I therapy. Breast feeding is a relative contraindication. Other relative contraindications in hyperthyroidism are: poor iodine uptake by the thyroid gland, severe hyperthyroidism with high hormonal values, presence of "cold" nodules in scintigraphy evaluation (FNAC indicating a benign lesion) and, infiltrative orbitopathy in Graves' disease.

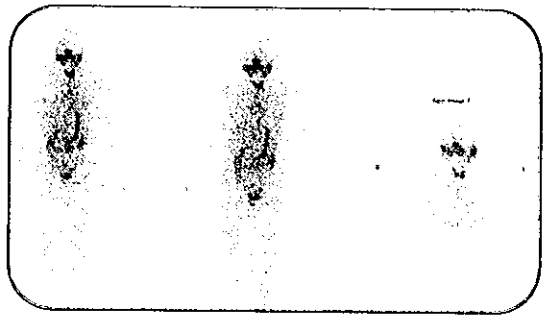


Figure 2: Post therapy scan showing residual thyroid tissues in a patient with DTC after total thyroidectomy [Picture courtesy: Nuclear Medicine Unit, Faculty of Medicine, University of Peradeniya]

Side effects of ^{131}I therapy

Long-term studies on radioactive iodine therapy have shown no evidence of teratogenicity or infertility in women. The absolute risk of carcinogenicity and mortality are very minimal or negligible. 131I therapy is generally well tolerated. Post-therapy hyperthyroidism and thyroid swelling can occur in patients with hyperthyroidism after treatment with 131I. Sialadenitis can develop in the parotid glands as a side effect compared to the submandibular glands.

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Review articles

Lipoprotein(a) and coronary artery disease

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Abstract:

Introduction: Lipoprotein(a) [Lp(a)] is a genetically determined molecule of low-density lipoprotein containing an apolipoprotein component1. Diet or environmental factors do not appear to exert a significant influence on its serum concentrations2.

Plasma Lp(a) concentrations are mainly determined by the LPA gene, race and ethnic factors significantly influence serum concentrations. Levels have been found to significantly vary between individuals and populations, ranging from 0.2 to 200mg/dl. In general, higher concentration are noted in populations of African descent compared to Asian, Oceanic, or European populations2.

This molecule was first discovered in 1963 by Kåre Ingmar Berg, a Norwegian professor in medical genetics3. Subsequently many studies have shown that LP(a) is an independent risk factor for atherosclerotic cardiovascular disease. Population studies have revealed increased levels of Lp(a) in a general population is associated with increased risk for myocardial infarctions.

Pathogenicity

Lp(a) appears to exert its pathogenicity through three modalities4

1. Inflammation

Inflammation plays an important role in plaque formation and rupture. Lp(a) appears to be susceptible to oxidative modification and produce an oxidation specific epitope. This epitope is an important mediator of the inflammatory process

2. Atherosclerosis

Atherosclerotic plaques develop as a result of an inflammatory process leading to a sub endothelial accumulation of lipid laden cells. Lp(a) appears to augment this process resulting in significant disease occurring at an earlier age.

3. Thrombosis

Under normal conditions the coagulation system maintains a fine homeostatic balance. Lp(a) appears to encourage thrombosis through inhibiting the fibrinolytic process by preventing plasmin- mediated thrombolysis. High concentrations of Lp(a) may even inhibit to some degree the efficacy of thrombolytic medications, resulting in poor outcome with therapeutic thrombolysis.

Current and emerging therapies

Therapeutic options currently employed for lipid lowering therapy do not provide an adequate reduction in Lp(a) levels. Existing management guidelines do not offer definitive recommendations on addressing Lp(a) levels. This is primarily due to the fact, that there were no proven medications effective at significantly reducing Lp(a)to desirable levels. Several treatment modalities are available and are being used to treat these patients.

• PCSK9 Inhibitors

PCSK9 inhibitors have demonstrated a reduction in LP(a) levels by around 25%. It can be used as a monotherapy or in combination with Niacin5. However more data and studies are required on this medication.

• Inclisiran

Inclisiran is a small interfering RNA (siRNA)which is capable of inhibiting PCSK9 protein synthesis in the liver. Initial studies indicate that this medication may provide similar reductions in Lp(a) levels as compared to PCSK 9 inhibitors1

• Niacin

Although Niacin is effective in producing a moderate reduction in Lp(a) levels (15-25%), it is no longer recommended as monotherapy as studies failed to demonstrate any cardiovascular benefit1. Also due to potential adverse reactions, recent guideline do not recommend the use of niacin as a way to reduce Lp(a)4

• Atorvastatin

A meta-analysis done on Atorvastatin shows that there may be some benefit in administering this medication6.

• Lipoprotein apheresis

Another therapeutic option available is lipoprotein apheresis. In countries such as Germany elevated levels of Lp(a) is considered an indication for extracorporeal lipoprotein apheresis1,6

Future perspectives

In Sri Lanka we are facing a progressively younger cohort of patients with coronary artery disease. Many patients do not have conventional risk factors. Some have a significant family history for coronary artery disease. It would be interesting to know if Lp(a) plays a significant role in the pathogenesis of these patients.

Furthermore, population studies will help to determine the Lp(a) plasma concentrations in the Sri Lankan population. This may have implications for future treatment and preventive strategies.

Also, as currently there isn't any effective and easily administrable conventional medication available to reduce serum Lp(a) levels, a window of opportunity is available to consider research on traditional medicines in this area.

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Case Reports

A Case of Chickenpox complicated with of Post-infectious Thrombocytopenic Purpura

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Abstract:

Introduction: Chicken pox is a self-limiting benign viral infection; occasionally, it may cause complications (1-3). Mild thrombocytopenia associated with chickenpox is well described in pediatric population (around 1-2%), but severe thrombocytopenia with bleeding is rare.

The incidence of idiopathic thrombocytopenic purpura among children with varicella is estimated to 1:25000 and ITP associated with varicella accounts for 1.9% of pediatric ITP cases (4).

We report a case of severe thrombocytopenia, possibly immune mediated bleeding in the form of epistaxis in a child recovering from chickenpox, who is successfully treated with intravenous immunoglobulin.

Case report

A 2 year-old girl was presented to us with the history of epistaxis for one day duration. The girl had a history of fever for 2 days followed by pleomorphic vesicular skin lesions 6 days back. There was no history of cough, vomiting, icterus and bleeding from other sites. The patient was afebrile and recovering from chickenpox.

On examination the patient was conscious and oriented, with pulse – 120/min, respiratory rate – 24/min, temperature 370C by axilla, blood pressure 100/70 mmHg and maintaining 99% saturation on room air. The patient had numerous dried scabbed lesions scattered over the body and few petechiae over both upper and lower extremities. There was no active bleeding after admission. There was no hepatosplenomegaly, lymphadenopathy, ecchymosis, bruises, bony tenderness and no past and family history of bleeding disorders or transfusion of blood components.

Laboratory investigations were as follows.

Investigations	05/08/2021	05/08/2021	06/08/2021	07/08/2021
Hb	12.7g/dl	12.2g/dl	11.7g/dl	10.7g/dl
HCT%	34.2	34.8	33.3	31.0
RBC (×106/micL)	4.56	4.09	4.34	3.90
MCV (fL)	74.9	81.4	80.1	79.6
MCH (pg)	27.8	27.5	26.8	27.5
MCHC (g/dL)	37.1	38.8	33.5	34.6
WBC (micL)	11000	11820	4960	7280
PLT (×103/micL)	16000	3000	12000	77000
RET%		1.44%		
B.Urea (mmol/L)		4.9		

S.Creatinine (4.3mmol/L)		34		
PT/INR (Seconds)		13.9/1.07		
aPTT (Seconds)		38.3		
CRP (mg/L)		1.6		
ESR (mm/1st hr)		12		
ALT (U/L)		35		
AST (U/L)		22		
Na+ (mmol/L)		138		
K+ (mmol/L)		3.9		

The blood picture was reported as marked thrombocytopenia with few reactive lymphocytes.

The child was treated with intravenous immunoglobulin 1g/kg per day for 2 days. As a result of treatment, there was no new rash on the skin, and platelets began to increase gradually and reached the level of 77 x 103/microL on clinical blood analysis on the 3rd day of treatment. The child was discharged from the hospital in satisfactory condition on the 4th day. The patient had normal FBC on follow-up examination with the platelet count of 550 × 103/MicroL in a week after.

Discussion

Our case demonstrates that thrombocytopenia is one of the rare but well-known complications of primary varicella zoster infection and its early diagnosis is essential for appropriate management of the patient.

Although various mechanisms have been implicated in the pathogenesis of Post-varicella ITP, including decreased bone marrow production of platelets, disseminated intravascular coagulation, virally induced platelet aggregation followed by phagocytosis or lysis, direct viral invasion of platelet precursors and viral-derived neuraminidase causing enzymatic desialylation of platelets followed by removal of abnormal platelets by reticuloendothelial system, the main mechanism implicated is immune-mediated platelet destruction (5-8). Platelet-specific IgM antibody against various platelet surface glycoproteins (GP), namely, GPIb, GPIIb, GPIIIa, and GPV were found in patients with varicella-associated thrombocytopenia by immune blotting technique (6,9).

Today, it is proved that the mechanism of Post-varicella purpura fulminant is thought to be due to development of anti-protein S antibodies. These antibodies lead to acquired transient severe protein S deficiency, which results in disseminated intravascular coagulation and micro vascular thrombosis (10). Treatment is symptomatic, including fresh frozen plasma to treat protein depletion, anti-thrombin III and heparinisation against thrombus formation, and anti-inflammatory drugs (steroids) (11). However, new therapies such as intravenous prostaglandin E and prostacyclin are being introduced (3).

Both corticosteroids and intravenous immunoglobulin can be used to treat varicella-induced thrombocytopenia. Corticosteroids during the incubation period of varicella can lead to the development of visceral varicella, so they should be used in recovery phase past the incubation period (5). Studies found early platelet count recovery within three months of intravenous immunoglobulin treatment, predicting a short duration of disease and favorable outcome in children with newly diagnosed ITP (12).

Diagnosis of ITP is based on findings of clinical examination and peripheral blood smear in all cases, and bone marrow aspiration in some. Bone marrow examination is not indicated in acute ITP with typical features, which includes, Hb level of >10 g/dL (6-12 months of age) or >11 g/dL (older than 1 year), total WBC count of >5 × 109/L (6-60 months of age) or >4 × 109/L (>6 years), neutrophil count of >1.5 × 109/L (6 months-6 years) or >2 × 109/L (>6 years), and a platelet count of <50 × 109/L (13). Children who did not meet the above criteria are categorized as atypical ITP for whom we must perform a bone marrow examination.

Although thrombocytopenia can be a finding of severe varicella, isolated thrombocytopenia in otherwise healthy children who are recovering from chickenpox has an excellent prognosis (5). This complication could be avoided by anti-varicella zoster virus vaccination (14,15).

In our case, the child has thrombocytopenia when she was recovering from the illness. RBC

and WBC counts and coagulation profile were within normal limits range. Rapid improvement in thrombocytopenia with immunoglobulin treatment is suggestive of immune mediated mechanism of thrombocytopenia.

Conclusion

Thrombocytopenia is commonly reported with varicella having various immune and non-immune mechanisms with an overall excellent prognosis, but it can also lead to life-threatening bleeding complications such as fatal intracranial hemorrhage.

The resulting positive effect of intravenous immunoglobulin for post-infectious ITP allows us to recommend its use in this state. However, further research is needed to clarify the mechanisms of development of this complication.
As some studies found the varicella vaccination could prevent this outcome, we suggest to consider vaccination against varicella for children.

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Case Reports

A case report of kawasaki disease in a child with dengue haemorrhagic fever

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Abstract:

Introduction:
Kawasaki disease is a medium vessel vasculitis where highest incidence occur in Asian children. Although the etiology remains elusive, certain clinical features support an infectious origin [1].

As some other infections are being proved and dengue infection is rarely been reported, further studies would be required to conclude whether dengue is an etiological agent for the Kawasaki.

Dengue is the most common arbo viral infection in Asia. Dengue hemorrhagic fever , Dengue shock syndromes are severe forms of the disease present with an acute febrile episode to plasma leakage followed by recovery with the rash, rise in the platelet count , resorption of the fluids.

Since Dengue as well as Kawasaki presents with fever and rash , their co-occurrence may pose a clinical dilemma [5]. However, it is uncommon to have two diseases occurring in a patient at the same time.

Kawasaki disease shock syndrome is a rare haemodynamically unstable phenomenon in the acute stages of disease. It can also present without any of the typical features. So consideration of this phenomina is difficult as it clearly mimics dengue shock syndrome.

Case presentation

A three year old , previously healthy Srilankan muslim boy presented with the history of fever of five days and features suggestive of Dengue fever. He was monitored for dengue febrile phase and on day seven of illness noted to have USS evidence of leaking. Critical phase monitoring was done and he was afebrile for a day. At the same time child developed an erythematous generalized rash with the right hand swelling. It was initially considered as dengue recovery rash with the cannula site infection. On day nine child developed multiple joint pain and cracked lips. Erythematous,tender,warm extremities were also noted by day ten with stepladder pattern of fever. IV Cefotaxime was empherically started after the blood culture.

On day thirteen mucosal changes ,bilateral cervical lymphnodes (R>L) were noted along with persistent fever spikes. Mother reveled there was nonpurulent conjunctivitis in the initial two days

Although the initial investigations were suggestive of Dengue haemorrahic fever, by day twelve there were leukoctosis, anemia, thrombocytosis with elevated transaminases , hyponatremia, hypo albuminemia, elevated CRP and ESR. USS abdomen showed no evidence for hydrops of Gallbladder. 2D echo was done which were normal twice in a week interval.

As the child fulfilled the criteria for typical Kawasaki disease ,IVIG was started with the high dose of aspirin until being afebrile for 48hours followed by low dose of aspirin for the next 8 weeks.

Although clinical improvement was seen he had persistent arthritis involving Knee, Hip ,cervical joints and high ESR for eight weeks. Expert opinion was taken from Consultant Paediatric rheumatologist inview of excluding any other rheumatic disease probabilities. He was kept on NSAIDs for six week and the ESR was monitored.

There are documented cases of Kawasaki disease with prolonged arthritis.

Both clinically and biochemically complete recovery was noted by eight weeks of illness with the normal 2D Echo. He is now being followed up at general paediatric clinic once in three month without any complications

Discussion

This child presented during the dengue outbreak at our district. Not only the fever, but also the leukocytopenia, thrombocytopenia , tender hepatomegaly and USS confirmed plasma leakage led us to suspect dengue. Dengue infection was also confirmed by dengue antibodies. Recovery from dengue was also satisfactory according to the clinical improvement.

Although the new fever spike after the clinical improvement was considered initially as possible secondary bacterial infection then the diagnosis of Kawasaki disease was made as the child fulfilled the criteria. Recovery with clinical improvement was noted with treatment where high ESR and the persistent arthritis settled after eight weeks of illness.

Even though dengue fever triggering Kawasaki is a rare presentation, this case is one of those significant presentation.

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6. Kawasaki Disease in a 2-year-old Child with Dengue FeverAnbazhagan Jagadeesh1 & Sriram Krishnamurthy1 & Subramanian Mahadevan1

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Case Reports

An uncommon cause of dyspnea in rheumatoid arthritis. The rheumatoid lung nodules.

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Abstract:

Introduction: Rheumatoid arthritis (RA) is a multisystem inflammatory disease with erosive arthritis and extra-articular manifestations. Thoracic manifestations of RA have a wide spectrum starting from the cricoarytenoid joint arthritis down to alveolar level, pulmonary vasculature and pleura. Rheumatoid lung nodules (RLN) are a distinct form of RA associated lung disease. We present a middle aged female with seropositive RA and RLN.

Case report

A-59-year old female with seropositive RA for two years on methotrexate and low dose prednisolone; presented with progressively worsening shortness of breath on exertion for two months. Her joint symptoms were in remission. She denied symptoms suggestive of other connective tissue diseases. History was negative for orthopnea, paroxysmal nocturnal dyspnea, ankle swelling, easy fatigability, constitutional symptoms, hemoptysis, recurrent sinusitis, skin rash, contact or past history of tuberculosis and occupational exposures to silica or coal.

Physical examination revealed an averagely built lady without fever, pallor, lymphadenopathy or ankle edema. She had no signs of active joint disease, joint deformities or subcutaneous rheumatoid nodules. Signs of other connective tissue diseases were absent. Respiratory system examination revealed no evidence of interstitial lung fibrosis, pleural effusion or exertional desaturation. Cardiovascular system didn't reveal signs of heart failure or pulmonary hypertension. Organomegally or focal neurological signs were not present.

Complete blood count showed a neutrophil leukocytosis, thrombocytosis and a mild normochromic normocytic anemia. Blood film was suggestive of anemia of chronic disease. Erythrocyte sedimentation rate and C-reactive protein were raised. Blood culture, urine culture and procalcitonin were negative. Chest radiograph appeared normal. Transthoracic echocardiogram didn't reveal evidence of cardiac failure or pulmonary hypertension. Metabolic panel including electrolytes, renal and liver functions were within normal limits. Rheumatoid factor (RF) and anti-cyclic citrullinated peptide (anti-CCP) titers were raised suggestive of active RA. Antinulcear antibody, extractable nuclear antigen panel, antineutrophil cytoplasmic antibodies and angiotensin converting enzyme were negative. Full lung function test didn't show a restrictive airway pathology. High resolution computed tomography (HRCT) of chest showed 4 calcified nodules located peripherally and subpleurally in right upper lobe. Three nodules were 10mm x 8mm and the other was 5mm x 4 mm in size. (Figure 01) They didn't show central cavitation, necrosis, pneumothorax or empyema. Apart from mild traction bronchiectasis involving right upper and middle lobe, rest of the lung parenchyma appeared normal without radiological features of RA associated interstitial lung diseases, organizing pneumonia or mosaic attenuation. Sputum pyogenic culture, fungal culture and acid fast bacilli were negative.

Figure 01 showing peripheral subpleurally located RLN

Bronchoscopy didn't reveal endobronchial lesions. Bronchoalveolar lavage (BAL) differential cell count revealed a neutrophil predominance. Malignant cells were absent. BAL pyogenic culture, fungal culture, gene-xpert for tuberculosis (TB) and aspergillus galactomannan antigen was negative. Bronchial brush and transbronchial biopsy from right upper lobe revealed inflamed bronchial mucosa without granulomas or malignant changes. A lung biopsy was not done due to the risk of progression of the underling lung pathology.

Broad spectrum antibiotics were commenced at the outset due to raised inflammatory markers with background immunosuppression. Antibiotics were continued for ten days. Following a diagnosis of RLN with the negative septic screen, antibiotics were omitted. Doses of methotrexate and prednisolone were escalated. She had a marked improvement in her symptomatology. Currently she is closely followed up at pulmonology clinic and awaiting a repeat radiological evaluation.

Discussion

RLN are a rare form of RA affecting the lung with an incidence of less than 1%. [1] Risk factors for the development of RLD includes; male gender, smoking, subcutaneous rheumatoid nodules, positive RF and antiCCP. [2] RLN can either develop without any temporal relationship to the disease activity or rarely precede the joint disease. Majority of RLD are asymptomatic, though RLN can become secondarily infected, undergo cavitation, rupture into pleural space leading to pneumothorax, pyopneumothorax and bronchopleural fistula formation; giving rise to cough, dyspnea, hemoptysis and fever. [3] Chest radiograph usually appears normal. HRCT demonstrates single or multiple nodules, located subpleurally and peripherally, in upper and middle lobes of the lung. Size of nodules vary from 0.5mm to 7cm. Differential diagnosis of RLN includes; granulomatosis with polyangitis, lung metastases, tuberculosis, histoplasmosis, sarcoidosis, amyloidosis and silicosis. Multiplicity, smooth border, cavitation and satellite nodules favors RLN over metastases. [4] In the absence of complications, RLN requires no specific treatment although in certain instances; treatment of articular disease with biological agents such as rituximab have shown to regress the size of nodules. [5]

Conclusion

RLN are a distinct form of RA associated lung disease. Majority of RLD are asymptomatic although emergence of complications can give rise to morbidity.

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Case Reports

A vegetable-induced severe hemolytic crisis in a possible G6PD Deficient child

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Abstract:

Background : Glucose-6-Phosphate Dehydrogenase (G6PD) is the first enzyme of pentose phosphate pathway (1) and the only source of reduced nicotinamide adenine dinucleotide phosphate (NADPH) required by the red blood cells to combat oxidative stresses (2). G6PD Deficiency is caused by point mutations in the highly pleomorphic G6PD gene in the X chromosome. Over 400 G6PD single nucleotide polymorphism (SNPs) have been identified with 186 SNPs being associated with the loss of G6PD activity and stability (3). Since the G6PD deficiency is X-linked, males are classified as either normal or deficient in G6PD whereas females classified into three G6PD phenotypes, normal, intermediate and deficient (4).

It is the commonest enzyme deficiency worldwide affecting 400 million people (5, 6). Globally, the highest prevalence of G6PD deficiency occurs in Sub-Saharan Africa where the malaria is also prevalent (3). It is more prevalent in regions where malaria was endemic in Sri Lanka as well as malaria endemic regions in the Middle East, Mediterranean and South East Asia (5-8). Due to this overlap, it has been suggested that G6PD deficiency may have arisen from negative selection by malaria (9, 10). The overall prevalence of G6PD deficiency in Sri Lanka is less than three percent of the population (6). But in some villages where malaria has been endemic the frequency of occurrence is high as 20.9% (6).

G6PD deficient people are usually asymptomatic until exposure to an oxidative stress in the form of a drug (Primaquine), a vegetable (Fava beans), a cosmetic (Henna), a household chemical (Naphthalene) or an infection that can result in acute hemolytic anemia (5-8). The basic management strategy in G6PD deficiency is avoiding oxidative stresses (5). However blood transfusions and iron and folic acid supplements are administered following an episode of acute hemolysis (5).

Acalypha indica (fig.1) a tropical plant that grows various parts of the world including Sri Lanka, is used in herbal medicine and as a local food (10). Intravascular hemolysis has been reported in humans with G6PD deficiency after ingesting this plant (11-13) and one case report also described methemoglobinemia (14).

We report a case of a nine year old boy presented with classic symptoms and signs of an acute hemolytic crisis following ingestion of acalypha indica, later diagnosed with G6PD deficiency.

Case Report

A nine year old school going boy was transferred from a local hospital to our institution with the history of fever, headache and dark urine for one day duration. The boy was apparently well before this illness and denied past history of similar events. There was no history of upper respiratory tract infection or skin sepsis with in the past month. He denies history of contact with muddy water or soil polluted with animal excreta.

He is the elder son of a non-consanguineous parents. The younger sibling who is also a boy (6years) is doing well. He was delivered by normal vaginal delivery with a birth weight of 3.5kg. His newborn period is uneventful and not complicated with neonatal jaundice. He denied any diagnosed hematological disease affecting his brother or relatives from the maternal side of his family.

He was managed for few episodes of respiratory tract infections at outpatient department until the age of 2 years. Since then he was healthy and not admitted to hospital for any reason. He is developmentally normal with a good school performance.

Upon examination there was severe pallor and lemon tinge icterus (Fig. 2). His pulse rate was 118/min, blood pressure was 100/70 mm mercury, his lungs were clear to auscultation and his liver was felt 1cm below the right costal margin and spleen was also felt 1cm below the left costal margin. The findings of precordial and nervous system examinations were normal. A urine sample was obtained from him clearly indicated hemoglobinuria as shown in fig. 3.

Further details revealed that he had eaten cooked leaves of acalypha indica one day prior to this admission. He denied ingestion of any other drugs or food that may induce hemolysis.

We started a workup for the possibility of hemolytic anemia. Table 1 summarizes his laboratory investigation results.

Blood picture showed normocytic normochromic red cells, polychromatic cells, spherocytes, bite cells and blister cells, suggestive of an oxidative hemolysis.

A hemolytic crisis triggered by acalypha indica in a G6PD deficient person was diagnosed. A consultant hematologist's opinion obtained and packed cell transfusions were given twice during his ward stay. He was also started with an oral macrolide (Clarythromycin 500mg twice daily for 7 days). His urine color was cleared on 5th day after admission (fig. 4). Once his symptoms subsided on 6th day after admission he was sent home with adequate doses of ferrous sulphate, ascorbic acid and high dose folic acid (5mg daily) supplements and with a list of drugs and meals to be avoided.

Reticular cell count two weeks after discharge has returned to normal. He was again referred to hematologist for the confirmation of diagnosis.

Table 1

Test	Reference range	Day 01 (at LH)	Day 01 (at THB)	Day 02	Day 03	Day 04	Day 05	Day 17
WBC (×109 /l)	4.00 – 12.00	11.12	9.01	9.93	10.58	8.39	8.39	5.89
Neutrophils%	50 – 70	61	64.4	54.1	56.7	47.4	44.8	
Lymphocytes%	20 – 60	31.4	26.5	32.4	30.3	39.2	43.0	
Hemoglobin (g/dl)	12 – 16	6.8	5.9	5.6	8.5	9.2	10.0	13.3
Hematocrit %	35 – 49	21.2	18.4	17.8	25.6	27.7	30.4	
Platelets (×109/l)	150 – 450	289	304	283	260	278	273	258
LDH in U/L	81 – 234			1260				
Reticulocyte count%				7.05				
ALT in U/L	12 – 78	41	28					
AST in U/L	15 – 37	63	127					
ALP in U/L	46 – 116		244					
Serum bilirubin Total in micro mol/l	3.4 – 17.1		190.6	124.5				
Serum bilirubin Direct in micro mol/L	0 – 3.4		17.68	5.3				
GGT in U/L	15 – 85		17					
Blood urea (mmol/l)	1.8 – 6.3	5.4	7.5					
Serum creatinine (mg/dl)	26 – 88	70	43					
Serum sodium (mmol/l)	136 – 145	134	140					

Serum potassium (mmol/l)	3.5 – 5.1	3.9	4.2					
CRP	0 – 5	41	45.2					
Dengue NS1 antigen		Negative						
Covid 19 – RAT		Negative						
ESR		65						
UFR Albumin Pus cells Red cells		3+ 1-2 0-	2+ 12-15 20-25	1+ 10-15 Nil				
DAT		Negative						
Total protein	64 – 82	80	74					
Serum albumin	34 – 50	50	39					
Serum globulins	22 – 48		35					
Urine culture		No growth						
Serum inorganic phosphorous	0.8 – 1.6		1.6					
Serum calcium	2.1 – 2.6		2.5					
INR			1.6					
APTT			32					
Epstein barr antibodies IgM (ELISA) Epstein barr antibodies IgG	< 0.89			Negative Positive				
Mycoplasma antibodies IgM				< 1:40 Negative				
Blood group				A Rh +				
Hepatitis B Surface antigen	< 0.89			1.17 (border line)				
Hepatitis C antibodies	< 0.89			0.13				
Chest X-ray				Normal				

Discussion

Diagnosis

At presentation we have considered two possibilities in this case, infection induced antibody mediated intravascular hemolysis or hemolytic crisis in a G6PD deficient child. As he didn't come out initially with the history of ingestion of acalypha indica and presence of febrile illness, we have done extensive investigations to rule out infection induced antibody mediated intravascular hemolysis. The Direct antibody test was negative. Epstein barr virus IgM antibodies, Mycoplasma IgM antibodies and Anti HCV were done at medical research institute found to be negative. Hepatitis B surface antigen was compatible with borderline results. Once he come out with the story and with the observation of urine sample we have concluded the possible diagnosis of G6PD deficiency in this child.

Anyhow the exact diagnosis of G6PD deficiency either by demonstration of reduced G6PD activity in RBCs or by electrophoretic and molecular analysis methods yet to be confirmed in this child at the time of writing this case report.

Severe hemolysis

On day 2 his hemoglobin was 5.6g/dl. He had eaten a large portion of the vegetable dish that may be contributed to severe hemolysis. There is a

published study on predictors of severe hemolysis with G6PD deficiency following exposure to oxidants stress in pediatric population. They have reported those predictors are male gender, negative family history, presence of fever and vomiting, a younger age and high alkaline phosphatase levels (15). Our child had all those factors except the younger age.

Conclusion

Consumption of acalypha indica leaves as a vegetable can results in life threatening hemolysis in some G6PD deficient children. Screening and awareness programs regarding G6PD deficiency in high prevalent areas of our country can reduce the probability of such children getting severe hemolysis as in this case.

Taking a detailed history and observation of a urine sample are the key approaches for early diagnosis in a male child with similar presentation in future for an improved outcome.

Figure 1 – Acalypha indica

Figure 2 – Lemon tinge jaundice

Figure 3 – Cola color urine suggestive of hemoglobinuria

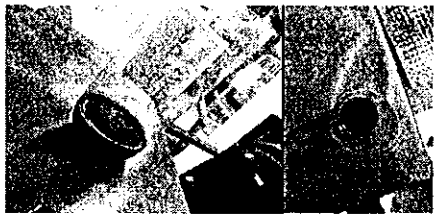

Figure 4 – Improved urine color after the treatment

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Case Reports

Calciphylaxis of the penis in end stage renal disease (ESRD) patient

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Abstract:

Introduction :- Calciphylaxis is a rare disorder of intravascular calcification that is primarily observed in patients with longstanding end-stage renal disease (ESRD). Patients develop necrotic skin ulcers [1].

Here we present the case of middle-aged male with ESRD and painful necrotic ulcers of his glans penis who was clinically diagnosed to be having penile calciphylaxis and later succumbed to the illness. This case depicts a distribution of calciphylaxis which is uncommon and carries high mortality.

Diagnosis of calciphylaxis is mainly clinical. Though biopsy of the lesion remains gold standard for diagnosis it is not done for typical cases due to the potential worsening of ulcer and sepsis [2].

Contemporarily there is no consensus on the optimal treatment for calciphylaxis and even with appropriate management, calciphylaxis is a lethal disease [1].

Keywords :- Calciphylaxis,Penis, ESRD

Case presentation

A 42-year-old man with a history of Diabetes mellitus, hypertension and end stage renal disease (ESRD)on hemodialysis, presented with necrotic lesion on the glans penis developing over the past 2 weeks duration. There were no ulcerative blackish lesions involving anyother part of the body. The patient did not give a history of trauma to the areas of ulceration or exposure to topical applications to glans penis. He had no history of smoking / illicit drug use. Of note, the lesion on his glans penis was recently treated with a 1-week course of oral amoxicillin–clavulanic acid without improvement.

The patient was afebrile and had a blood pressure of 150/80 mmHg. Physical examination revealed a blackish eschar like necrotic lesion on the glans penis with overlying purulence (Fig. 1)

Laboratory results obtained during admission are in Table 1.

Investigation	Results
Blood urea nitrogen	40 mg/dL (reference range 7–21 mg/dL)
Serum potassium	5.1 mmol/L (3.5–4.7 mmol/L)
Serum calcium	9.1 mg/dL (8.5–10.1 mg/dL)
Serum phosphorous	1.5 mg/dL (2.5–4.9 mg/dL)
hemoglobin A1c	6.9% (≤ 5.6%)
hemoglobin	10.3 g/dL (13.0–17.0 g/dL)

The patient underwent a computed tomography (CT) scan of the abdomen and pelvis and CT angiogram	extensive intravascular calcification in internal pudendal artery. No obvious thrombus seen.

Although the patient tolerated treatment without appreciable side effects, he experienced no improvement in the digital or penile lesions; in fact, he developed several more painful, necrotic ulcers. He was discharged home with plans to continue regular hemodialysis. On follow-up, his wounds remained painful with minimal progression. He succumbed to his illness in one month time.

Discussion

Calciphylaxis is a devastating rare disease that results in calcification of media of dermal arterioles and capillaries of skin and adipose tissue resulting in necrosis. The precise mechanism of pathogenesis is unknown, it is believed that calcium deposition in vessels result in endothelial damage and microthrombi formation, which reduces luminal caliber leading to ischemia and tissue necrosis [1].

Skin biopsy is the preferred method of confirming calciphylaxis; however, its role in practice is arguable as it can result in non-healing ulcers. So, biopsy is not needed for a patient with ESRD and the classic presentation of a painful necrotic ulcer covered with a black eschar [2].
some sources even argue that biopsy is contraindicated in penile calciphylaxis because of low diagnostic yield, as well as the particularly high risk of sepsis or acceleration of necrosis [3].

There are multiple risk factors for Calciphylaxis including dialysis dependent ESRD factors diabetes mellitus, high calcium and phosphate product, warfarin use and others. Our patient had long standing diabetes and was on dialysis. Due to its rich vascular network, calciphylaxis of the penis is even less common [3].
Further diagnostic testing is not necessary if the presentation is characteristic; however, if imaging is obtained, it may reveal intravascular or soft tissue calcification in corresponding areas. CT could be performed subsequently as the most sensitive modality to assess the extent of vascular and soft tissue calcification, necrotizing soft tissue infection [4].

The mortality due to calciphylaxis is approximately 64% predominantly as a result of sepsis. It carries a very poor prognosis with less than 1-year survival rate [5].

This case highlights the importance of recognizing penile calciphylaxis though rare, due to the fact it carries grave prognosis.

Informed consent
Informed consent was obtained from the patient.

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Case Reports

Eosinophilic Granulomatosis with Polyangitis: The Journey to Diagnosis A diagnostic challenge of a classic case of a rare disease

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Abstract:

Eosinophilic granulomatosis with polyangitis is a rare form of small to medium sized vasculitis that is characterized by asthma and eosinophilia that can later progress to multisystem involvement with skin, neurological, pulmonary cardiac and rarely renal involvement. The disease is a diagnostic challenge especially in the early course of the illness due to the phasic nature of presentation as it commonly mimics simple asthma and atrophy. Distinguishing between when to further evaluate or not may be ambiguous at presentation. We present a case of EGPA in a 57- year- old driver initially coming with episleritis and hearing impairment in the background of adult onset asthma, initially managed as per leptospirosis due to high inflammatory markers with conjunctival suffusion, with age- related hearing impairment. The diagnosis was not apparent until the next few months, during which time he gradually developed evidence of pansinusitis, peripheral length-dependent sensory motor neuropathy, mononeuritis, and progressive dyspnea with worsening of asthma symptoms.

Diagnosis was established following sural nerve biopsy evidence of vasculitis with positive P-ANCA and the patient fulfilled American College of Rheumatology diagnostic criteria for EGPA His HRCT was showing an isolated small nodule and evidence of right lower zone bronchiectasis. Most features over the time course were consistent with EGPA, but eye involvement and bronchiectasis were described less frequently in the context of EGPA. He was initially started on isolated low dose steroids due to minimal organ involvement, but later added azathioprine following which he attained marked symptomatic improvement.

Introduction :- Eosinophilic granulomatosis with polyangitis (EGPA) is a rare necrotizing vasculitis with extravascular granuloma formation. With a prevalence of 1.8/100 000 population, it is the rarest out of all ANCA associated vasculitis (AAV) which is the umbrella term for microscopic polyangitis (MPA) , granulomatosis with polyangitis (GPA) and EGPA(1) (2). Although categorized under the same entity AAV, EGPA is differentiated from other entities by persistent asthma, hypereosinophilia, and the low prevalence of ANCA positivity (3). EGPA was first described by churg and Stauss who established three major histological criteria for diagnosis consisting of infiltration of eosinophils, necrotizing vasculitis and extravascular granuloma(2),that are not demonstrable in many patients. American Colleague of rheumatologists (ACR) has established a more practical guidance for diagnosis consisting of six criteria out of which at least four need to be met to fulfil diagnosis(6).

The phasic nature of the disease is well established. They being prodromal phase, eosinophilic phase, and vasculitic phase (4). European Respiratory Society endorsed taskforce on EGPA described a novel entity hypereosinophilic asthma with systemic manifestations (HES), when the ACR criteria are not fulfilled but the patient has evidence of eosinophilic asthma. Recent studies confirmed a definite clinical and biologic profile overlap between the two groups of EGPA and HES, thus may influence future classification and updates in AAV(5)

Case report

We report a case of a 57-year-old driver from Hataraliyadda, a village in Central Province Sri Lanka, who presented with a two-week history of bilateral painless red eye and left-sided episodic hearing impairment, tinnitus, ear discharge with chronic cough and scanty clear sputum, which exacerbated at night.

His CRP and ESR were elevated, thus managed for leptospirosis with IV ceftriaxone and oral doxycycline and discharged with a follow-up plan at clinic level with repeat ESR.

Following discharge, he developed severe arthralgia, myalgia with a backache and severe bilateral lower limb painful paresthesia for which he was evaluated a month later.At the time his ESR had been persistently high for which he underwent myeloma screening, that was negative and a bone marrow biopsy which was a reactive marrow.

His third admission within 4 months was for persistent paresthesia, reappearance of red eye, hearing impairment, and evidence of sinusitis.
He gave a history of adult onset asthma with recent worsening, although not on regular inhalers. There were no skin rashes, chronic ulcers, history of night sweats or fever, polyuria, polydipsia, hematuria or frothy urine. His diet included protein based products.

On examination his sclera were red with conjunctival suffusion. Scattered rhonci and crepitations were present in bilateral lower zones of lungs, left more than right. Lower limbs were normal in tone and power with slightly diminished ankle jerk. Bilateral sensory loss was noted in stocking distribution up to mid-calf level. Joint position and vibration sensations were preserved. Rhomberg test was negative. Upper limb examination was unremarkable. Examination of the ear canal was normal. There was no sinus tenderness. However bi-lateral hearing impairment was noted.

His ESR was above 100. Although eosinophilia was not recorded initially, persistent eosinophilia was later noted. Blood picture had normocytic, normochromic and hypochromic, microcytic red cells with moderate rouleux formation, mild eosinophilia, a few hypersegmented neutrophils and moderate thrombocytosis. Mantoux and sputum for AFB were negative.
His basic renal function and liver profile were unremarkable. UFR was normal with no dysmorphic RBC. Complement levels were high with negative rheumatoid factor and ANA.
2ECHO was normal and ultrasound scan abdomen and KUB revealed a left-sided kidney of 6.3cm and right-sided kidney of 11cm.

Table 1: A summary of basic lab tests during each admission

	Normal	1st admission	2nd admission	3rd admission
WBC (×103/µL)	4-10	14.5	10.8	19.66
Neutrophils (×103/µL)	2-7	8.5(59%)		12.15 (61.9%)
Eosinophils (×103/µL)	0.02-0.5× 103/µL			1.69 (8.6%)
Lymphocytes (×103/µL)	0.8-4 × 103/µL			4.62(23.5%)
Monocytes (×103/µL)		2.32 (16%)		1.09 (5.6%)
Haemoglobin	11-16 g/dL	12.2	9.5	
MCV	80-100fL			77.7fL
Platelets ((×103/µL)	150-450	399	317	581
ESR (mm)	12mm	100mm	114mm	126mm
CRP (mg/L)	<10 mg/L	76		120
C3 (mg/dL)	90-180 mg/dL	221 (90-180)		
C4 (mg/dL)	10-40 mg/dL	45.5 (10-40)		

S.Creatinine (µmol/L)	65-120µmol/L	110		95.2
BU (mmol/L)	2.8-7.2 mmol/L	5.08		
Uric acid (mg/dL)				
AST (U/L)	<35U/L	16.2		49
ALT (U/L)	<45U/L	22.1		37
T.protein (g/dL)	6.6-8.3g/dL	7.9		6.6
Albumin (g/dL)	3.5-5.3g/dL	4.6		3.3
Globulin (g/dL)		3.3		3.3
Total Bilirubin (µmol/L)	5-19µmol/L	7.7		5.35
Direct Bilirubin (µmol/L)	1.7-6.8µmol/L	2.23		2.2
Indirect Bilirubin (µmol/L)		5.5		
ALP (U/L)	30-120U/L			246
GGT (U/L)	11-61U/L	38.9		120
Sodium (mmol/L)	136-146mmol/L	146		136
Potassium (µmol/L)	3.5-5.6mmol/L	4.7		4.6
Calcium (mmol/L)	2.1-2.55mmol/L	2.29	2.32	2.25
Phosphate (mmol/L)	0.81-1.45mmol/L	1.11		1.37
Magnesium (mmol/L)	0.7-1 mmol/L			0.74
CPK (U/L)	<171U/L	69.1	'99	10.1

ESR, erythrocyte sedimentation rate; CRP, C-reactive protein; AST, aspartate aminotransferase; ALT, alanine aminotransferase; LDH, lactate dehydrogenase; BU, Blood urea; CPK creatinine phosphokinase

The optic fundi were normal with left-sided episcleritis. Audiometry concluded bilateral sensorineural hearing impairment. Bilateral tympanic membranes and ear canals were normal.

NCCT brain showed sinusitis with sinus mucosal thickening. Rigid nasal endoscopy (RNE) showed right-sided grade 2 nasal polyps, left-sided hypertrophic nasal mucosa with normal post-nasal space and Eustakian tube opening. Mucosal biopsy wasn't done due to healthy looking mucosa.

Sinus CT showed soft tissue thickening in the bilateral frontal, ethmoidal and maxillary sinuses, consistent with pansinusitis. (figure1)

Tumor markers including PSA, CEA and AFP were normal.

Nerve conduction study indicated bilateral moderate sensory motor, large fiber length dependent polyneuropathy, sural nerve and mild left ulnar neuropathy more in favor of mononeuritis (figure 2)

Sural nerve biopsy showed no inflammatory changes within the neural tissue, but heavy inflammatory cell infiltrate predominantly consisting of lymphocytes with scattered neutrophils within the vessel walls of medium-sized vessels with fibrinoid necrosis consistent with medium sized vessel vasculitis in the perineural blood vessels (figure 3)

P-ANCA came positive with a negative C-ANCA but MPO and PR3 ANCAs were not available.

CXR (figure 4) showed nonspecific infiltrative shadows over the right lower zone. HRCT (figure

5) an 8mm sized lung nodule in the posterior basal segment of the right lower lobe, bronchiectasis involving the medial segment of the right middle lobe, with no evidence of interlobar or septal thickening. There were multiple prominent mediastinal lymph nodes (maximum diameter up to 8mm)

Finally, with the available clinical, biochemical and imaging evidence, a diagnosis of EGPA was made according to the ACR criteria. Patient was started on low dose steroid initially (0.5mg/kg) later adding concomitant azathioprine 50mg daily.

Figure 1: CT Sinus view with navigating protocol; note the soft tissue thickening of right frontal, ethmoidal, and maxillary sinuses.

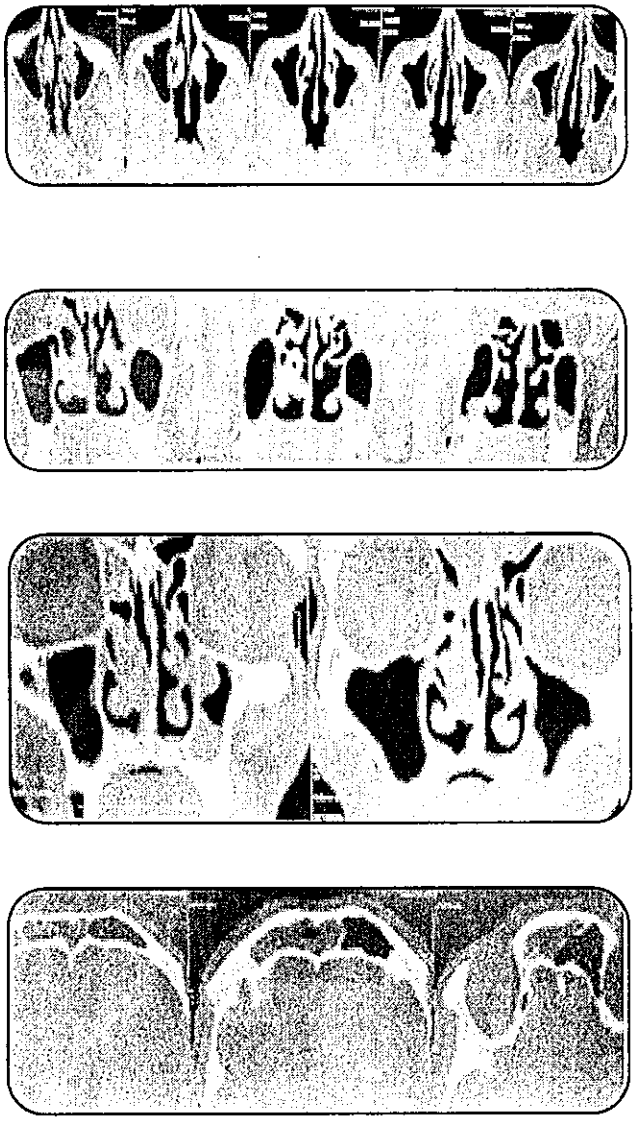


Figure 2: Nerve conduction study report

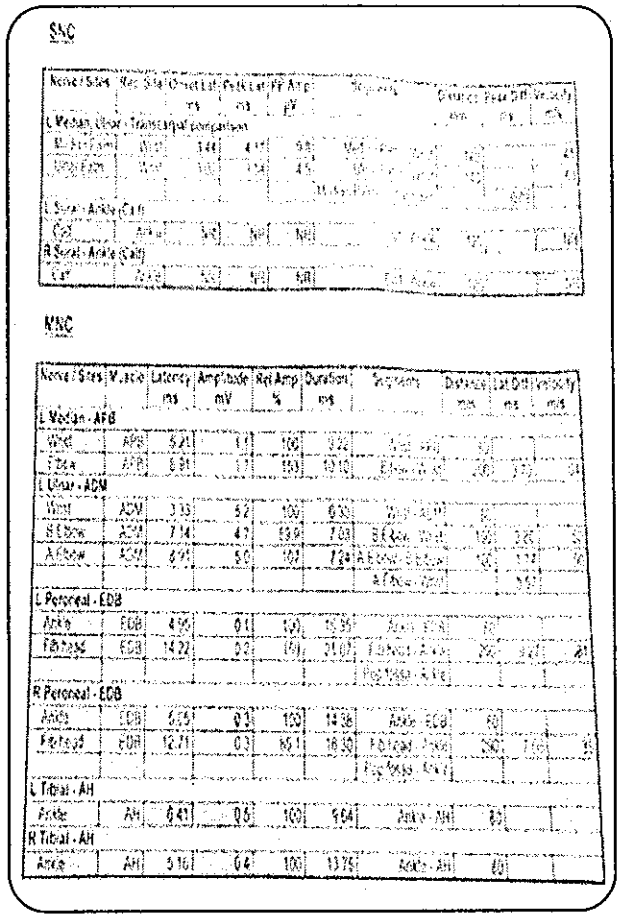


Figure 3 : Sural nerve biopsy specimens : (a) sections include neural and perineural soft tissue, (b) inflammatory cell infiltrates predominantly consisting of lymphocytes, note focal fibrinoid necrosis (d) The neural tissue shows minimal inflammation ; note the inflammatory cell infiltrating the medium sized vessel walls (H&E × 100)

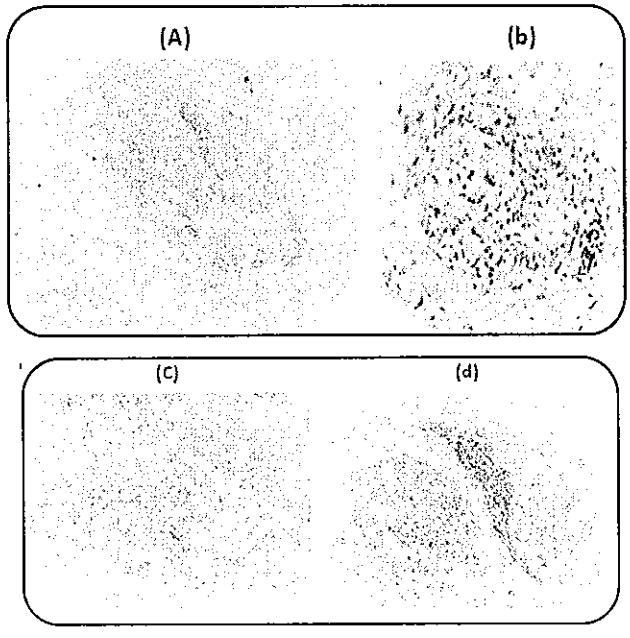


Figure 4 : CXR

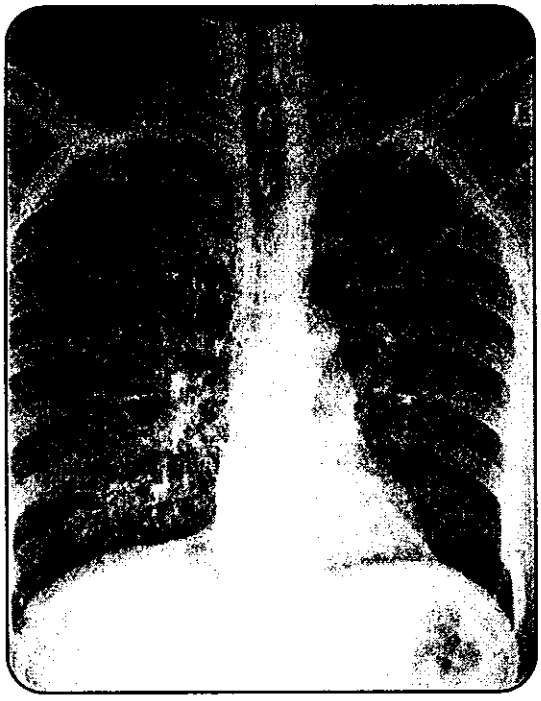
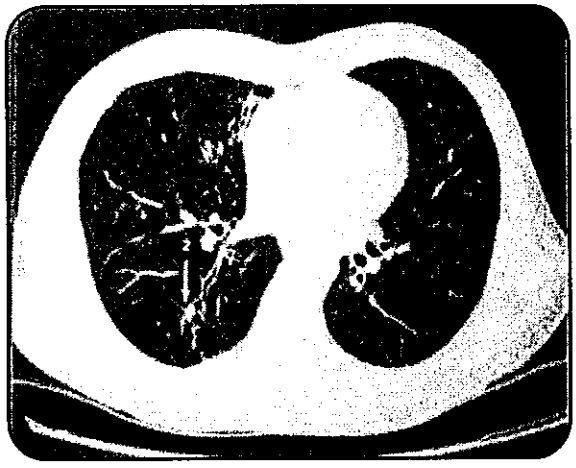
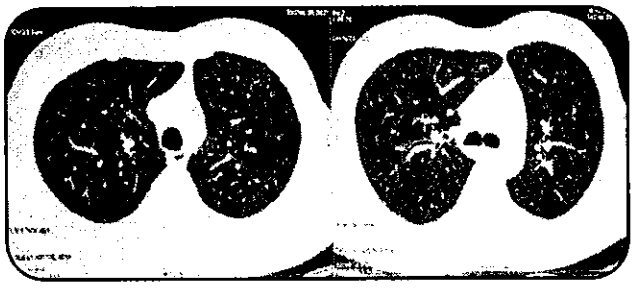


Figure 5: High resolution CT pointing to a small nodule, a few dilated bronchioles on the right lower zone can be appreciated.





Discussion

At the time of diagnosis our patient had most features of the disease fulfilling ACR criteria (6)(7). However isolated features of EGPA namely sinusitis and bronchial asthma having a benign course in most, even in the presence of eosinophilia are often due to atopy. However, we emphasize the need to follow these patients up, especially if inflammatory markers are high.

Eye signs are not common in EGPA. When present, literature describes ischemic signs more often than scleritis or episleritis. Ocular manifestations are of 2 categories , either idiopathic -orbital inflammatory syndrome-like presentation or ischemic vasculitis (8). Conditions such as periscleritis, marginal keratitis, and orbital apex syndrome were described, although quite rare. Commonest symptom reported had been redness in most studies. Ischemic vasculitis ranged from retinal artery and vein occlusion to ischemic optic neuropathy and retinal vasculitis, this group in contrast presented with transient or permanent visual loss. Ophthalmic manifestations are described approximately 5 to 6 years after the initial diagnosis of asthma, thus ophthalmologic examinations are recommended at least 5 years after the initial presentation.

In contrast otological and rhinological manifestations were commonly described in literature. They are multifactorial and have variable severity and often readily treatable with good response.

The few published reports on the otological manifestations of EGPA , had limited audiogram data with both sensorineural and mixed hearing loss, latter being more common (9) (10)(11)Although the causes were not always transpicious possibilities such as eustachian tube dysfunction, otitis media, interna, and auditory nerve dysfunction are postulated.

Bronchiectasis in the setting of EGPA is obscure (12) and he being a non-smoker didn't have other identifiable risk factors for development of bronchiectasis.

The airway manifestations in EGPA include centrilobular pulmonary nodules usually non cavitatory, V or Y shaped opacitis , dilatation and thickening of the bronchial wall, tree in- a- bud sign and mosaic pattern mostly noted in CT(13).Chest x-ray findings are diverse and may be transient, with no predilection to a specific zone. Transient

patchy opacifications, symmetrical bilateral axillary or peripheral opacities, diffuse or milliary opacities have been described, although nonspecific. A study conducted by Worthy et al. (14) describes bronchial dilatation, bronchial wall thickening, or both in 35% of the 17 patients studied, consistent with bronchial asthma which is a consistent manifestation of EGPA. Interestingly, our patient's clinical findings, CXR and High resolution CT scan (HRCT) were not comparable, possibly due to the transient nature of bronchial asthma and pulmonary infiltrates. The HRCT and CXR were not done simultaneously which also explains the discrepancy.

The symptoms of sinusitis, nasal polyps and peripheral neuropathy were vague in our patient but sinus CT confirmed pansinusitis. Overall, amongst the ANCA associated vasculitis, EGPA has higher incidence of peripheral neuropathy compared to other AAV. In a cohort study by Cattaneo et al.2007, neuropathy was described as an early manifestation of EGPA compared to other two with higher male predilection (15).

Hypereosinophilia can be easily missed in patients with EGPA, especially if not looking for it. Our patient's eosinophil count was around 1600, slightly lower than the usual range above 5000, however a count above 1500 is adequate for a diagnosis (6)
Treatment of EGPA has been revised in the latest treatment guidelines by ACR(16). While the guidance has endeavored to provide a generalized standard of care, they emphasize on adherence to guidelines to be voluntary and to individualize care. Although glucocorticoids with mepolizumab were given as the preferred option in the guidance, due to financial constraints, patient was initially on glucocorticoid monotherapy, later adding azathioprine as per guidance.

Conclusion

EGPA is a rare disease with manifestations that are common in the general population. The phasic nature of the disease makes it difficult to diagnose the disease during the early course where asthma and atopy may be predominant. Eye symptoms including episcleritis, although uncommon, can be a manifestation, and polyneuropathy or mononeuritis in an asthmatic may guide towards an early diagnosis. Imaging findings are variable and may not correlate with clinical examination due to the transient nature of lung manifestations. HRCT changes of bronchial asthma may be similar to bronchiectasis. Close follow up of patients with HES is necessary to differentiate from EGPA as patients may not fulfill all the diagnostic criteria of EGPA during the first few presentations to medical care.

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Case Reports

Giant lymphoepithelial retroperitoneal cyst

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Abstract:

Introduction :- Retroperitoneal cystic masses arising within the retroperitoneal space, but outside the major organ are uncommon. Primary retroperitoneal cyst formation is encountered with an incidence of 1/5750–1/250,000. As the retroperitoneum is a potential space, masses forming in it can grow to significantly large sizes before they present.

Usually, they are found incidentally. However, in some cases, they may attain a significant size without any other associated symptoms. Their symptoms are not specific and are mainly due to mass effect on adjacent structures. Typical cystic appearance in CT imaging with origin and involvement of adjacent structures and its characteristics may suggest specific diagnosis, but to achieve a definite diagnosis and to exclude malignancy histopathologic analysis is necessary. Complete surgical excision is the mainstay of treatment. This is a case of giant retroperitoneal cyst treated with a total surgical removal.

Case report

A 51-year-old female presented with generalized mild abdominal pain and abdominal distension for 1year duration. She also complained of feeling of stretching of abdomen particularly during squatting position and mild low back pain. There was no history of fever, loss of appetite and gross weight loss. Bowel habits were normal. There was increased frequency of micturition without any burning sensation. There was mild to moderate degree of dysmenorrhea, no dyspareunia. She is married and having two children born via normal vaginal delivery. Last childbirth was 17 years back. She doesn't have any medical illnesses and no family history of malignancy.

On examination, she is an average body built & nothing significant in general examination. Abdominal examination revealed fullness of almost whole of the abdomen with doughy consistency, no definite lump, no shifting dullness and no fluid thrill. Her per vaginal examination revealed fullness of right fornix and per rectal examination showed cystic fullness of pouch of Douglas with no definite lumpy feelings or ballotability.

Ultrasonography showed a large cystic lesion in the right lower abdomen arising from the adenexiae and extending up to her right hypochondrium with right sided mild hydronephrosis. It measured 18 * 12 cm in size. CT scan of abdomen reveled a unilocular thin-walled non-enhancing cystic mass in the right infra mesocolic space close to the ascending colon (Figure1). No solid components, no septations, no calcifications or other radiological features of malignancy were present. The mass was separated from the ascending colon and the retroperitoneal structures by small strands of adipose tissue.

Figure1: Sagittal and axial CT images shows a giant thin wall cystic lesion with significant mass effect on the intra and retroperitoneal structures.

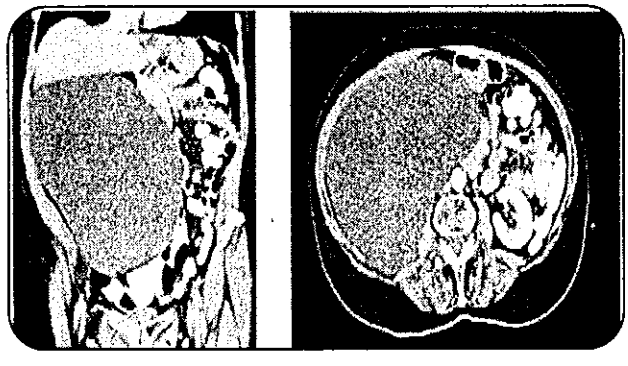

Figure2: Resection of retroperitoneal cyst which filled with milky or fatty substance

Blood tests showed white cell count (13 × 103/mm3) and ESR (68 mm/h). Her liver enzymes, serum amylase and serum lipase were in the normal range. CA 125 is 92 U/ml, but CA 19-9, alpha feto protein & CEA were in normal range. About 50 ml yellow-colored fluid was aspirated under ultrasound scan guidance and sent for histology. Cytological analysis revealed the presence of amorphous material with reactive mesothelial cells and scattered lymphocytes. There were no features suggestive of malignancy.

A midline laparotomy was carried out to remove the cyst. The cystic mass was in the retroperitoneal space and extended from the right iliac fossa upto the inferior surface of the liver and midline. The intraperitoneal structures were shifted to the left and upwards. Progressive blunt dissection enabled complete removal of the cyst from the adjacent tissues (Figure 2). No invasion into the surrounding structures were encountered. Uterus, both tubes and ovaries were normal in size. The specimen was sent for histopathology.

The cyst weighted around 3kg with maximal dimensions of 200mm 150mm 120mm. There were no septations noted within the cyst. The cyst wall had a smooth outer surface and appeared as yellow colored, thin, non-translucent with few areas of hard indurations. The cyst filled with the milky or fatty substance and it's devoid of any microbial growth.

Microscopically, the cyst wall composed of a thick fibrous tissue layer and aggregates of lymphocytes. The cyst wall is lined by the flattened cuboidal epithelium (Figure 3). There was no signs of atypia or malignancy were noted. The overall gross and microscopic appearance was suggestive of with a lymphoepithelial cyst of retroperitoneum. The patient was discharged on postoperative day five as the post operative period was uneventful. At her 6 weeks of clinic follow-up she was still asymptomatic and repeat CT shows no residual lesions or recurrence.

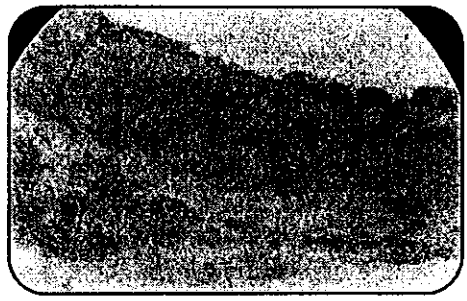

Figure 3: Lymphoepithelial cyst wall shows fibrous tissue and lymphocytes aggregates

Discussion

In this case report we discuss a patient with a giant cyst arising from the retroperitoneal space outside the major organs. Such "primary retroperitoneal cysts" are uncommon lesions. In most instances they are confined to the retroperitoneal space and typically not connected with adjacent structures. Most of them are small and encountered incidentally. Such large dimensions as in our case are rare in clinical practice.

Retroperitoneal cystic lesions can be classified as either neoplastic or nonneoplastic. Neoplastic lesions include, mucinous cystadenoma, cystic lymphangioma, cystic mesothelioma, cystic teratoma, Mullerian cyst, epidermoid cyst, pseudomyxoma retroperitonea and cystic change in solid neoplasms. Nonneoplastic lesions include pancreatic or nonpancreatic pseudocyst, lymphocele, hematoma and urinoma. Cysts that did not fall into the above mentioned categories are called benign idiopathic.

Lymphatic cysts include two subtypes. Chylous cysts which are arising from intestinal lymphatics or pancreas and lymphangiomas which forms out of retroperitoneal lymphatics and are not connected with the intestine. These lymphatic cysts are usually filled with a more fatty or milky substance. They can either be multilocular or unilocular and lined with flattened endothelium. Cystic lymphangiomas can grow into large volumes and have a tendency to cross from one retroperitoneal compartment to an adjacent one.

Its diagnosis remains difficult due to vague clinical presentation and lead to great challenge to the unsuspecting physician. Two-thirds of the cases present with chronic abdominal symptoms such as dull indistinct abdominal pain or symptoms secondary to the mass effect of the cyst (e.g. abdominal distension, palpable cystic or soft tissue mass, hydronephrosis). Non-specific symptoms such as referred pain, back pain, swelling of the

lower limbs and increasing weight have also been reported.

Radiological studies serve as an indispensable tool to describe these lesions. Approximately one-third of them are discovered incidentally on ultrasound of the abdomen or CT imaging. Contrast enhanced CT is the preferred imaging technique as this cross-sectional technique provides an excellent description of the lesion and a detailed analysis of the anatomic relationships with the adjacent structures. Density measurements gives radiologic evaluation of the content of the cyst. Malignant characteristics such as contrast enhancement of the cyst wall and solid nodules are absent and the majority of the cysts are unilocular. Magnetic resonance imaging studies can provide additional information about the precise location, possible connection between the cysts and intra-abdominal organs and content of the retroperitoneal cyst.

Complete excision is the preferred strategy of treatment especially when the cysts have grown to large volumes or become symptomatic. Both extra-peritoneal and trans-abdominal approaches can be used to excise the cyst depending on their location. A laparoscopic approach can also be undertaken in selected cases if enough expertise is available. Blunt dissection without damaging the cystic wall and preservation of the surrounding vital structures is utmost important. Cyst drainage and Marsupialization is an alternative if surgical enucleation is too difficult. But if the cyst wall is not completely removed, the recurrence rate is more with high chance of cyst infection.

Conclusion

This case highlights the facts that it is very uncommon to have a primary lymphoepithelial cyst in the retroperitoneal space. Contrast enhanced CT is the preferred imaging technique as this cross-sectional technique provides a detailed analysis of the anatomic relationships with the adjacent structures. Complete surgical excision is curative and required for definite diagnoses.

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Case Reports

Case report of a twin baby with hydrops fetalis possibly due to paroxysmal supraventricular tachycardia

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Abstract:

Introduction :- Hydrops fetalis is considered if there is an abnormal fluid collection in two or more fetal compartments (1). Earlier it was thought to be the cause of Rh isoimmunization, where recently other causes and associations of non-immune hydrops fetalis became responsible for at least 85% of all cases of fetal hydrops. It needs a detailed evaluation and earlytreatment for the better outcome.

Supraventricular tachycardia (SVT) is one of the most common conditions requiring emergency cardiac care in fetal, neonatal life (2) and may cause ventricular dysfunction and hydrops fetalis. Untreated cases of hydrops may easily cause death of fetus or hypoxic events.

Case report

Here in we present a case of a moderate preterm baby with the POA of 336 and the weight of 2.3 kg , who is the T1 of a monochorionic mono amniotic twin pregnancy of a non consanguineous parents. The twinning was identified by the week of ten with an uneventful antenatal period. Because of reduced fetal movement, mothergot admitted to the base hospital where the diagnosis is made as hydrops fetalis of the T1 and transferred to the tertiary care hospital CSHW.

Here, due to the persistent reduced fetal movements and the absent diastolic flow the mother underwent emergency section after two dexamethasone doses were given.

At birth the baby was edematous, had low APGAR which needed resuscitation and intubation. Initially the baby was kept on conventional ventilation and then to HFOV mode.

By six hours, circulatory insufficiency was noted and intravenous Dopamine, Dobutamine and Milrinone were initiated .There was bleeding through the ET Tube and deranged liver functions were noted. PCC and FFP were given according to the values during the stay.

On the day two baby developed tachycardia of >240bpm which were confirmed by ECG as SVT. The episode was terminated by the IV Adenosine. As there was a documented hypocalcemia, calcium gluconate bolus was given and continued as maintenance.

SVT episodes were noted another three times where all needed adenosine boluses. On expert opinion, Electrophysiologist suggested to start on propranolol until the baby is stable to carry out a proper electrophysiological study.

Due to the neutrophilic predominance, empirically intravenous antibiotics were given for ten days.

By day five the baby became stable on the respiratory aspect where we were able to wean off from HFOV. Day eight of the stay found to be icteric due to direct hyperbilirubinemia. Series of USS and investigations excluded biliary atresia and possible inspissated bile syndrome was managed with the antifailure regimen.

Further electrophysiological studies or the cardiac medications were not continued as the baby was clinically stable and thriving well. Due to the multiorgan involvement, inspissated bile syndrome and the renal involvement were investigated further. By day 20 the baby was discharged from MBU after establishing the feeding with a good weight gain.

Investigations

	3/9	3/9	4/9	6/9	9/9	11/9	15/9	19/9
WBC	30.7	26.4	13.4	9.7	8.1			
N%	57	66.5	81.7	48	52			
Hb	17.9	16.4	14.4	15	12.5			
Plt	157	120	87	90	168			
CRP	1.8			<6				
PT/INR	>12	3.65	5.2	3.157	1.6			
APTT	>180	67.4	60.2	65.9				
BU		4.4			4.1			
Scr		71.4			85.3			
Na^+		143.9			146.5			
K^+		3.3			2.9			
Ca^{2+}	2.3	3.05						
Mg^{2+}	1.7	2.5						
AST	6420	419						
ALT	550	353						
ALP	567							
Total Protein	37.9			46				
Albumin	25.6	36.9		34.2				
S. Bilirubin Direct/Indi-rect	45.8			177	248 89.2/158	216 103/112	196 123/72	206 107.1/98
B/Culture		No growth						

CXR - fluid collection in the pleura, peritoneum and the pericardium

USS - Small GB with smooth inner surface, no biliary dilatation
2DEcho - large PDA and a small ASD
Repeat 2Decho- small PFO

Discussion

Once the diagnosis is made , evaluation of hydrops begins with an antibody screening, detailed sonography of the fetus(es) and placenta, echocardiography and assessment for fetal arrhythmia, and middle cerebral artery Doppler evaluation for anemia, as well as fetal karyotype and/or chromosomal microarray analysis(1). The main factors are imbalance of interstitial fluid production and the lymphatic return due to congestive heart failure, obstructed lymphatic flow, or decreased plasma osmotic pressure.

In our case, the hydrops fetalis of the first twin has been identified on the day of the delivery by Ultra Sound Scan. Although our first consideration was twin to twin transfusion syndrome due to MCMA twin pregnancy, evaluation showed no features suggestive of that. As the mother and the babies had the same blood group the most common cause, isoimmunization was excluded. No documented fetal tachycardia or the abnormal fetal echocardiograms in the antenatal and postnatal periods. VDRL was non reactive in the mother.

As there were four episodes of SVT during the stay, we considered the complete diagnosis as Non immune hydrops fetalis of the twin one due to the paroxysmal supraventricular tachycardia complicated with respiratory distress, metabolic acidosis, liver dysfunction with severe coagulopathy and electrolyte imbalance.

Other possibilities may be hypoalbuminemia or multiple pregnancies causing the hydrops.

Sustained fetal supraventricular tachycardia with a heart rate of approximately 210 bpm may lead to increased atrial and venous pressures and congestive heart failure. In SVT, IV adenosine is preferred as cardioversion can be performed rapidly with limited adverse effects (2)The inspissated bile syndrome needs close monitoring and the supportive medications only.

The clinical significance of this presentation is, although the majority of subsequent pregnancies are not affected, the literature shows few recurrent

idiopathic cases have been reported which needs a specific attention. Regarding the further management of the baby, we have planned for a neurodevelopmental, growth, cardiology follow up at the clinic level.

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Case Reports

Hypercalcaemia: A sole manifestation of occult TB lymphadenitis.

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Abstract:

HTuberculosis (TB) is one of the infective diseases known to cause health burden worldwide. Affecting one third of the world population with significant morbidity and mortality. Sri Lanka is one of the endemic countries for TB in Southeast Asia. Even though pulmonary infection is more common, significant number of patients present with extrapulmonary TB and disseminated TB. TB has a wide array of clinical presentation which was reported time to time by several Clinicians. Here we report a case where hypercalcaemia was the only abnormal finding in a patient who was eventually diagnosed with TB lymphadenitis.

Introduction :- Tuberculosis is an airborne infection caused by Mycobacterium tuberculosis which is prevalent all over the world. Affects mostly the developing countries than developed countries because of the overcrowding, poor economic status and inadequate awareness. Affects both immunocompetent and immunosuppressed people with the latter group have high risk of infection and its complications. TB most frequently causes pulmonary infection though it is well known to cause multi system infection. Caseating granuloma formation is the hallmark of the infection, and it is known to cause hypercalcaemia as a complication of it.

Keywords :- Hypercalcaemia, TB lymphadenitis, granulomatous disease.

Case report

The 64-year-old male who is diagnosed with ischemic heart disease, bronchial asthma and hypertension presented with poor appetite with unintentional weight loss of 3Kg over the period of three months duration. He also reported exertional tiredness, constipation and nausea for the same duration. He did not have evening pyrexia, night sweat, respiratory symptoms or haemoptysis, bone pain, upper abdominal discomfort, palpable neck masses, lower urinary tract symptoms or haematuria, chest pain, dyspnea or dysphagia. He did not have past history or contact history of TB. He was not taking dicey diuretics for the control of his hypertension. Other than his routine medication, he did not take any vitamin supplements, calcium supplements or antacid medication. He was a teetotaler and lifelong nonsmoker. There was no history of high-risk sexual behavior. His past surgical history and family history were not significant.

On examination his BMI was 17.6 Kg/m2. He was alert and well oriented. He was neither pale nor icteric. There was no clubbing, cyanosis, lymphadenitis or ankle oedema. There was no erythematous skin rashes or neck lumps. Auscultation of the lung fields revealed, equal breath sounds in both sides and minimal wheeze. Abdominal examination did not reveal hepatosplenomegaly. Cardiovascular and nervous system examination were unremarkable.

On initial investigation he was found to have hypercalcaemia with albumin corrected calcium value of 3.32 mmol/L which was repeated and confirmed, with normal phosphate and magnesium levels. He also had renal impairment with serum creatinine of 157 umol/L. His complete blood count, fasting blood sugar, serum electrolytes, liver biochemistry, serum LDH level, UFR, serum uric acid were normal. ESR was 26 mm/first hour and CRP was 0.6 mg/L. Thyroid function test was normal. VDRL and retroviral screening were negative. CXR did not reveal any abnormalities. Ultrasound scan of the abdomen revealed multiple gallstones without

evidence of cholecystitis. He was treated with intravenous normal saline 3L/24 hours and oral furosemide 40mg daily with which his calcium level reduced to 2.74 mmol/L.

Further investigations were directed to find out the cause for hypercalcaemia. Neck ultrasound scan was negative for parathyroid gland abnormalities, but it revealed enlargement of the deep cervical lymph nodes. Intact PTH level was within normal range, 24-hour urinary calcium excretion was normal. CECT of the chest, abdomen and pelvis revealed level IV cervical lymph node and mediastinal lymph node enlargement. Upper GI endoscopy and lower GI endoscopy revealed normal studies. Screening for multiple myeloma was negative. Tuberculin skin test was negative and serum ACE level was normal. 1,25-OH vitamin D level was elevated. Since all the investigation were inconclusive, ultrasound guided true cut biopsy of right supraclavicular level IV lymph node was performed and sent for histopathology. FNAC sample was sent for Gene Xpert. Biopsy histology revealed granulomatous lymphadenitis with confluent granulomata with lymphoid cuffs which favoured the diagnosis of TB lymphadenitis which was further supported by positive Gene Xpert report. He was started on CAT 1 anti TB treatment (ATT) as for extra pulmonary TB lymphadenitis. His symptoms improved following treatment with combination ATT and the serum calcium level also normalized.

H & E stain of lymph node: Granulomata with lymphoid cuffs

Discussion

Mycobacterium tuberculosis is an obligate aerobic and facultative intracellular acid-fast bacillus with more predilection towards lung tissue. Because of its slow growing nature, human body's immune system act against it by delayed hypersensitivity type reaction with granuloma formation. At the center of granuloma, mononuclear phagocytes which are infected by the bacilli are seen. They are surrounded by highly differentiated, multi nucleated Langerhans giant cells, epithelioid cells and foam cells. The outermost part of the granuloma consists of a rim of lymphocytes. Caseating necrosis takes place at the center of the granuloma [2].

Manifestations of hypercalcaemia are nonspecific, such as personality changes, poor concentration, depression, anorexia, nausea, constipation, nephrolithiasis and pancreatitis [3]. Hypercalcaemia is symptomatic, usually with the serum calcium level more than 2.9 - 3 mmol/L. Complications of severe hypercalcaemia, at serum level more than or equal to 3.7 - 4.5 mmol/L are associated with electrocardiographic changes like bradycardia, AV block, short QT interval resulted in arrhythmias and CNS manifestations like stupor or coma. Therefore, it should be treated timely [1].

There are multiple causes for hypercalcaemia. Among that, primary hyperparathyroidism is the most common cause, occurs due to presence of parathyroid adenoma, hyperplasia or carcinoma. Malignancies associated with production of PTH related peptide (PTHrP) are the second most common cause for hypercalcaemia. Familial hypocalciuric hypercalcemia, vitamin D excess, side effects of the drugs, milk alkali syndrome and excess dietary intake are the other causes for hypercalcaemia [1,3]. Granulomatous inflammatory conditions like sarcoidosis, TB are known to cause high level of 1,25-OH vitamin D level. Macrophages in the granuloma, in higher rate, converts 25-OH vitamin D into 1,25-OH vitamin D [3,4] which enhances the absorption of calcium iron from gut and renal tubule resulted in hypercalcaemia. Where PTH level would be suppressed to normal or lower normal due to negative feedback mechanism by increased calcium level. There are several case reports have been published in the literature regarding hypercalcaemia associated with active pulmonary TB, miliary TB and hepatic TB, but less with TB lymphadenitis. Whereas hypercalcaemia was the only abnormality present in our patient, rest of the expected biochemical changes associated with TB were lacking, namely ESR which is usually elevated significantly in TB.

High level of calcium causes renal impairment by reduce glomerular filtration rate, increase sodium excretion with depletion of total body water [1]. Acute kidney injury was noted in our patient at the time of admission, which improved with the correction of hypercalcemia.

Conclusion

Since TB is endemic in Sri Lanka, which could be considered as a differential diagnosis of hypercalcaemia when appropriate. In the absence of typical clinical findings and typical investigation findings, infection with Mycobacterium tuberculosis is still possible. Therefore, TB specific investigation such as staining for AFB, Gene Xpert, culture for Mycobacterium tuberculosis, Interferone-Gamma Release Assays (IGRAs) and histopathological studies need to be performed from the body fluids and tissue samples which are collected from the patient. Since it is a completely curable condition with appropriate treatment, clinicians should have a lower threshold to suspect TB infection as differential diagnosis in appropriate clinical ground.

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Case Reports

Ichthyosis Netherton Syndrome

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Abstract:

Introduction :- Netherton syndrome is a rare autosomal recessive disorder which affects the skin, hair & the immune system. It is caused by the mutations in the serine protease inhibitor of Kazal type 5 gene (SPINK5) which is involved in skin and hair morphogenesis, as well as anti-inflammatory and antimicrobial protection of mucous epithelia. The incidence of the condition is 1 in 20000.

It is clinically characterized by a triad of congenital ichthyosiform erythroderma, trichorrhexis invaginata, and an atopic diathesis. Usually, the affected individuals present at birth with erythroderma. All of the skin is red and scaly. The hair shaft abnormality (Trichorrhexis invaginata) due to invagination of the distal portion of the hair shaft into the proximal portion is pathognomonic. It is often seen by trichogram or trichoscopy as "bamboo hair". Atopic manifestations include atopic dermatitis, asthma, food allergies, allergic rhinitis, urticaria, angioedema, anaphylactic reactions. Most individuals have elevated serum IgE levels & eosinophilia.

Keywords :- Netherton syndrome, erythroderma, atopy

Case Report

The case we report is a newborn of a consanguineous marriage. The baby was born via normal vaginal delivery at 36/52 period of amenorrhea with a weight of 2.6 kg at delivery. This baby had eczematous patches and secondary skin infection at birth. The first baby died of sepsis-related neonatal mortality at the age of 6 weeks. The third, fourth, and fifth babies are 8, 4, and 1 years old, respectively. Netherton syndrome was present in all of the newborns. In the first three months of life, all of the babies exhibit congenital icthyosiform erythroderma and have difficulty in gaining weight. Recurrent infections have never been a problem in the past. The company is currently booming.

Dermatological referral done. We started an antibacterial cream. We are planning to follow up in the immunology clinic for further investigations.

Fig 01 - Generalized Erythroderma & Scaling of skin

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Case Reports

Kikuchi disease with Evolving SLE

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Abstract:

Introduction :- Kikuchi disease is a self-limiting benign disease of unkown etiology that is characterized by cervical adenopathies and fever. Extranodal manifestations are rare. The skin may be affected, with nonspecific lesions. The diagnosis represents a challenge due to similar features to non-Hodgkin lymphoma, systemic lupus erythematosus (SLE), and viral infections. However, examination by an experienced pathologist will lead to correct diagnosis.

Here we present a case of kikuchi disease with evolving SLE.

Keywords :- Kikuchi, SLE,

Case report

This is a 24-year-old lady who presented with a history of fever for one month duration with associated rash involving face and upper torso for 2 weeks duration.

2 weeks after the onset of fever she has noticed that there were dark reddish brown macule involving ears, malar region and then it spread in couple of days to lower neck and shoulder region. No mucosal involvement.

She did not have other features of connective tissue disorder.She had no loss of appetite or weight loss.

Clinical examination revealed a febrile patient with enlarged and tender bilateral cervical adenopathy. She was neither pale nor icteric. Scattered erythematous macules were noted over the ears and upper trunk which were fading.

Abdominal examination revealed no palpable organomegaly. Respiratory, cardiovascular, Neurology and musculoskeletal examinations were unremarkable.

Laboratory studies are presented in Table 1.

FBC	WBC-9.8, N-79%, E-2%, L-15%, Hb-11.9, MCV-87,PLT-345
CRP	23
ESR	87
Procalcitonin	0.2
LFT	NAD
UFR	NAD
S.Ca	8.9
LDH	202
Blood picture	Viral infection
Blood cultures	No growth
ANA	Negative
Rheumatoid factor	Negative
C3,C4	Normal
USS neck and abdomen	Multiple cervical lymnodes largest on r/s level IV 2x2.3 cm No goitre, No organomegaly
Lymphnode biopsy	Necrotizing lymphadenitis
Skin biopsy	Favours cutaneous SLE
EBV/CMV/Toxo Serology, retroviral studies	Negative

Patient was started on HCQ, prednisolone and naproxen and the fever settled.

The rash gradually faded off with no scaring.she was discharged with a plan to follow up at the clinic.

Discussion

Kikuchi disease is a rare benign histiocytic necrotizing lymphadenitis, described in young women, usually characterized by cervical lymphadenopathy and fever. Histopathology of the involved lymph nodes differentiates Kikuchi disease from several more serious conditions.It may mimic including Infections like EBV,CMV,Toxoplasmosis and retroviral infections and other lymphoproliferative disorders.(1)

The young lady in this case presented with ongoing fever for more than 4 weeks and cervical lymphadenopathy and macular rash. Since all infection panels were negative and the lymphnode biopsy was suggestive of kikuchi disease the diagnosis was made without any doubt. The skin biopsy report was in favor of systemic lupus erythromatosis and she did not have any other features of SLE upon presentation and ANA was also negative.

Since SLE per say can present with PUO and lymphadenopathy with rash initially it was challenging whether to label this case as kikuchi disease. Considering the negative ANA, normal C3,C4 and absence of other features of SLE she doesn't full fill the EULAR/ACR 2019 criteria of SLE.

Many of the symptoms, cutaneous manifestations, biological and histopathological features of Kikuchi disease are similar to those of SLE, and differentiating between the two entities is challenging.(2)

Several reports have emphasized the importance of Kikuchi and SLE association. Kikuchi disease can precede or coexist with SLE (3).

Some authors recommend antinuclear antibody (ANA) screening at diagnosis and close follow-up, especially in patients with cutaneous lesions for the early detection of an autoimmune disease (4).

Since there are few cases where SLE was subsequently diagnosed few years after SLE she is being followed up at clinic.The presence of weight loss, arthralgia, skin lesions, and ANA was associated with the development of SLE(5).

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Case Reports

A case of laryngeal tuberculosis in an immunosuppressed patient- An uncommon presentation of a common

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Abstract:

Introduction:- Laryngeal tuberculosis(TB) is rarely seen in today's practice and could be primary or secondary depending on the pathogenesis. (1) primary laryngeal tuberculosis develops when tuberculous bacilli directly invade the larynx while secondary disease occurs via bronchogenic, hematogenous, or lymphatic spread of the bacilli from a secondary site. (2) Patients commonly present with dysphonia, dysphagia, odynophagia, stridor, and constitutional symptoms .(3)It is important to remember that isolated laryngeal tuberculosis can happen.

True vocal cords appear to be the commonest site affected by laryngeal tuberculosis(4). With the use of anti-tuberculosis treatment, the incidence of laryngeal tuberculosis has gone down dramatically (3)But it remains a highly contagious disease and a diagnostic and management challenge.

Case Report

A 44-year-old who was a diagnosed patient with dermatomyositis with Interstitial lung disease(ILD) for two months who was treated with multiple immunosuppressive medications including I.V methylprednisolone, followed by oral prednisolone and I.V cyclophosphamide and Hydroxychloroquine presented to us with a new-onset fever which was intermittent without chills, rigors, or night sweats. She also complained of one week duration of dysphonia without any hoarseness of voice. There was mild dysphagia and odynophagia. She didn't have any cough, sputum production, urinary tract symptoms, loose stools, or any other symptom suggestive of a focus of infection. She had lost 3kg during last month despite on prednisolone although her appetite was good.

Examination revealed a febrile averagely built woman with a BMI of 21.9 who was pale without icterus. There was no clubbing, lymphadenopathy, or ankle oedema. Respiratory system examination revealed a respiratory rate of 16 per minute, central trachea, and a few bibasal end-inspiratory fine crepitations which were compatible with the previous admission clinical findings. The central nervous examination was normal while peripheral nerve examination revealed a slight reduction in proximal muscle power with normal reflexes. Cardiovascular, abdominal, breast, per vaginal, and per rectal examinations were normal.

Her full blood count showed a WBC count of 5600/µl with the neutrophil count being 55% and lymphocyte count being 41% and normal hemoglobin level and platelets count. Erythrocyte sedimental rate was 67 and C reactive protein level of 38. Blood, sputum, and urine cultures and Mantoux test were negative. Sputum AFB was negative. Chest x-ray revealed minimal opacity involving bilateral upper zones but Repeat HRCT showed changes suggestive of early pulmonary tuberculosis involving both upper zones in addition to previous findings of ILD.

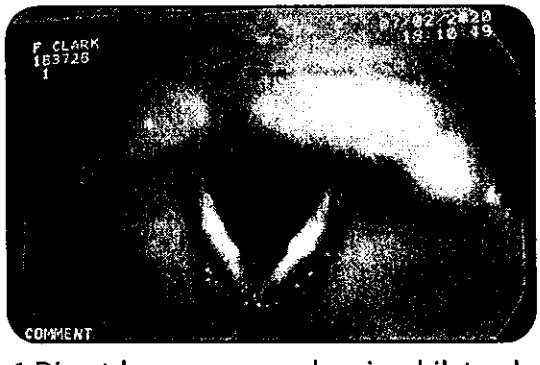

Fig1:Direct laryngescopy showing bilateral vocal cord nodules.

Fiberoptic laryngoscopy revealed bilateral vocal cord nodules that were suggestive of laryngeal tuberculosis. Biopsy taken from the right vocal cord lesion revealed granulomas with the caseating center and Langerhans giant cell and lymphocytic infiltration which was suggestive of tuberculosis.

She was started on the standard anti-tuberculosis regimen with the combination of isoniazid, rifampicin, pyrazinamide, and ethambutol. Oral dexamethasone 0.5mg b.d for 3 days was given.

Once stable immunosuppressive therapy was maintained with the lowest possible dose of prednisolone (20mg mane) and HCQ. Further Cyclophosphamide doses were withheld.

After two weeks of anti-tuberculosis therapy patient clinically improved with no dysphonia, odynophagia, or dysphagia or fever and was discharged with an antituberculosis drug regimen and her regular medications.She was clinically well for two months without recurrence of symptoms but admitted with a bacterial pneumonia 2 months after where she passed away following sepsis and multiorgan failure.

Discussion

Dysphagia and oral mucosal disorders are well documented in dermatomyositis.(5) However the clinical presentation of this patient suggested an ENT infection as a most likely diagnosis. Her recent hospital admissions and immunosuppressive status were the risk factors for her to get tuberculosis.

So specially when an immunosuppressive patient presenting with fever and dysphonia, dysphagia, odynophagia, or stridor, it is worth considering laryngeal tuberculosis as a differential diagnosis. Current guidelines warrant treatment of laryngeal tuberculosis with standard six-month combination therapy with two months of intensive phase but further data is needed about the necessity of a longer duration of treatment/intensive phase especially in immunocompromised patients as in this case.

Dermatomyositis is known to be associated with many malignancies with an increased risk is in the first 3-5 years after the diagnosis.(6) commonest were cervical, lung, ovarian, pancreatic, gastrointestinal, and bladder malignancies.(7) (8) Although laryngeal carcinoma is not common in the background of dermatomyositis a case of laryngeal rhabdomyosarcoma was noted in literature.(9)So the exclusion of secondary malignancy was a primary object in the diagnosis assessment in our patient. The biopsy confirmed granulomatous inflammation with caseous necrosis which was diagnostic of tuberculosis with the clinical picture.

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Case Reports

Leptospirosis with pulmonary hemorrhage successfully managed with therapeutic plasma exchange

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Abstract:

Introduction :- Leptospirosis is a zoonotic disease caused by Leptospira species which are spirochetes. Sri Lanka reports 3000-5000 suspected cases per year with a case fatality rate of 1-2%. (1) Treatment of pulmonary hemorrhage in leptospirosis is supportive with mechanical ventilation and the use of corticosteroids and therapeutic plasma exchange (TPE) is still being evaluated. We are presenting a successfully treated case of Leptospirosis and pulmonary hemorrhages with the use of TPE.

Case Report

A 67 year old previously healthy Sri Lankan farmer from Kegalle, presented with fever and severe myalgia for 3 days and reduced urine output for one day without any other systemic symptoms. He was a teetotaler. On examination he was alert, had icterus and conjunctival suffusion. His pulse rate was 96 bpm and blood pressure was 100/65 mmHg. He had tenderness in the right hypochondrial region and respiratory system examination was normal.

His full blood count showed white blood cells 6.16×109/L (4.5-11×109/L) , neutrophils 86.8% (40-70%), lymphocytes 7.4% (20-40%), platelet count 64×109/L (150-450×109/L), and hemoglobin 11g/dL (14-18 g/dL). C-reactive protein was 375mg/l (<5 mg/L) and creatinine was 5.4mg/dL (0.6-1.2mg/dL). Serum bilirubin was 26µmol/L (5.1-17µmol/L) with direct bilirubin of 19µmol/L (1.7-5.1µmol/L). AST was 56U/L (0-45U/L), ALT was 167U/L (0-55) and INR was 1.4 (<1.1) Arterial blood gas revealed pH 7.32 (7.35-7.45), lactate 3 mmol/L (0.6-2.0 mmol/L), HCO3 12mmol/L (21-28 mmol/L), pCO2 24 mmHg (35-45mmHg) and pO2 92 mmHg (80-100mmHg).

He was managed with intravenous Ceftriaxone and hemodialysis considering the possibility of leptospirosis. The next day he became tachypneic and on air oxygen saturation was 84%. Chest x-ray showed multiple patchy opacifications in both lungs. There was a hemoglobin drop from 11g/dL to 9g/dL. For the possibility of a pulmonary hemorrhage he was started on IV methylprednisolone 1g daily for 3 days and supported with non-invasive ventilation. TPE was started as the last option since there was no improvement after methylprednisolone pulses. On the first day of TPE 2641 mL plasma was extracted and 2132 mL fresh frozen plasma (FFP) was given together with six units of platelets. During the TPE next day, extraction of 2810 mL total plasma volume and transfusion of 2500 mL FFP was done. The 3rd cycle was done on the following day with extraction of 2700ml plasma and transfusion of 2700ml FFP. By this time the patient's oxygen demand gradually reduced and he became polyuric. Serum creatinine, blood urea and bilirubin started to become normal. Patchy opacifications seen on the previous chest x-ray were not seen in the repeat chest x-ray. Leptospira DNA was detected by real time quantitative PCR confirming the diagnosis. He was discharged on the 14th day from admission. In his follow up visit after 1 week he was found to have normal renal functions and he was in complete clinical recovery.

Discussion

This patient was a farmer with a high risk of exposure to leptospira and presented with clinical and biochemical features of leptospirosis with multi-organ failure. After three consecutive cycles of TPE

the patient had a remarkable improvement with the reversal of acute kidney injury and resolution of pulmonary hemorrhages.

Plasma exchange has been thought to be effective in severe leptospirosis in a few ways. In the leptospiremic phase, there is tissue damage due to severe inflammation. In the immune phase similar damage is caused by immune complexes. Once antibiotics are started, the release of endotoxins by the organism's death contributes to the above damage (Jarisch–Herxheimer reaction). (2) Plasma exchange can reduce the tissue damage by removing immune complexes and endotoxins. Removal of bilirubin reduces the nephrotoxic effect on renal tubules and helps in rapid recovery from the renal insult. (3) In a study done at the Teaching Hospital Karapitiya, Sri Lanka, 53 confirmed patients with leptospirosis had a 36.4% mortality rate after TPE, 21.4% after intravenous immunoglobulin and TPE, and 92.8% without TPE or intravenous immunoglobulin . (4) In a case series by Trivedi et al 114 patients with pulmonary hemorrhages secondary to leptospirosis underwent two TPE cycles and one dose of cyclophosphamide, and 30 were not given TPE. The survival rates were 64.4% and 16.6% in the treated and untreated groups respectively. (5) In conclusion, we observed a significant role of therapeutic plasma exchange in the survival of a patient with severe Leptospirosis with acute kidney injury and pulmonary hemorrhages. Further studies are recommended to evaluate the effectiveness of plasma exchange in severe leptospirosis.

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BATTICALOA MEDICAL JOURNAL
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Case Reports

A case report on possible immunization induced Multisystem Inflammatory Syndrome in a Neonate

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Abstract:

Introduction :- The global pandemic of coronavirus disease (COVID-19), caused by the novel coronavirus SARS-CoV-2 has affected all aged population in the recent past with higher incidence of clinical manifestations presently called multisystem inflammatory syndrome in neonates, children as well as in adults. Severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection during pregnancy may increase the risk of stillbirth, neonatal death, preterm birth, low birth weight, fetal distress, and neonatal asphyxia (1). As there are several new practices and new concerns emerging through, we need more cases and evaluation to recognize the spectrum of MIS-N, which is imperative for timely initiation of appropriate management.

Here we are going to present a case of possible MIS N in a neonate immediately after birth possibly due to the immunization of the mother.

Case Report

Term healthy baby with the birth weight of 3.625kg is the first born baby to a nonconsanguineous parents. Mother was vaccinated against the COVID 19 and the second dose has been taken two weeks prior to the delivery. Elective cesarean section was done at the base hospital with the indication being elderly primi. This was a planned pregnancy and the antenatal period was uneventful. The baby was not resuscitated at birth but in 10mins, the baby developed grunting and respiratory distress followed by drop in saturations.

The baby was stabilized with nasal prong oxygen at base hospital. At fourteen hours age the worsening of distress and significant low saturations associated with metabolic acidosis raise the suspicion of a cardiac disease.

Baby was resuscitated, ventilated and transferred to LRH for a echocardiogram and then to tertiary care hospital for further management. Initial echocardiogram revealed moderate PDA, PFO, Biventricular hypertrophy, bilateral large pleural and small pericardial effusion and Severe persistent pulmonary hypertension.

On admission as the baby was connected to HFOV immediately and was started on inhaled NO. Bilateral needle thoracocentesis was done soon after admission and 5cc fluid was drained bilaterally. Inotropes were initiated to maintain the blood picture. The baby was ventilated for nearly four days where we were able to wean off the HFOV and the NO on day two. There were multisystem involvement like seizures, coagulopathy, acute kidney injury in the initial two days of the birth.

Investigations showed high inflammatory markers- LDH, Ferritin, D dimmers, CRP where the cultures were negative. Pleural fluid full report showed some features of exudates where the culture was sterile. Due to the suspicion of MIS-N due to the multi system involvement and high inflammatory markers, the antibodies against COVID 19 was sent for both mother and the baby although the RAT and the PCR were negative. IgG anti bodies were positive for both. IV Penicillin, IV Meropenem, IV Teicoplanin given for 14days. One dose of IVIG was given.

There was a quick response after the IVIG dose to the baby. IV Dopamine and the antiepileptic medications were gradually weaned off. The baby

was discharged on day15 with the further follow-up care at the clinic level.

Discussion

Paediatric multisystem inflammatory syndrome in any category has been on the increasing trend of the pandemic which making the diagnosis and treatment challenging. As there are still researches going on in the pathogenesis, clinical spectrum and management, more and more studies will resolve the clinical dilemma.

In this scenario, the idea is to identify the possibility of getting Multisystem inflammatory syndrome in the neonates in the absence of the clinical disease or the exposure where the chances are due to the immunization and the acquired antibodies from the mother.

The possibility of perinatal vertical transmission of SARS-CoV-2 seems to be very rare. Although the most cases present as asymptomatic or mild infection with uncomplicated course, the literature, indicates that a few cases need intensive care (2,3). An intrauterine infection is suggested when a mother tests positive for SARS-CoV-2 within the period of 14 days before birth to 2 days after birth, with the detection of the virus in the neonatal respiratory tract in the first 24 hours of life, with either the persistence of swab positivity after 24 h of postnatal life or positive SARS-CoV-2 IgM in the first 7 days of life (4). The low placental expression of canonical receptors, with negligible co-transcription of angiotensin converting enzyme (ACE2) and transmembrane protease serine 2 (TMPRSS2) in the placenta necessary for the virus entry, may explain this low risk for vertical transmission (4).

With all these background details in our scenario we were able to come to possible diagnosis of MIS-N as we excluded other causes of the presentation. Although there are no evidence for a clear infection or exposure of the COVID- 19, the positive IgG levels in both the mother and the baby raise the suspicion of MIS N. But we need many more case reports and researches to prove the immune mediated diseases following the vaccines.

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Case Reports

Recurrent Kounis Syndrome due to Amoxicillin induced anaphylaxis

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Abstract:

Introduction :- Kounis Syndrome is the concurrence of acute coronary syndrome with conditions associated with mast cell activation, such as allergies or hypersensitivity and anaphylactic or anaphylactoid insults [1] Since its first description in 1991 it has been extensively studied and expanded. Three variants have been described with different pathophysiology: Due to coronary vasospasm (Type 1), plaque erosion and rupture (Type 2) and stent thrombosis with histological infiltration by eosinophils and mast cells of the thrombus (Type 3) [2].

Recognizing this syndrome as separate clinical entity is crucial in clinical practice as it has a unique management. Certain drugs used in Acute coronary syndrome such as Beta blockers may be harmful in Kounis Syndrome.

This case report aims to highlight the importance of early recognition of this syndrome amongst patients presenting with chest pain. To the best of our knowledge recurrent acute coronary syndrome due to Kounis syndrome due to the same agent is very limited in literature.

Case Report

A 60-year-old male presented with sudden onset rash, swelling of face and lips five minutes after taking amoxicillin for an infected wound. He also complained of an ischemic type of chest pain. He had a 30-pack year smoking history and regular alcohol consumption but no other vascular risk factors. He had a similar episode four years back where he developed chest pain and a rash few minutes after taking amoxicillin. He had documented ECG changes suggestive of an ischemic event during the previous episode. He has no history of other allergies.

On examination he had fascial and lip swelling with a generalized erythematous rash. He had bilateral rhonchi on auscultation. His blood pressure was 90/60 and had a tachycardia of 110 bpm.

ECG showed deepening T inversions from V1- V6 (Figure 1). Troponin I was 0.07 (more than the 99th percentile). 2D echo showed a mild hypokinesia of anterior wall with a EF of 50-55% and grade 1 MR.
A diagnosis of Kounis Syndrome was made with anaphylaxis to amoxicillin.

Patient was managed with IM Adrenaline 0.5ml (1:1000), IV hydrocortisone 200mg and iv chlorpheniramine 10mg. He received Stat doses of Aspirin 300mg, Clopidogrel 300 mg and Atorvastatin 40mg and was continued with Aspirin 75mg nocte, Clopidogrel 75mg nocte, Atorvastatin 40mg nocte, subcutaneous enoxaparin 60mg twice daily for 6 doses. He also received prednisolone 10mg and chlorpheniramine 4mg for five days.

Patient had an uneventful recovery. He did not give consent for coronary angiogram.

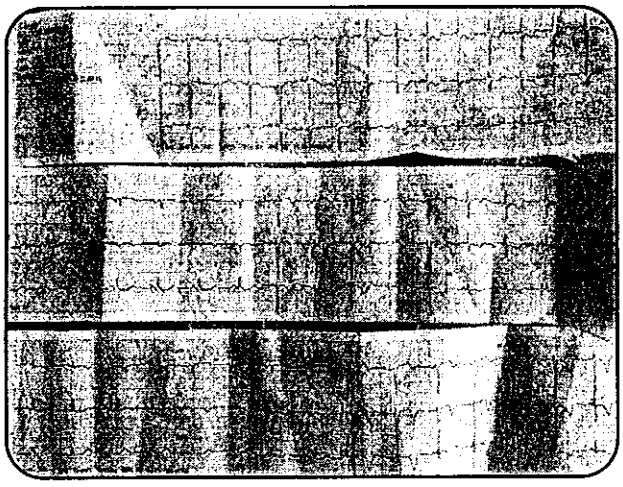

Figure 1: Serial ECG taken during admission

Discussion

Kounis Syndrome was first described in 1991 by Kounis and Zavras as a syndrome of allergic angina pectoris [3]. Postmortem examination of coronary artery specimens in 20 patient who died of myocardial infarction by Kovanen et al. in 1995 revealed higher levels of mast cell degranulation at sites of plaque rupture and erosion as compared to adjacent or distant sites [4]

Various triggers have been implicated with Kounis syndrome including drugs, food and environmental agents. Studies show that Nonsteroidal anti-inflammatory drugs is the most frequently trigger (60.7%), followed by drugs for cardiovascular disease (19.6%), antibiotics (17.6%), and anesthetics (9.8%) [5] Amongst drugs the commonest implicated is amoxicillin/clavulanic.

Although the diagnosis is clinical coronary angiogram could be performed which will show coronary artery vasospasm in the acute setting. Further allergic reactions could be proved by high serum tryptase levels and eosinophilia in blood. We did not perform Tryptase levels as an obvious clinical diagnosis of anaphylaxis could be made. Eosinophilia was not observed in our patient.

Management will depend on the Type of Kounis Syndrome. Type 1 will require only the management of the allergy/anaphylaxis while other types would additionally require the standard management of acute coronary syndrome. Since an angiogram was not performed in the acute setting, we assumed the patient to have Type 2 Kounis syndrome due to multiple risk factors in this patient for coronary artery disease.

Case reports of acute coronary syndrome following adrenaline injection were found on literature review. This was thought to be unlikely as our patient was symptomatic with ECG changes prior to administration of adrenaline and since only a therapeutic dose of adrenaline was used.

Conclusion

Kounis syndrome remains underdiagnosed in the emergency setting. It should be an important differential diagnosis amongst patients admitted with chest pain. This case report highlights the need to add the management of Kounis syndrome in the management guidelines of acute coronary events.

Conflict of Interest

None

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Case Reports

Recurrent Stroke and Sjogren Syndrome.

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Abstract:

Introduction :- Primary Sjögren's syndrome is an autoimmune disease of the exocrine glands with lymphocytic infiltration into lacrimal and salivary glands [1] with possible multisystemic sequalae [2]. Neurological complications in Primary Sjogren syndrome are often due to peripheral neuropathy through small-vessel vasculitis. There are case reports of Recurrent Ischaemic Stroke associated with Primary Sjogren syndrome, either as the initial presentation or as the sequalae [2]. We report a patient with Recurrent Ischaemic Stroke diagnosed to have Primary Sjogren Syndrome after many years of initial presentation.

Case presentation

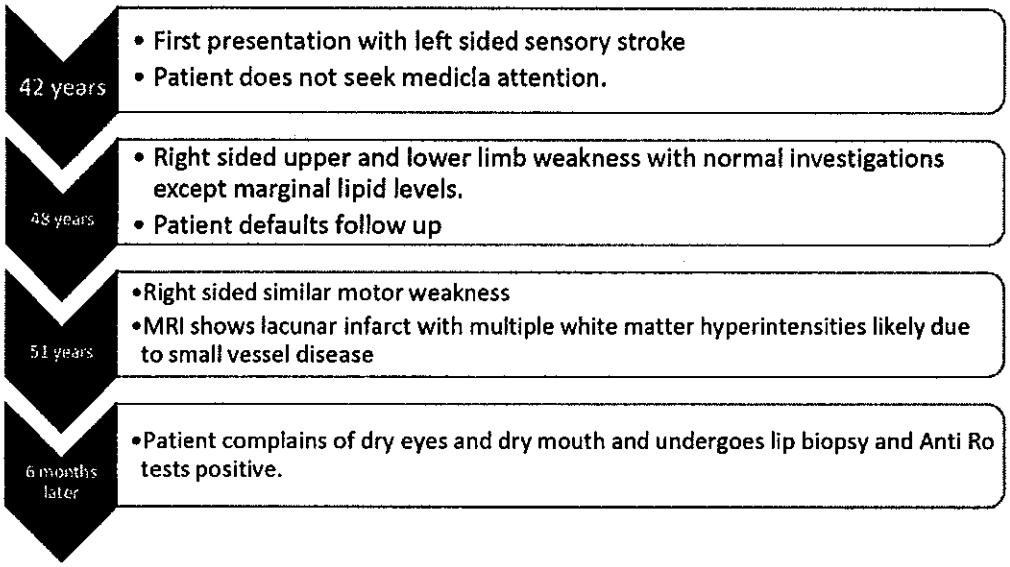
A 52 year old woman presented with the thirst episode of Acute Ischemic Stroke. She was not a known patient with Hypertension or Diabetes Mellitus, but her lipid levels were documented to be in the upper limit of normal 4 years back.

She recalled that at the age of 42 she had developed her first stroke with left sided upper and lower limb numbness which gradually improved over few weeks. She had not sought medical attention as it did not impair her daily functions. The second episode of stroke occurred six years after, at the age of 48, with right sided upper and lower limb weakness of MRC grade 3. She had sought medical attention at a hospital and underwent a Non contrast CT scan which did not show any haemorrhage or obvious infarct. She had undergone screening for hyperlipidemia which revealed a total cholesterol and LDL level towards the upper limit of normal. Rest of the evaluation including blood pressure monitoring, fasting blood sugar, carotid doppler, clotting profile including PT/INR and APTT were normal. She continued to take Clopidogrel and Atorvastatin for few months and defaulted treatment and clinic follow up.

This time she presented to us with right sided upper and lower limb weakness with a power of MRC grade 4. She underwent an MRI brain which revealed acute lacunar infarction with multiple white matter hyperintensities likely due to small vessel disease. She still had marginal total cholestrol and LDL level. She was restarted on dual antiplatelets and high dose statin.

After 6 months of follow up the patient complained of dry eyes, dry mouth and vaginal dryness for 6 months duration. She denied any other connective tissue disorder features such as alopecia, photosensitive rashes, miscarriages, arthritis, recurrent fever, weight loss, loss of appetite, dysphagia or Raynaud phenomenon. She did not have a family history of connective tissue disorders. She underwent a Schirmer test which was positive and her biopsy revealed lymphocytic sialadenitis of the minor salivary lobules consistent with Sjogren syndrome. As the ACR/EULAR 2016 criteria for the diagnosis of Sjogren syndrome was met with she was diagnosed with Primary Sjogren syndrome in the absence of other connective tissue disorder features. She was positive for Anti Ro antibodies and an ANA titre of 1:80 with fine granular pattern. Ds DNA was negative. Meanwhile the patient was referred to the Rheumatology clinic as such patients benefit from Hydroxychloroquine for relief of Sjogren syndrome symptoms and for consideration regarding need for other immunosuppressant therapy or steroids.

Figure 1



Discussion

Sjögren syndrome is a autoimmune disease characterized by keratoconjunctivitis sicca and xerostomia . Central nervous system involvement includes multiple sclerosis-like symptoms, including acute and chronic myelopathies, cognitive dysfunction, subacute aseptic meningitis, encephalopathy, psychiatric symptoms, chorea, and seizure[3]. Stroke is an uncommon manifestation of Sjögren syndrome, and recurrent strokes are an even rarer.

Ischaemic Strokes in Sjogren Syndrome patient could be due to accelerated artherosclerosis or vasculitis. A strong correlation has been found between the presence of anti-Ro antibodies and vasculitis in patients with Primary Sjögren syndrome [4]. In our patient, since the lipid levels were towards the upper limit of normal during the second episode of stroke, we suspect accelerated atherosclerosis with small vessel vasculitis as the cause of Recurrent Ischaemic Stroke.

A systemic review and meta analysis done in 2017 by Jirapat Teerakanok, et al. found no statistically significant increased risk for ischemic stroke in Primary Sjogren Syndrome patients compared to the normal population, but concluded that more studies are needed to clarify the association between these two conditions [1].

The treatment of primary Sjögren syndrome patients with central nervous system involvement is similar to that of patients with other systemic autoimmune diseases affecting the central nervous system and is based on experts' experience. The treatment mainly includes immunosuppressive and possibly intravenous immunoglobulin and rituximab [5].

Conclusion

Clinicians should actively look for features of Sjogren syndrome in patients, especially females with Recurrent Ischaemic Stroke.

Abbreviations
MRC- Medical research council
ACR/EULAR- American College of Rheumatology/ European League Against Rheumatism

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Case Reports

A young male with Aortic regurgitation diagnosed as Takayasu Arteritis

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Abstract:

Introduction :- Takayasu Arteritis is a well known yet rare form of large vessel vasculitis. It is also known as pulseless disease and affects predominantly the aorta and its main branches [1]. In this disease there is vessel wall inflammation leading to vessel wall thickening, fibrosis and thrombus formation [1]. Women under the age of 40 years are predominantly affected by Takayasu Arteritis with the highest prevalence reported in East Asia, especially Japan and Korea[2]. But Takayasu arteritis has now been recognised worldwide in both sexes, still with female predominance in East Asia. We present a common presenting complain evolving into a rare disease later.

Case report

A 22 year old male patient awaiting aortic valve replcement was referred to our Rheumatology unit by the Cardiothoracic unit for high ESR. On questioning the patient revealed having loss of appetite and loss of weight of 15% of bodyweight over a period of 6 months with left arm claudication pain radiating from shoulder to forearm. He did not have chronic cough, contact history of tuberculosis, inflammatory type of back pain, high risk sexual behaviour, rashes or genital ulcers. There was no past history suggestive of Rheumatic heart disease or family history of Connective tissue disorders or Ankylosing spondylitis. On examination he had an absent left brachial pulse with normal capillary refilling time in left nailbeds. Pressure discrepancy was present between both arms with the right arm pressure being 130/40mmHg and left arm pressure being 120/40mmHg. There was also a right carotid bruit. Although the patient did not give a history suggestive of carotidynia. Investigations revealed an ESR of 104 mm with a CRP of 40 mg/L. Mantoux test, HLA B27, ANA,C-ANCA, P-ANCA were negative. Arterial and venous duplex scan of left upper limb did not reveal any thrombosis.

The past medical history of the patient includes an initial presentation 9 months ago with subacute onset of shortness of breath worse with exertion leading to hospital admission and the diagnosis of Aortic regurgitation and Aortic root dilatation by 2D echocardiography. We did not find old records of ESR or CRP during the first presentation.

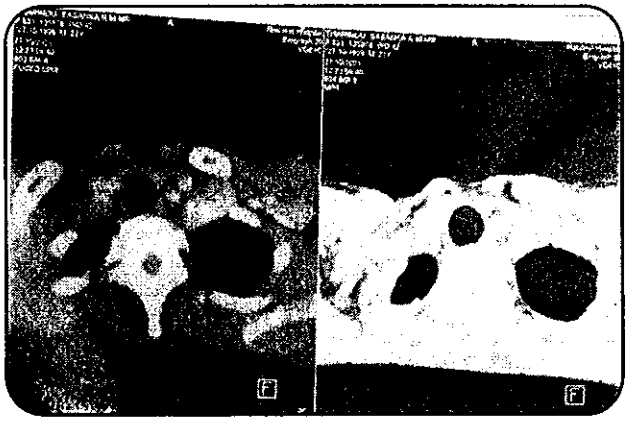
After the referral to our unit, due to the high clinical suspicion of inflammatory large vessel vasculitis in this young patient with aortic root dilatation, absent left brachial pulse with arm claudication, carotid bruit with high ESR and CRP, the patient underwent a CT Aortogram which only showed aortic root dilatation. He was planned to undergo PET CT scan and meanwhile was started on IV methyl prednisolone pulse 500mg for 3 days followed by oral prednisolone 1mg/kg and Mycophenolate mofetil 500mg Bd.

Patients ESR, CRP swiftly declined after the initiation of corticosteroids with an ESR decline from 104 to 30mm 1 month after initiation of corticosteroids and patient claimed to have improvement in left arm claudication. Meanwhile the PET-CT was reported as increased wall thickness with increased metabolic activity in the ascending, arch and proximal descending aorta which is likely due to inflammation (figure 1). The patient was diagnosed with Takayasu Arteritis based on American College of Rheumatology classification criteria due to the

patient having arm claudication with absent left brachial pulse, blood pressure difference of at least 10mmHg between both arms and age at disease onset less than 40 years. .

Patient's Prednisolone dose was titrated down and is currently maintained at 10mg per day. Patients ESR is currently normal at 18mm with absence of new arterial stenoses, for 8 months duration. He is awaiting review by the cardiac surgeon for aortic valve replacement since there is clinical and biochemical evidence of reduced inflammation.

Figure 1 : PET CT of the patient showing increased metabolic activity in the proximal descending aorta.



Discussion

Takayasu's arteritis is also known as pulseless disease, occlusive thromboaortopathy, and is a chronic granulomatous inflammatory arteritis affecting large vessels, predominantly the aorta and its main branches [1]. Vessel inflammation leads to wall thickening, fibrosis, stenosis, and thrombus formation. Aortic regurgitation and heart failure are well described and are usually preceded by constitutional symptoms, limb claudication, pulse and blood pressure discrepancies and vascular bruits [3].

Nipun Lakshitha, et al. have reported a similar case in a female patient presenting with isolated aortic root dilatation evolving into Takayasu arteritis, in Sri Lanka [3]. The above patient has also responded well to glucocorticoids with a good reduction in ESR. Timely diagnosis of Takayasu arteritis is crucial as prognosis of surgical interventions with ongoing vascular inflammation is unfavourable. We suggest periodic detailed clinical examination in young patients with aortic root dilatation to prevent missing such an important diagnosis.

Conclusion

Takayasu arteritis should be considered as a differential diagnosis in any young patient presenting with Aortic root dilatation.

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Case Reports

Constrictive Pericarditis: A Notoriously Elusive Diagnosis

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Abstract:

Constrictive pericarditis (CP) is a rare and frequently overlooked cause of chronic ascites leading to misdiagnosis and reduction in the quality of life of patients. We present a case of a 69 year old lady with a past history of tuberculous pericarditis presenting with ascites for 4 months duration with poor response to diuretics. The elevated JVP first stirred the possibility of a CP with the ascitic fluid analysis revealing a high protein content with an elevated serum-ascitic albumin gradient (SAAG) and the diagnosis was confirmed through a trans-thoracic echo (TTE) and a thoracic CT.

Introduction :- Constrictive pericarditis (CP) occurs due to pericardial fibrosis and thickening causing a non-compliant rigid pericardium leading ultimately to a diastolic heart failure7. In Sri Lanka it is mostly secondary to tuberculous pericarditis6. Patients with CP may present to non-cardiac specialties with either symptoms of fluid overload or symptoms related to a reduced cardiac output2, 9. Usually CP tends to get overlooked and the patient will be commenced on diuretics which will provide symptomatic improvement. However in the event of no other identifiable cause, CP must be actively evaluated for, otherwise it is easily missed with diagnostic delays having being reported for up to 10 years9, 14. The importance of early diagnosis is that it is completely reversible with a pericardiectomy[12].

Case Report

69 year old Sri Lankan female with hypertension, diabetes and hypothyroidism and a past history of treated tuberculous pericarditis 3 years back, presented with progressive abdominal distension over 4 months with a medical research council (MRC) grade 2 shortness of breath, orthopnoea, paroxysmal nocturnal dyspnoea and peripheral edema. There was no associated exertional angina or a productive cough and her urine output had been normal without any frothy urine or haematuria. Several similar episodes of admission were noted over the past 4 months where she had responded poorly to diuretics.

On examination her haemodynamics were stable. She was pale with bilateral pitting ankle edema. There was no clubbing or peripheral stigmata of cirrhosis. Cardiovascular system examination was normal except for the elevated jugular venous pressure (JVP) up to the angle of the jaw. The respiratory system examination revealed bi-basal fine end inspiratory crepitations and abdomen revealed a horse-shoe dullness and was otherwise normal.

Her primary laboratory evaluations were normal except for a mild hypochromic microcytic anaemia. The electrocardiogram, the cardiac bio-markers and the inflammatory markers were normal. The liver function tests revealed elevated transaminases (AST- 82 ALT- 54). The proteins, coagulation profile and thyroid profile were normal as was the hepatitis B, C serology, serum caeruloplasmin, anti-nuclear antibody (ANA) and tumour markers including CA-125. The urine analysis, serum creatinine and blood urea were normal. Ultrasonography did not show any evidence of cirrhosis but revealed an engorged inferior vena cava (IVC) with gross ascites and diffuse omental thickening. Upper gastrointestinal endoscopy detected no oesophageal varices.

Our main query was a tuberculous peritonitis, but the peritoneal fluid analysis revealed a high protein of 49.1 mg/dl with an elevated serum-ascitic albumin gradient (SAAG) which was not compatible with a tuberculous ascites. Staining for acid fast bacilli, TB PCR and tuberculosis culture were negative. There were no malignant cells in the cytology. Diagnostic laparoscopy revealed no omental deposits or peritoneal nodules and the omental biopsy

showed a reactive mesothelium without evidence of malignancy or granulomatous inflammation. Contrast enhanced computed tomography (CECT) showed a thickened pericardium of 10 mm without any pericardial effusion or calcification. The 2D Echo showed a preserved ejection fraction accompanied by bi-atrial dilatation and septal bounce with a dilated IVC without inspiratory collapse and a thickened pericardium. Therefore, a diagnosis of constrictive pericarditis secondary to tuberculousis was made.

Discussion

Constrictive pericarditis, resulting from a thickened, fibrotic pericardial sack, is a known, but often overlooked cause for ascites. There is a myriad of causes for CP including those occurring post cardiac surgery, post radiation therapy and autoimmune causes5. Post infectious causes for CP include tuberculous and purulent pericarditis7. Tuberculosis is regarded as the commonest cause of acute pericarditis and pericardial effusion in Sri Lanka even though statistical studies are lacking6. It is reported that the cases of tuberculous pericarditis is less than 4% in the developed world but in the developing countries with a high prevalence of TB, this is almost 70% 5, 15.

Despite the advanced imaging facilities available today, CP is still a difficult diagnosis to make. These difficulties have been reported for over a century with records of liver transplantations done for presumed cryptogenic cirrhosis due to the diagnosis of CP being missed1, 2, 10. The clue to diagnosing CP is the raised JVP and in any patient with an unexplained elevation of JVP, the possibility of CP should always be entertained 12, 16.

Analysis of the peritoneal fluid will be helpful in the differentiation of CP by the revelation of a high SAAG above 11g/l with high total protein level (>25g/l) of ascetic fluid9. In already established cirrhosis even though the SAAG is high, the ascitic fluid proteins will be low3, 13. The ascitic fluid protein will be high in cardiac causes of ascites which had not resulted in cardiac cirrhosis. CP might lead to the formation of frank chylous ascites and chylothorax as well8, 11.

The 2015 ESC guidelines recommend trans-thoracic echo (TTE) as the first line imaging modality for patients suspected of CP18 and the ventricular septal shift was found to have the highest sensitivity (93%) and of the utmost importance in diagnosis17. The other features which are reported to increase the diagnostic sensitivity are the variation in the mitral inflow velocity with respiration and the late diastolic flow reversal of the hepatic vein in the doppler study17. Further evaluation could be done by cardiac CT/MRI and the cardiac catheterization studies. Our patient underwent a CT, which is much more widely available and easier to access, and it demonstrated a grossly thickened pericardium. The ESC guidelines recommend cardiac catheterization if other noninvasive methods fail to confirm a diagnosis of CP20. Accordingly it was the specialist opinion that a catheterization study was not indicated in our patient.

Conclusion.

As demonstrated by this case report, CP is a notoriously elusive diagnosis since clinicians tend to overlook the clues pointing towards it. Diagnostic delays of up to 10 years have been reported, hence the need to include it as one of the differential diagnoses early when a patient presents with an otherwise unexplained ascites.

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Case Reports

Fatal Fulminant Hepatic Failure: Possible Relation to the Pfizer BioNTech Vaccination

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Abstract:

Introduction :- The pandemic of COVID 19 has hit the world more than 2 years now, which had a high demand of vaccine due to the rapid spread, high morbidity and mortality rate. There are several vaccines authorized by at least one national regulatory authority. Even though the above vaccines have excellent safety profile, there have been several mild side effects as well as some major fatal side effects. Here, we are reporting a case on Pfizer induced fatal fulminant hepatic failure.

Case Presentation

A 15-year-old, previously healthy girl who got her Pfizer BioNTech vaccine, developed intermittent vomiting after 8 days of her vaccine with loss of appetite with yellowish discoloration of sclera. She was admitted to a local hospital, and she has been conscious, rational and hemodynamically stable, found to have high transaminases and treated as viral hepatitis and discharged after a week of admission.

Since discharge she was having recurrent vomiting with epigastric pain and later, developed gum bleeding with hematuria and presented with altered level of consciousness few days after discharge from local hospital.

On Examination, she was deeply icteric, GCS was 11(E-3, V-3, M-5) and there was no neck stiffness. Blood pressure was 110/70mmHg and having a tachycardia of 110 bpm with hepatomegaly and lungs were clear.

WBC	19420	17420
Haemoglobin	10	8
Platelets	322,000	156,000
AST	1562	685
ALT	1282	895
T.Bilirubin	256	120
D.Bilirubin	442	351
Albumin	36	32
ALP	217	
GGT	42	
Plasma Ammonia	15.2	
S.Creatinine	65.6	
ESR	08	12
CRP	<5	<5
PT/INR	5.4	3.56
Ferritin	600	

Table 1 : Baseline investigations

Hepatitis A, B, C serology	Negative
EBV, CMV, HIV serology	Negative
Dengue and Leptospirosis serology	Negative
Anti-Nuclear Antibody	Negative
Anti-Mitochondrial antibody	Negative
Anti-smooth muscle antibody	Negative
Anti-Liver Kidney Microsomal Antibody	Negative
Ceruloplasmin Level	27 (15-60)
Paracetamol Level	Negative

Table 2 : Infectious and immunology work up

Ultrasound scan showed hepatomegaly and a hypoechoic liver with regular margin and no intra or extra hepatic dilatation or focal liver lesion was seen. The same day, GCS deteriorated and transferred to Intensive Care Unit. She developed right sided focal seizures and she was electively ventilated. NCCT brain showed cerebral oedema without herniation and EEG showed severe hepatic encephalopathy. Immediate supportive care started since admission, and IV N-Acetyl Cysteine infusion started with plasmapheresis. Liver biopsy was planned but not done due to the ascites and coagulopathy. Despite aggressive measures and immense effort, the patient's condition deteriorated and unfortunately patient died.

Figure 1 : Perivenular necrosis with viable ductular proliferation

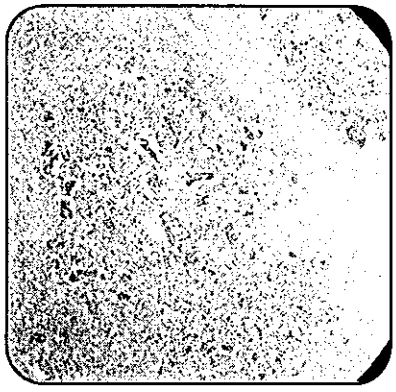

Figure 2 : Massive necrosis of liver

Discussion

Acute Liver failure due to drug induced liver injury (DILI), has been always a challenge as there are no diagnostic tools available and this is a diagnosis of exclusion. There are two types of chemical toxicity: intrinsic and idiosyncratic toxicity. Intrinsic toxicity is dose dependent with predictable regularity where idiosyncratic toxicity is not dose dependent and may occur any time after exposure with variable morphologic lesions. (1).

Roussel Uclaf Causality Assessment Method (RUCAM) is used to assess the relationship to the drug and liver injury and is used along with nR criteria which assesses the liver injury pattern. Novel biomarkers such as Glutamate dehydrogenase, high mobility group box 1, keratin-18 have been found even though their evidence for reliable utility is still accruing (2).

In this case, the review of history and medications did not reveal any other reason for acute liver injury and prior to this presentation our patient had no chronic illnesses. Acute hepatic failure can occur with vaccines even though it is more common with prescribed and nonprescribed drugs. So, clinicians should be vigilant in patients showing signs and symptoms of hepatotoxicity usually after 5 days but less than 3 months following the vaccine.

Conclusion

In summary, we presented a case of fulminant hepatic failure following Pfizer vaccine due to the idiosyncratic DILI, given no other cause in our patient after extensive work-up. The purpose of this case report is to raise awareness of potential and fatal side effects of COVID-19 vaccines.

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Case Reports

Vertebral artery trauma – an area that can be overlooked

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Abstract:

Introduction :- Blunt vertebral artery injury (VAI) frequently occurs in forensic practice. However, injuries to the vertebral artery are easily ignored or overlooked because of its relatively deep location[1]. VAI commonly occurs in penetrating injuries to the neck, such as firearm injuries, stab injuries, etc. Nonpenetrating injuries also play a role in VAI, such as blunt force trauma to the neck, hyperextension, and lateral rotation[2]. Isolated basal subarachnoid hemorrhage (SAH) is essentially associated with the VAI. SAH is usually massive and extensive. Level 3 (extracranial part) of the vertebral artery is the common area involved, but the intracranial part (Level 4) also can be involved and cause basal SAH[3]. Alcohol intoxication is significantly associated with VAI. Victims who sustain a blow to the head and neck are frequently under the influence of alcohol. After the development of the SAH, they used to lose their consciousness rapidly and could die immediately or following several hours or days[3], [4].

Case report

A 48-year-old manual laborer had consumed alcohol with his son-in-law at his home. After some time, they both ended up in a heated argument under the influence of alcohol. As a result, the son-in-law assaulted the father-in-law by the back of a thick-bladed knife found in the kitchen to the deceased back of the neck. Deceased developed convulsion after 30minutes and later collapsed. Relatives of the deceased rushed him to The Teaching Hospital Jaffna, and he has pronounced dead at OPD. An inquest was requested by the medical officer in charge of the OPD. The Magistrate has visited the hospital and ordered an autopsy. Only a linear abrasion was found on his back right side of the neck during the external examination. The musculoskeletal dissection revealed a contusion over the right upper posterior neck. There was a basal subarachnoid hemorrhage found while opening the cranial cavity. Careful exploration of the vertebral artery has shown an intracranial vertebral artery (Level 4) laceration on the right side.

Figure 1: A linear abrasion showed on the right side of the posterior upper neck.

Figure 2: Contusion on the right side of the posterior upper neck showed following Musculoskeletal dissection

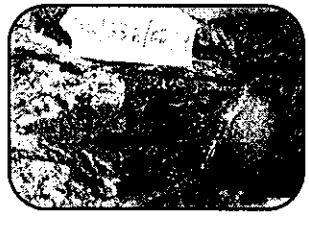

Figure 3: Basal subarachnoid hemorrhage while the brain was insitu.

Figure 4: The base of the brain showed subarachnoid hemorrhage around the brain stem and cerebellum

Figure 5: Digital subtraction angiogram of the vertebral artery showing the injury.

Discussion

The vertebral artery (VA) originates from the subclavian or innominate artery and enters the transverse foramen of the sixth or fifth cervical vertebrae (Level 1). The artery passes through a series of foraminae until it reaches the base of the axis (Level 2). Then, it turns posteriorly to enter the foramen transversarium in the posterolateral part of the ring of the atlas and perforates the posterior atlantoaxial membrane to enter the foramen magnum (Level 3). It then joins the contralateral VA to form the basilar artery (Level 4), which supplies the brain stem and cerebellum[5]. Assault behind the ear leads to trauma to the VA of that level (Level 3) as it is not protected by bone. This type of injury can extend to the intracranial portion of the vessel (Level4). Sometimes, injury can occur only in the intracranial part due to stretching of the vessel. And resulted in damage to thinner blood vessels and caused basal subarachnoid hemorrhage, which is fatal[6]. The case reported above also developed basal SAH, the following tear on the right intracranial part of the VA due to overstretching of the VA as a result of blunt trauma on the right upper posterior neck.

Carol K. Lee et al. stated that 50% of the traumatic vertebral artery injury (TVAI) was associated with alcohol toxicity[7]. Koszyca et al. studied 151 cases from English literature between 1971 to 2002 of TVAI. They stated that 89.7% happened following the assault, and 81.5% of them were under the influence of alcohol at the time of the incident[8]. Intoxication with alcohol may adversely affect neuromuscular coordination and could lead to overstretching of VA. It may also cause relaxation of the surrounding muscles and enhance the effect of mechanical rotation and extension. Vasodilatation and increased blood pressure due to alcohol intoxication may exacerbate the arterial rupture[8].

Torn vertebral arteries are often challenging to document at autopsy because of their relatively concealed and inaccessible location in the transverse foramen and foramen magnum and because the VA is usually cut to remove the brain, and the cut may pass through the region of the arterial trauma[9]. Careful removal of cervical spines as a block and keeping it in 10% formic acid for ten days to lysed the bone will give you a clear view of the VAI. Extensive histological examination is also helpful to locate the injury and demonstrate the associated pathologies such as degenerative changes and atherosclerosis. A relatively easy way to evaluate the integrity of the vertebral artery from its origin to its termination into the basilar artery is to perform a post mortem angiography. Any extravasation of contrast indicates a defect in the vessel that can be further explored with the dissection of that particular area of the artery[9]. In our case, we couldn't perform post mortem angiograms as our institution had limited facilities.

Conclusion

Vertebral artery injuries resulting in fatal subarachnoid bleeding are rare but a well-known phenomenon encountered in forensic settings. Careful gross, histologic examination and postmortem angiography are recommended in all cases of basal subarachnoid hemorrhage where trauma to the neck has occurred or was suspected before the death. Vertebral artery angiography probably holds the most significant promise for precise diagnosis and research.

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