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PRESIDENT'S MESSAGE



BMJ, Broadening the Horizons

Research, innovation and quality improvement are instrumental to bring developments to Health system and patient care. BMA having identified and prioritized the importance of research among medical fraternity stood up to publish its journal, BMJ biannually from 2019.

Embedding a culture of constantly seeking innovation & quality improvement in medical service is admirable along with safety of the patient. Research on one hand innovate, introduce and develop new methods, techniques and technologies to uplift medical field. More importantly on the other hand it will unearth some overlooked subtle complications, adverse events and inefficiencies which in long run are detrimental for patient safety and quality of care.

Research will improve comprehension, analytical skills, critical thinking and judgement. Collaborative, multidisciplinary, multicenter studied effectively enhance team work undermining destructive narcissistic ego of many medical professionals. Research as the language of medical professionals which speaks out ones' caliber and image, should be encouraged among junior doctors and incorporated in to medical curriculums.

Like any successful programme which continue to evolve, the diversity of publications – ranging from molecular studies to clinical trials; locally, nationally and internationally would be the ultimatum for BMJ. Broadened horizons and untiring efforts of editorial board of BMA paved the way for this second publication of BMJ in 2019.

I wish very best for this unprecedented piece of work of the Editorial Board to be a continuum and welcome you to take part of it as authors & readers.

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Are we heading towards another dark age?

We are all familiar with the philosophical, scientific and mathematical achievements of the ancient Greeks. Hippocrates, Socrates, and Archimedes are household names. The intellectual achievements of these individuals happened during a historical period known as the classical age. The accomplishments of this era were achieved through sound reasoning, critical thinking and a driving curiosity to understand the universe, the foundations of life and the reasons for our existence. However this classical era came to an end around 476AD. This was followed by a period known as the dark ages. It was a time when Western Europe faced profound demographic, cultural, intellectual and economic deterioration. Although controversies and exceptions exist, it is generally accepted that during this period scientific innovation and rational thinking became second place to religious dogma, superstitions and unfounded popular beliefs. Science was looked upon with suspicion and in many aspects considered to be magic and sorcery. Many who chose to think differently and challenge accepted norms through scientific thinking, were considered threats to society and needed to be eradicated by imprisonment, torture or death.

Science was all but dead in Western Europe until the renaissance once more resurrected scientific methods, the benefits of which we take for granted today.

Now the question arises is it possible for humanity to regress back to a similar state of intellectual deprivation. Many would choose to disagree, citing the enormous technological feats of modern man. Information is everywhere, new information is created daily at mind boggling rates, never before in the history of mankind have we had such easy access to such a vast repository of ever growing information. But does growth of information equate to growth of knowledge? T. S Eliot famously stated "Where is the knowledge we have lost in information?"

Information becomes knowledge only through a process of internalization. This process of internalization can only be achieved through the hard work of focused thinking, critical analysis and rational arguments. Unfortunately society

today does not seem willing to spend the time and effort to undertake the tedious process of reasoning; rather we feel that any knowledge can be obtained from the internet. The difference between information available at ones finger tips and internalized knowledge is blurred, with many people even intellectuals struggling to understand the difference. By failing to understand this difference we are drowning ourselves in information and unwittingly losing the concept of true knowledge.

This problem is especially evident in the younger generation. They are conditioned towards obtaining information easily. For past generations assimilation of knowledge was a slow and tedious process, consisting of much laborious reading, many times unrewarding. This process resulted in a lot of gaps in information, which had to be filled by rational thinking and critical judgement. This methodology is alien for our internet generation. The need for rational reasoning in order to discover knowledge, and the joy which comes with that "Ureka moment" has been lost. Now it's just a matter of finding the right search words in Google, and all the information verified and more often unverified are poured into our brains. We have lost our patience to listen and understand a slow, calm exposition of scientific reasoning as evidence of careful, trustworthy thinking. Rather people of today are more likely to give credence and accept someone with a manic, energetic, and confident style of presentation.

The internet has all but done away with credible expert advice. Everyone on the internet can be an expert. And many people tend to accept and follow these so called experts based on their internet status of likes and shares. People feel that if a site or blog has more "likes and shares" then it must be credible because more people are following. Unfortunately this is not scientific rationale, rather it is group mentality or to put it in a crude way "mob mentality". Just because a majority believes in a concept or idea that does not mean that it is the correct view.

This is where the problem arises. As more and more people begin to place their trust in information created outside

the scientific method, false information, pseudo-science, and unfounded fears will continue to rise; until we will reach a tipping point where the scientific method and scientific reasoning will take second stage to public opinion and dogmatic viewpoints. If such an eventuality occurs then we will undoubtedly begin to see a deterioration in knowledge, health, culture and economy. In all respects we will be looking into another dark age.

Although it may be hard to imagine such a ghastly future the ill omens of future perils are already part of our daily lives.

In our practice of medicine we see that many doctors lack a clear understanding of the scientific principles on which effective therapies are built upon. Rather they rush to the safety of guidelines. Preferring to blindly follow a set of guidelines without fully appreciating the context on which the guidelines were created. Also we have the so called "experts" and "concerned citizens" who hound and harass healthcare professionals on social media, their views are based on false information and misconstrued scientific knowledge, ultimately misleading the common man.

This problem is further compounded by grave mistakes of certain professionals and professional bodies, created by not upholding scientific principals above personal agendas. Twisting and falsifying research data to suit preconceived ideas, or narrow minded agendas has put further strain on the trust people have with the scientific establishment. Alternate therapies and pseudoscientific justifications are on the rise, with more and more people subscribing to these views. This is not to argue that alternate therapies do not work, rather if they did work then there must be a scientific reason for them to do so.

As the threat of pseudo-science grows even professionals fail to understand the difference. The vaccine controversy is an excellent example. Despite the utter dearth of scientific research pointing towards significant medical issues with vaccination, and the enormous body of scientific

knowledge showing the benefits of vaccination yet we see an ever increasing number of people including professionals choosing to embrace an anti-vaccination stance. Their viewpoints bearing an uncanny resemblance to the superstitious dogmas rampant during the European middle ages.

Recently we experienced another sad reminder that science may soon be pushed aside when a scientifically sound method of waste disposal was questioned even by health care workers, based on pseudo-scientific assumptions and misinformation.

As people are ever increasing misguided by misinformation, and as people are ever increasingly content to allow themselves to be misguided, and as long as people choose not to invoke rational thinking into the process of decision making, and rather rely on others to make decisions for them, then we as a people cannot expect a bright future. Intellectual illiteracy will result in a vicious cycle of misinformation begetting misinformation until the norms of human society as we know it today collapses into disarray.

So what can we do to prevent this catastrophe? This is where the theme of our scientific session takes on special importance, "Attitude Transformation and society development though Professionalism".

We carry with us as professionals the responsibility to safe guard and promote scientific methodologies. To fulfil this responsibility we must set aside our differences in personal opinions and dogmatic viewpoints and rather embrace scientific medicine in all aspects of our practice. We must be aware of the difference between true science and pseudoscience, while understanding that the boundaries of science are ever changing. We must use the methods of scientific thinking to enhance rational thinking, analytical abilities and problem solving skills within our society. It is only through this form of attitude transformation that we can hope to achieve true social development and lead ourselves towards a progressive future.

Dr. G.R. Francis.



Perception of Nursing and Midwifery staff on continuous foetal heart monitoring during labour at Teaching Hospital, Batticaloa, Sri Lanka: A Mind mapping analysis

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Abstract :

In Sri Lanka mothers have their baby delivered at a standard health care facility. Sri Lankan state health sector has developed well and caters majority of the delivery. Monitoring the Foetal Heart beat in mothers admitted to the labour ward for confinement plays a significant role in the neonatal outcome. Such monitoring has taken turns from simple and intermittent auscultation to continuous electronic monitoring.

The continuous foetal heart monitoring (CFHM) has been introduced to the labour ward Teaching Hospital, Batticaloa has been introduced in the year 2015. This study was conducted among the 25 nursing and midwifery staff working in the labour ward via an interview regarding their perception on the aspects of this method. Their views were categorised by developing a mind map and analysed under four themes.

The study reveals the necessity for training on this mode of CFHM to the staff and also to optimize the workload to the staff and the machine by developing guidelines on its usage to be selective. Childbirth however, is a natural process and a personalized care by the labour ward staff need not be overlooked.

Keywords:

Foetal heart monitoring, delivery, mind mapping, staffs' perception

Introduction:

Sri Lankan Health system has taken a significant amount of involvement in ensuring the pregnant mothers having their confinement in a standard health care facility. Such health care facility could mainly be in the state sector or in the private sector as well. Recent figures show that almost every woman has the delivery occurs in such facility.

The Sri Lankan state health system has shown a very significant development; thus, the hospitals and maternity homes are equipped either with basic or with emergency Obstetric care facilities and manpower. The Annual Health Bulletin 2014 published by Ministry of Health which contained statistics from 2008 that out of the total reported deliveries 99.6 - 99.7 % of deliveries

occurred in a health care facility (Annual health Bulletin 2014)[1]. Similar trend has been continuing as reported in the Sri Lanka Demographic and Health Survey (2016) [2] that in Sri Lanka Nearly hundred percent of births take place in a health facility: ninety-four percent were delivered in public-sector health facilities, five percent in private health facilities and only 0.5% at home or some other place. In estate sector nearly one percent (0.7 percent) of deliveries was outside the health facilities.

Sri Lanka continues to maintain an effective form of health care provision by maintaining a very low level of maternal morbidity and mortality. Similarly efforts are taken to minimize the perinatal morbidity and mortality by a vigilant form of monitoring of the mothers admitted to the labour ward for delivery by instituting effective foetal heart monitoring.

Foetal heartbeat (FHB) Monitoring during labour has taken turns from the simple form of auscultation by Pinnard stethoscope (Sudha Salhan 2007) [3] to electronic monitoring

such as Doppler ultrasound (Callagen, Rowland & Goldman 1964; Trudinger 1996) [4,5] and cardio toco graphy (Baker & Kenny 2006) [6]. It has also been developed from intermittent monitoring, to continuous monitoring. Cardio toco graphy (CTG) is the electronic form of monitoring which is widely practiced (Alfirevic, Devane, & Gyte 2006)[7].

Since the year 2015 continuous FHB monitoring has been employed at the labour ward, Teaching Hospital, Batticaloa. This practice is currently ongoing. This method of foetal heartbeat monitoring has been introduced to the nursing and midwifery staff who have been working in the labour ward at Teaching Hospital, Batticaloa were much familiar with intermittent monitoring by using Pinnard stethoscope or Doppler. Thus their perceptions about this newly introduced method becomes imperative to develop an understanding towards its continuous usage. This study therefore aimed at assessing the perceptions of the staff of the newly introduced method of FHB monitoring. Staff working in the labour ward of THB since 2016 were recruited.

Materials and Methods

The study was conducted at labour ward, Teaching Hospital, Batticaloa in the year 2017. Nurses and Family Health Officers (FHO) who involved in the care of laboring mothers and conducting the delivery involved in this study. Twenty-five such officers were interviewed regarding their perception on the continuous CTG monitoring and were requested to make their views in writing. The interview was conducted by the Nursing sister-in-charge of the Labour ward who received prior instructions from the investigator without disclosing the identity of the participants and was completely blinded to the investigator. The transcripts of the interview were then analyzed qualitatively and the perception notes were categorized in a mind mapping format. Ethical clearance to conduct this study was obtained from the Ethical Review Committee of the Faculty of Health-Care Sciences, Eastern University, Sri Lanka. Informed consent was obtained from each participant.

Results

A total number of 25 staff working in the labour ward took part in the study. Perceptions of the staff had been grouped into 18 statements. Based on the nature of the statement they are categorized into four broader themes such as

- Advantages of continuous FHB monitoring,
- Issues with CTG machine,
- Issues with the system and
- Recommendation by the staff.

They were developed mind map (Fig 1). Statements in each theme were then analyzed.

A. Advantages of continuous FHB monitoring

Almost every staff felt that this method has advantages specially with the regular situation of inadequate number of staff assigned

to the labour ward duty. They also felt that this method of monitoring has an advantage of minimizing perinatal deaths and improving the neonatal outcome.

B. Issues with CTG machine

Staff made their observation that due to continuous working of the machines they often are not working properly. There had been problems with the probes. Further as noted by a few participants that "we should have to take responsibility of the machine for not working properly". Some said "we only know to switch on and switch off the machine". Other statements made by a few are 'noisy' and also it is an electronic device and nothing like Pinnard which makes direct FHB monitoring.

C. Issues with the system

Over 60% of the staff noted that they have lack of knowledge in identifying the abnormality in the FHB and thus find difficulty in foetal monitoring. Further they note that 'when the machine is broken or not working staff to become responsible'. Thus, they also feel frustrating continuously working in the labour ward and at times avoid attending to duty.

D. Recommendation by the staff

One of the suggestions made by the participants is to minimize the use of continuous CTG and be selective. Thus, by making certain guidelines agreed upon by the different Obstetric teams working in the labour ward the use of CFHM would be effectively be undertaken and the burden to the machines as well as the staff would be under control. Although not suggested the quantity, the staff pointed out that 'An adequate number of CTG machines must be made available in the labour ward'. Participants of this study made a recommendation that they require 'training on CTG patterns and the operation of the machine'.

Discussion

Looking into these perceptions and into the relevant literature the perceptions raised by the participants of this study are similar as felt by the staff working in the labour ward of other hospitals (Devane et al. 2012; Altaf et al. 2006)[8,9]. Especially poor staffing levels, busy clinical environments and increased medicalization or industrialization of childbirth have been cited in the studies (Smith et al., 2012).[10]. In our population also, many prefer to have their childbirth in a major hospital with Consultant care.

Monitoring in the labour ward is principally aimed with the safety of the laboring mother as well as the foetus. Thus, the responsibility of the staff working the labour ward become significant.

Effectiveness of the foetal heartbeat monitoring depends on the training obtained by the health care staff working in the labour ward and the availability of suitable equipment in adequate quantity. Cardiotocography has been introduced and in operation



Influenza in Pregnancy

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Abstract :

The influenza virus is RNA virus and is classified into four subtypes, influenza A, influenza B, influenza C, and influenza D. One of the subtypes of influenza A, the H1N1 strain, also known as swine flu, is especially of high risk for development of complications in pregnant women. The influenza A virus infection is difficult to diagnose clinically because its presenting symptoms are similar to those of the common cold but are more severe, last longer, and can be potentially life-threatening. Its substantial morbidity and mortality impact can be mitigated by education of women and their physicians as well as by vaccination and use of available preventative and therapeutic modalities. The aim of this article updates the knowledge about influenza in pregnancy.

Key words: Influenza and influenza in pregnancy

Introduction

Influenza virus is a single-stranded, enveloped RNA virus from Orthomyxoviridae family. Its incubation period is 2-5 days and the transmission is mainly air-borne, i.e., droplet inhalation or direct transmission or contact through hands and fomites. It is classified into four distinct generations such as influenza A, B, C, and D depending on antibody responses to glycoproteins, hemagglutinin (HA), and neuraminidase (NA) on the surface of the viruses(1).

One of the subtypes of influenza A, influenza A (H1N1) strain called swine flu was first identified in April 2009 and the outbreak has since reached pandemic status at that time. So far, there have been 4 pandemic attacks worldwide: Spain in 1918 (unknown strain but suggestive of avian-like H1N1), Asian flu in 1957 by H2N2, in Hong Kong in 1968, and Mexico in 2009. After each pandemic attack, healthcare personnel have become more aware of its lethal complications. (2) (3).

Clinical features

Influenza is a contagious respiratory illness caused by flu viruses. It can cause mild to severe illness, and at times can lead to death. The flu is different from a cold. The flu usually comes on suddenly. People who have the flu often feel some or

all of these symptoms. The typical uncomplicated influenza A syndrome is a symptom complex that overlaps with a number of other respiratory illnesses. There is an abrupt onset, with high-grade fever (38–40°C) being the most prominent symptom. Respiratory symptoms such as rhinorrhoea, nasal congestion and sore throat are present, but are overshadowed by the systemic symptoms during the first 3 days of illness. The influenza virus tends to remain in the respiratory tract and does not usually cause a viraemia. The cough frequently changes from a dry, hacking nature to one that is productive of small amounts of sputum that are usually mucoid but can be purulent. After the fever and upper respiratory tract symptoms resolve (usually within 7–10 days), cough and weakness can persist for 1–2 additional weeks(4)(5).

Features of influenza include headache, myalgia, malaise, anorexia, sore throat, nonproductive cough, sneezing, and nasal discharge; these symptoms are not pathognomic for influenza, and asymptomatic infection can occur. The pulmonary complications of influenza include pneumonia (viral and bacterial), croup, asthma, and bronchitis. Complications of swine flu in the general population appear similar to seasonal influenza. Myocarditis and pericarditis have been observed, usually associated with a marked tachycardia. The prognosis is unclear, though influenza-related myocarditis usually has a good prognosis for recovery (6).

In addition to Reye's syndrome, a range of neurologic complications have been noted, including confusion, convulsions, psychosis, neuritis, Guillain-Barré syndrome, coma, transverse myelitis, and encephalomyelitis. Influenza has also been associated with the toxic shock syndrome, myositis, myoglobinuria, and renal failure. People with asthma may experience asthma attacks while they have the flu, and people with chronic congestive heart failure may experience worsening of this condition that is triggered by the flu (7)(8).

Pathophysiologic mechanisms

The pathophysiologic mechanisms underlying increased influenza risk to pregnant women and their fetuses are unclear. Significant anatomic and physiologic changes during normal pregnancy include changes that increase the risk of respiratory failure and complicate the treatment of respiratory illness. These changes include elevation of the diaphragm to accommodate the uterus, increased respiratory rate, increased intra-abdominal pressure, decreased chest compliance, and as a consequence, increased risk of aspiration. Decreased functional residual capacity due to a greater expiratory volume can lead to alveolar collapse. Because increased tidal volume is necessary to meet increased oxygenation needs, minute ventilation is increased, leading to falling arterial CO₂ partial pressure and compensated metabolic acidosis. These cardiopulmonary changes and the increased respiratory rate needed to compensate for the metabolic acidosis make pregnant women more susceptible to respiratory compromise, predispose to the development of pulmonary edema, and make such complications more difficult to treat(9)(10).

Effects on pregnancy

In Sri Lanka, influenza has a substantial contribution for maternal deaths in recent past years (about 10% of total maternal deaths in 2015 and 2016). On the other hand it is associated with increased risk of adverse pregnancy outcomes such as spontaneous abortion, preterm birth and fetal distress. Increased severity of influenza in pregnant women is thought to be related to normal physiologic changes that occur during pregnancy. The disease may be more severe in pregnant women with comorbidities such as diabetes, heart disease and bronchial asthma.

Prevention

Prevention of influenza infection in pregnant women and their newborns begins with efforts to limit exposures, including hand washing, respiratory hygiene and cough etiquette, and implementation of infection control precautions and environmental procedures in the healthcare settings that these individuals frequent. If hospitalized, droplet precautions should be instituted, and all persons coming within three feet of the woman should wear a surgical mask. Education of family members as well as pregnant women is a very important component of prevention. This

becomes even more important once the baby is born, as proper hand hygiene prior to handling the baby is an essential component of prevention of transmission to the newborn(11)(12).

Antiviral treatment

All pregnant mothers with severe/complicated disease or signs of progression of the disease (or even suspected cases) should be treated with the antiviral Oseltamivir. Treatment with antiviral medications should begin without waiting for collecting specimen or laboratory confirmation. Chemoprophylaxis is not recommended in pregnancy. Oseltamivir is safe for use even in the first trimester (13)(14). Treatment with Oseltamivir to a lactating mother is not a contraindication for breastfeeding. Antiviral drugs will help to relieve symptoms and reduce viral shedding, but therapy is more beneficial if started within 12-48 hours. Neuraminidase inhibitors are used and they are as an adjunct to immunization. Oseltamivir is taken orally and zanamivir is administered via inhalation. Both are category C drugs but oseltamivir is preferred over zanamivir due to its well evidenced results from clinical experience with a guaranteed systemic absorption. Efficacy of antiviral agents during pregnancy, and especially in severely ill pregnant women, is unknown. Clinical studies in non-pregnant women indicate maximal efficacy when given early in infection or used for prophylaxis, before development of severe disease (15).

Labour and newborn care

It is vital to afford routine intra-partum and postpartum care with attention to specific complications related to child-birth, the postpartum period or the newborn. The newborn baby should not be detached from the mother even if she has seasonal influenza infection. Mothers should wear a disposable /surgical face mask and practice hand hygiene before and while feeding or handling the baby. Support the mothers to initiate and continue breastfeeding and to breastfeed frequently and exclusively on demand. If mother is ill, she should be helped to express her breast milk and feed it to the infant. Newborns of infected mothers should be observed for development of infection. Newborn infants are unlikely to have typical influenza signs. Influenza or its complications in newborn infants may begin with less typical signs such as apnoea, fever, fast breathing, cyanosis, excessive sleeping, lethargy, feeding poorly and dehydration. Newborn infants with severe or deteriorating illness and those at risk of more severe or complicated should promptly be treated with anti-viral drugs (16). Immunization is recommended to pregnant women especially during flu season. They are low-cost interventions that have been shown to have substantial benefits for both mother and baby(17).

Breastfeeding

Breastfeeding is encouraged in H1N1 influenza although H1N1 transmission through breast milk is unknown; breastfeeding strengthens the neonatal immune response and infants who are

bottle fed may be prone to getting a viral infection. If the infant needs to be isolated from the infected mother, the infant should receive bottle feedings of expressed breast milk until the mother and infant can be reunited.

Conclusion

Influenza in pregnancy is a significant and under-appreciated public health problem. It causes considerable morbidity and mortality and impact can be alleviated by education of women and their physicians as well as by vaccination and use of available preventative and therapeutic modalities. Public health measures that increase vaccination rates are key to these efforts. It is important that physicians educate their patients regarding the increased severity of influenza infection in pregnancy and that influenza vaccination be offered to every pregnant woman, as well as every woman considering becoming pregnant.

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Dealing with mass casualties: An outlook

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Abstract :

It was one of those rare and calm non casualty weekend, as it was an Easter Sunday, time for peaceful worshipping and a perfect get together with families and friends at home. Most of the patients have been discharged on previous day with appropriate plans of management to spend the holy day with their loved ones. I had only about ten patients in the ward to attend to. Most of them were with chronic wounds, either staying in the ward to receive their course of antibiotics or awaiting a wound toilet in the casualty theatre that evening. Ward was void of all the hullabaloo of a double casualty weekend. In short, it was one of those beautiful Sunday Mornings in the life of a surgical trainee. What could possibly go wrong?

I was doing rounds with one of the intern house officers and a nursing officer who was pushing the drug trolley ahead of us and delivering drugs to the patients, suddenly exclaimed "Doctor did you hear that noise?" I didn't hear any noise. I was busy calculating the dose of insulin my patient needed, to control his blood sugar levels. My intern, then said "Miss, something must have fallen down on the floor above." "No it seemed like a sound of a blast, doctor." We dismissed her concern with a "nah" and went on to advise our patient regarding the proper diet control in addition to Insulin injections.

A few minutes later, a minor staff who has gone with a patient to the ECG room, came in wheeling the patient with an air of excitement "Doctor, people down there are talking that there was a transformer blast in Zion Church and lots of small children who went for Sunday school have got inju-". Before he could finish, another one came in half running, uttering breathlessly "Doctor there was a bomb blast in Zion Church!" Before I could comprehend what was going on, I got a call from a colleague in 'Accidents and Emergency Department, in a rasping voice filled with an element of urgency "Machan we are expecting a mass casualty, can you come as soon as possible to the A & E? We need more hands it seems. Pass this message to the nursing in charge too, we need more nursing staff as well."

I made a gesture to my intern to tag along and together we hurried down the corridors, making our way to A&E. On our way, my intern got a call from his mother in Colombo, who has told him that there was news about bomb blasts in Colombo churches, and asking him to stay safe with motherly concerns. She was obviously at that moment wasn't aware of the blast in Batticaloa. My smart intern was also keen in not mentioning her about the 'news' he has just heard. He asked her and 'thatha' to be safe repeatedly with apprehension and not to worry about him as he was in one of those safest premises-the hospital. We knew something serious was going on. Our pace got its momentum and we saw familiar faces from other wards, doctors and nursing staff, all heading towards A & E.

We never would have imagined what we saw there even in our worst night mares! The serenity of that beautiful Sunday morning was shattered by sirens of ambulances, beeping of various monitors and of course by heart wrenching cries of those who have lost their families and friends!

Introduction

Over the last decade major incidents or mass casualties have been a rare phenomenon. The majority of the junior doctors would have gone so far entire career without facing one. However, modern day clinicians cannot afford to go unconcerned about it, especially when there is a significant rise in recent number of terror attacks worldwide.

While many practitioners are well trained regarding the management of severely injured individuals, only very few are competent in managing a mass casualty. In current circumstances it is of utmost importance to train the clinicians in view of managing a major incident with the limited resources available.

Definition for a mass casualty are numerous in literature. Mass casualty or a major incident can be defined as an unexpected natural or a man-made incident that results in large number of victims who need medical attention in a short period of time, thus overwhelming the resources of the health care system available at that point in time.

Natural disasters such as floods, earth quacks, landslides, tsunami and man-made unintentional incidents such as plane crash, train crash, gas leak or intentional incidents such as grand massacre, arson, active shooting are few examples of so called mass incidents.

When dealing with major incidents one should be aware of the fact that these situations impose an additional work load on top of the hospitals or health care system's usual work load. This usual work load doesn't go away i.e. A 60 years old patient with a sudden onset of crushing type of central chest pain with probable STEMI may present while one is involved in the management of mass incident and therefore near optimal service must be rendered in both circumstances.

Liaison of Emergency Services During a Mass Casualty

Various emergency services are usually involved in the management of a mass casualty. The police force and other security officers are often responsible for the overall command and control of the situation. It is their duty to preserve the scene and related forensic evidence if the major incident has resulted from a criminal act.

If the major incident has resulted from arson or a blast injury, firearm and rescue services play an important role in controlling the damage and providing safety.

Ambulance services are crucial in collecting and transferring the victims to nearby hospitals according to the triage. A properly organized pre-hospital service should be able to efficiently transfer the most seriously injured patients to the most appropriate hospitals (depending on the availability of resources needed to treat the patients at hand) and least injured victims later or to other hospitals as needed. Shortages in this service will result in single hospital being overloaded with large number of victims of varying degree of severity, leading to inadequate care to the victims.

Triage at the site of major incident

Triage is a rapid assessment and categorization of casualties according to their physical signs in to a set of treatment groups which correlates with the urgency of their need for treatment.

A correctly conducted triage will result in those who have greatest need receiving rapid treatment while those who can withstand a small delay without detrimental effects, being treated subsequently.

Triage is split in to two stages - sieve and sort. Sieve phase represents the initial categorization of patients in to P1- P4 based on respiratory rate and heart rate / capillary re-filling time. This is usually accomplished in matter of seconds. The next stage sort is much more labour intense and involves the measurement of systolic BP and GCS.

Moreover, patients in a triage category can change over time in either direction. Therefore, it is very crucial to re-assess and re-triage the patients in appropriate intervals to ensure that the deteriorating victims are identified early and are given adequate care without further delay.

Disaster response in the hospital setting

General considerations

Despite the fact that every disaster varies in nature, every hospital should have pre-structured plan of management that is applicable to all disasters. Ideally an incident command system is the most appropriate first step in response to major incidents. Incident command system is a standardized approach to coordinate an emergency response between multiple agencies. It has specific hierarchy with pre allocated functional roles to everyone involved in the care. It should be based on the functional demands, with flexibility so that it can be deployed in any major incidents according to the number of victims and the nature of the disaster.

Every hospital should frequently conduct disaster drills so that everyone involved in the management of a major incident can provide a better care in the time of need. It is crucial to understand that the ability to manage an inflow of severely injured victims is greater among well-trained responders than readily available volunteers at the time of incident.

Once the hospital is informed about a mass casualty, the pre-devised disaster management plan should be implemented, assigning the duties as specified in the plan. As the Emergency Department is the center of mass casualty care, the emergency department nursing officer in-charge must make sure that ED is cleared of current patients accordingly (patients who need inpatient treatment are transferred to wards while other non-emergent patients are discharged) and all qualified staff are reported to duty. He/she must ensure that adequate number of critical care beds are created in intensive care units, coronary care units and special baby units based on the information received. All non-emergent procedures and operations should be postponed except

for ongoing surgeries and operation theaters should be made available for the care of disaster victims. Blood banks should be notified and transfusion products should be made available.

It is important to bear in mind that large number of patients may self-present if the incidence has taken place within the reasonable distance of the hospital in addition to those arriving via ambulances. This can cripple the efficient function of the ED until they are appropriately triaged.

The safety of hospital should be ensured especially in the cases of massacre/active shooting with the help of security officers such as army officers, police, CIDs as there is a risk of secondary attacks and perpetrators might as well be among the victims.

The goal of disaster management is to provide the minimum acceptable care for the greatest number of patients rather than the usual standard treatment. For instance, damage control surgeries preferred rather than definite care. In addition, mass casualties often lead to situations where moral and ethical values are taken in to consideration while making difficult decisions.

Even though the ED seems to be the important setting in the management of a major incident, its role in the disaster response is short lived, may be for initial few hours. It is actually the other parts of the hospital such as intensive care units, operation theatres and wards that provide major contribution in the care of victims for a variable duration lasting from days to weeks and even months.

Triage in the hospital

The most experienced medical officer especially with the type and severity of the injuries unique to the mass incident being addressed, should be in charge of categorizing the patients according to the likelihood of survival, severity of injury and available resources.

Communication

It is of great importance to establish a large information center to help families and friends to locate their loved ones and to receive regular updates about them once they are removed

from ED for further specific management. Nursing officers can extend invaluable service by explaining patient's condition and the necessary treatment. Appropriate steps should be taken to ensure correct patient identification. Moreover, communication networks can be shutdown either by law enforcement authorities or overload, therefore it is wise to be prepared for such challenging situations.

Recovery and Rehabilitation

A mass casualty can impact a person's life in physical, emotional and socio-economical standpoints. The most important part in the response to major incident is thus to provide ample opportunities for the patient and his family to rebuild themselves. Medical care provided in the hospitals lead successful physical recovery while various other organizations both government and non-government extend their help in the community by giving counselling, fundraising to reestablish an economic source, housing etc. Eg: Tsunami (2004) rehabilitation programs.

Summary

In view of recent rise in mass casualties, especially due to terror attacks, it is of utmost importance that every clinician is competent enough to deal with such adversities, in an efficient manner by being able to provide good quality service, with limited resources. Similarly, a properly formulated major incident plan of management and preparedness will make a huge difference in the quality of outcomes.

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A Novel technique of botulinum toxin injection to upper head of lateral pterygoid muscle & observation for improvement TMJ disc derangement disorder.

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Introduction

TMJ disc derangement disorder is the most common disorder of TMJ affecting young population in the world. Almost all the patients attending to maxillofacial clinic either suffer from unpleasant click, pain, restricted mouth opening or any combination of those symptoms depending on severity & stage of the disease. This condition often interfere with patient's lifestyle resulting mental stress. Various treatment modalities have been proposed in medical literature for the condition showing that there is no single curative treatment option for this condition. First line treatment is medical treatment. Monthly follow up is essential for adjustment of therapy. Some patients do not get satisfactory results with those treatment & most of them become chronic in maxillofacial clinic without subjective satisfaction. We hypothesize the lateral pterygoid muscle is the muscle responsible for TMJ disc derangement disorder due to its chronic spasm. We paralyzed the lateral pterygoid muscle & observed the improvement for pain, mouth opening & click. The results were rapid & predictable. Almost all the patients were satisfied. We used this technique as first line therapy for disc derangement disorder.

Material & Method

Patient selection

We have selected 21 patients with age range between 25-35 years irrespective of gender. All the patients were medically fit & well. There were no history of previous hospitalization or on regular prescriptions. Previous Botox injection & allergy history were excluded. All selected cases were unilateral disc derangement disorder without history of prior treatments for the condition. Eleven cases were diagnosed as anterior disc displacement with reduction & ten cases were anterior disc displacement without reduction. Diagnosis were made on clinical ground. Degenerative joint disorders & other TMJ disorders were excluded by clinically

& radiologically. Other facial pain disorders also excluded. Ethical approval was taken. Patients were explained the procedure & informed consent was taken.

We recorded pre injection pain score (VAS), Click frequency, Mouth opening (Inter incisal opening) as clinical monitoring tool.

Armamentarium

25 units of Botox(Ref - 1) were mixed with 01 ml of 0.9 percent normal saline & Loaded in 01ml insulin syringe. This was done by the same operator to avoid the bias. 26G, 05cm long dental needle was used for the injection. The needle length was determine by observing series of adult skull & 1:1 ratio adult CT scan which was taken in radiology department for other purposes in different patients in same age group (eg. Suspected head injury patients). This is to minimize radiation exposure of our study group patients & minimize the cost. The length was measured from anterior surface of condylar head to upper most point of lateral surface of lateral pterygoid plate. The average length obtained was 4.0cm. Additional 01 cm was added to cover the soft tissue thickness over the zygomatic arch. This was determined by observing the soft tissue window of CT. Finally we used the 26G dental needle which is 05cm length for the injection.

Surface marking & Injection Technique

Lower border of the zygomatic arch was drawn upto tragus. Anterior border of condylar head was marked while the patient kept in occlusion. The point 5mm from the anterior surface of condylar head & inferior border of zygomatic arch was marked. It was used as point of insertion. Pt was kept in Dental chair in sitting position with back rest at 90 degree to horizontal plane. Head was positioned at shoulder level of the operator. Topical

anaesthesia was applied for 5min over the injection site. Needle was inserted through the point of insertion while pt was asked to keep in occlusion. The direction of needle was 45 degree lateral to vertical plane running through the lateral border of zygomatic arch & 5-10 degree inferior to horizontal plane running through the inferior border of zygomatic arch. This direction was already defined by previous CT study & skull study. Further we used ultrasound guided needle insertion for first two pts & confirmed our direction of insertion. However our USS couldn't visualize the pterygoid plate. Echogenesity was poor at that depth.

Needle was advanced until lateral pterygoid plate is hit. We confirmed the bony hit & withdrawn needle about 2mm & then aspirated to confirm needle is not within the vessel & 5U of Botox released. Then again the needle was withdrawn 5mm & aspirated & 5 U released. This was repeated along the path of

needle removal which is same as needle insertion path. Empty needle & syringe was discarded according to regulations. Pressure pack was placed over the needle prick for 10 min with ice pack on top of it. Pt was observed for vital signs for 20 min & sent back to home with instructions. We observed blood pressure, respiratory rate, GCS, Pulse before the procedure & after procedure for every 10 minutes for 30 min. Only one case showed dizziness, high blood pressure, tachycardia, light headedness. Even though we can't explain it, this could be due to anxiety as well. Others didn't show any systemic upset.

Pt was reviewed every two weeks up to six weeks to assess Pain, Click frequency, Mouth opening & any post injection complaints:

Results

Our results are as follows.

Case	Age	Sex	M.O Pre	Pain Pre	Click Pre	M.O 2wk	M.O 4wk	M.O 6wk	Pain 2wk	Pain 4wk	Pain 6wk	Click 2wk	Click 4wk	Click 6wk
1	30	F	30	8	NO	16	36	41	2	0	0	60	NO	NO
2	28	M	26	7	NO	30	43	45	1	0	0	80	NO	NO
3	30	F	30	5	Yes	32	30	22	0	5	4	Yes	Yes	Yes
4	25	F	35	6	Yes	40	42	46	2	0	0	NO	NO	NO
5	32	F	40	4	Yes	41	43	43	2	1	0	70	85	90
6	27	F	30	5	NO	37	53	56	4	3	1	NO	NO	NO
7	28	M	32	7	Yes	39	42	45	0	0	0	NO	NO	NO
8	24	F	25	6	NO	40	43	44	0	0	0	25	50	NO
9	34	F	40	5	Yes	44	44	46	0	0	0	85	NO	No
10	26	F	25	6	NO	29	26	26	3	4	4	N/C	N/C	N/C
11	30	F	40	4	Yes	43	43	46	1	0	0	NO	NO	NO
12	29	F	24	7	NO	40	43	47	3	0	0	30	70	100
13	26	F	21	7	NO	29	32	44	2	0	0	80	NO	NO
14	27	M	42	5	Yes	44	44	46	1	0	0	95	95	NO
15	31	F	20	6	NO	22	27	36	2	0	0	80	NO	NO
16	28	F	29	4	Yes	30	38	41	2	2	0	90	NO	NO
17	33	F	10	8	NO	10	31	37	2	0	0	90	NO	NO
18	29	F	20	6	NO	24	28	33	3	0	0	90	NO	NO
19	26	M	38	5	Yes	40	42	46	0	0	0	80	NO	NO
20	30	F	40	4	Yes	45	46	45	0	0	0	Yes	NO	NO

(Abbreviations : F- female, M- male, Pre- before injection, M.O - inter incisal measurement of mouth opening, Wk- weeks after injection, Post injection click improvement was assessed with visual analogue scale 0-100 percent (100 percent improvement shown as NO)

We reviewed patients every two weeks interval. All patients showed improvement of pain score 3 or below 3 within first two weeks of injection. 15 of them showed complete resolution of pain within second two weeks. Others showed persistent pain score of 3. Nearly 17 patients showed complete resolution of pain after 6 weeks. Others were within mild pain score. Clicking within first two weeks about 70 percent.

Discussion

TMJ is the only atypical synovial joint exist in head & neck area where the mandibular condylar head articulate with temporal bone & a fibrous disc in between two articulating surface. It plays role of mandibular movements therefore it is commonly affected by disc derangement disorder which is a one category of many TMJ disorders. Majority of young population in the world

suffering from this condition resulting in high number of patient attendance in maxillofacial clinic in every year. Most of these patients are suffer from unpleasant click (A noise from the joint), progressive worsening joint pain, disordered mouth opening & later restricted mouth opening. Various aetiological factors & pre disposing factors have been described in medical. Micro trauma & macrotrauma to the joint are the main aetiological culprits (Ref - 16). Micro trauma includes; parafunctional habit like nail biting, pencil/ needle biting, Premature incisor contact & anterior displacement of jaw due to dental malocclusion, bruxism, habitual protrusion of lower jaw to mask the prognathic maxilla . Macrotrauma includes; direct hit to the joint, iatrogenic trauma like excess mouth opening during dental procedure etc. Mental stress is a major predisposing factor for this condition. It act like a vicious cycle of the condition. It means mental stress causes

the TMJDD & Vice versa. Therefore there is no specific aetiological factor for this condition & because of lack of aetiology clinicians are struggling to manage this disorder resulting in unsatisfactory results.

There are various types of treatment modalities have been described for TMJDD including medical treatment, invasive treatment, and surgical treatments. None of these became a successful option & often clinicians combine these options to get satisfactory results. Even in our center we did same kind of multimodal treatments to get certain results but there wasn't complete improvement. We used to introduce medical treatment & physiotherapy first (NSAID, Steroids, Muscle relaxants & antidepressants, Retrusive jaw exercises, Bite raising appliances, Heat therapy, Massaging). But even with these options it take so long time to improve the condition & some patients end up with unchanged results. Then we tried with needle arthrocentesis which also often gave temporary results. We were reluctant to do surgical procedure like eminectomy, discectomy, disc repair as it could result in high chance of intra articular ankylosis & facial nerve damage.

According to aetiopathogenesis we hypothesize that the lateral pterygoid muscle is the major muscle play a role in this disorder as it is the major muscle contributing of mandibular translator movement along the articular eminence on mouth opening. Therefore we thought paralyzing this muscle with BOTULINUM TOXIN TYPE- A (BOTOX) (Ref - 1) should alleviate the problem.

Therefore we have selected new cases came to our unit from 01st /January/ 2017. All cases were unilateral TMJ disc derangement disorders. All of them were disc displacement with reduction & without reduction. They haven't treated before with any treatment modality except simple analgesics were taken by patients as their own. Other facial pain related conditions were excluded. Patients were informed about condition & consent was taken after explanation of the new procedure. We have recorded the presenting signs & symptoms in three aspect 1. Pain score 2. Click frequency percentage according to patient 3. Mouth opening (we recorded the inter incisal opening). Our idea was to inject the toxin extra orally through zygomatic sigmoid notch. Because this approach gave the total paralysis of upper head of the lateral pterygoid muscle belly & easy access. Our idea was to inject the solution from lateral pterygoid plate to muscle insertion to articular disc. It was a challenge to identify the needle point of insertion & depth of injection. To tackle this problem we analyzed the 1:1 CT scan axial cuts through the condylar head & pterygoid plate which was taken for other purposes in our radiology department such as head injured patients, Intracranial pathology patients. We also analyzed the laboratory natural adult skull to measure the length from anterior surface of condylar head to mid-point of lateral pterygoid plate. This effort was to eliminate radiation risk to our pts & minimize the cost for the procedure. Finally we

could be able to get the average needle length & direction as mentioned above (in Material methods). For first two cases we injected the toxin with ultrasound guide. Once we confirm our direction of insertion is correct then we did not do any further ultrasounds for needle insertion. This helped the procedure to be done in clinic set up. Prior application of topical anaesthetic cream & ice pack placement over preauricular were helped to get reasonable analgesic effect during injection. We did not give sedation as this technique is new & we would expect some CNS effect/complications after the injection. We used low dose of toxin as about 25 unit according to muscle size & literature report of intra oral pterygoid paralysis techniques. Yozuda et al has described a similar technique of injection but it doesn't describe the exact injection needle insertion point. Here we have introduced a point of insertion of needle exactly. (Ref - 14,15)

Conclusion

Our novel technique of botulinum toxin injection to lateral pterygoid was safe in selected 20 patients & improvement of disc derangement condition with our technique supported our hypothesis. This technique improved the patient's condition satisfactorily. Further studies may be necessary to analyze the results in larger samples.



Injection point

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CENTRAL LINE ASSOCIATED BLOOD STREAM INFECTIONS (CLABSIs) Frequently asked questions (FAQs)

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1. What are central line associated blood stream infections (CLABSIs)?

Blood stream infections (not related to an infection at another site) in a patient on central vascular lines for ≥ 48 hrs, are called CLABSIs.

2. What are the central lines, associated with CLABSIs?

- Neckline - Internal jugular venous line
- Subclavian venous line
- Femoral venous line
- Peripherally inserted central catheter (PICC lines): not commonly
- used in SL

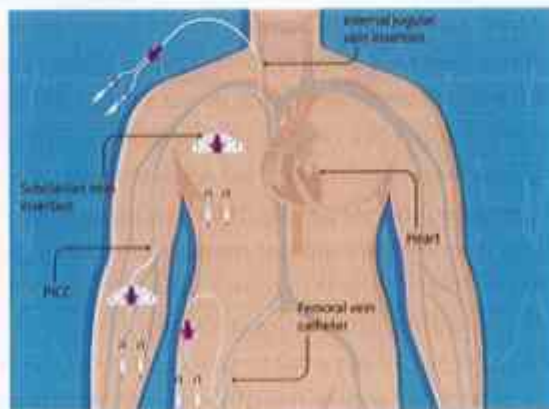


Figure 1: Central Venous Catheter

3. Why are CLABSIs important?

CLABSIs increase morbidity, mortality and healthcare cost. Placing a central line is an integral part of critical care management. Therefore, patients following major surgeries, patients with Dengue Hemorrhagic Fever or with any other conditions which require critical care can develop CLABSIs. CLABSIs can be the immediate cause of death in those patients.

4. What is the pathophysiology of CLABSIs?

When a patient receives critical care in an ICU for ≥ 48 hrs, he/she gets colonized with multi-drug resistant organisms (MDROs). Figure 2 illustrates the mode of entry of microorganisms causing CLABSIs. When a central line is placed, natural skin barrier is bypassed by the skin organisms including MDROs and organisms enter the blood stream in between the skin and the line. Organisms form a biofilm around the central line. Biofilms protect the organisms from the antibiotics and immune components. Biofilms act as source of infection which can only be eradicated by removal of the line.

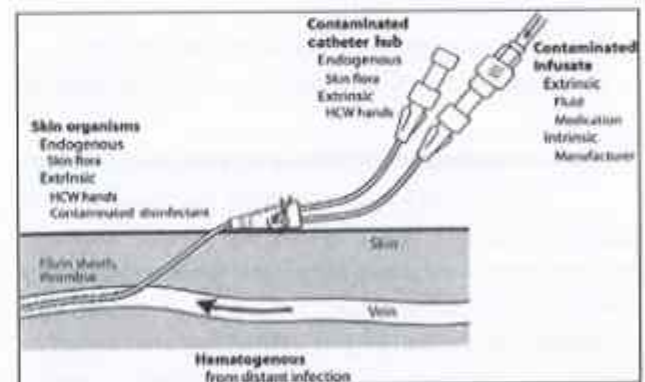


Figure 2: Mode of entry of microorganisms causing CLABSIs.

MDROs enter through contaminated central line hubs through which the medications are injected. Contaminated medications and fluids also can be the source of infection and infections at other sites can also be the source. Contaminated healthcare workers' hands are the major mode of transmission which contribute to skin colonization, hub colonization and contamination of medication while preparing. As the central lines directly open into the superior or inferior vena cava, any organisms which enter through/along the line will easily reach heart. If the patient has damaged heart valves, he/she can develop infective endocarditis of right heart. Quality and quantity of organisms entered and the body's reaction to the organisms determine the clinical outcome.

5. What are the challenges in managing CLABSIs?

- Limited choice and availability of effective antibiotics due to the multi drug resistant nature of causative agents.
- Cost and toxicity of the effective antibiotics (Glycopeptides, Colistin, Amikacin, Streptogramins etc.) are much higher than that of other common antibiotics.
- Potency of the antibiotics effective against MDROs are less than that of common antibiotics.
- Persistent source of the infection – the infected lines, cannot be removed in many cases as limited vascular access.
- Comorbidity of the patients – Diabetes mellitus, Chronic Kidney Disease, Chronic Liver Cell Disease, Malignancy, acute trauma and severe infections (Eg. Dengue, Sepsis, Etc.) can cause immunosuppression which worsens CLABSIs.
- No surveillance data available to plan effective management protocol.
- Inadequate supply of necessary items – consumables as well as equipment (alcohol hand rub, surgical spirit, transparent dressing, kidney trays, etc.) are not available in adequate amounts at present Sri Lankan settings.

6. How can CLABSIs be prevented?

- Good hand hygiene compliance of Health Care Workers (HCWs).
- Following aseptic non-touch technique (ANTT) in insertion and maintenance of central lines.
- Ultrasound scan guided insertion of central lines.
- Review the line daily to decide to remove or keep the line.
- Adequate staff training using simulation on insertion and maintenance of central lines.
- Applying transparent dressing at the site of insertion.
- Appropriate cleaning and disinfection of touchable surfaces.

7. How does hand hygiene compliance of HCWs prevent CLABSIs?

When HCWs practice the WHO's 5 moments of hand hygiene as indicated in Figure 3 by using alcohol hand rub (if hands are visibly clean) or using soap and water (if hands are visibly dirty or dealing a patient with diarrhoea), transmission of infections from one patient to another will be significantly reduced. And it will reduce colonization of MDROs on patients' skin and central line hubs.



Figure 3

8. How does aseptic non touch technique (ANTT) prevent CLABSIs?

Aseptic non touch technique means, not touching the critical parts which come in contact with patient's sterile areas, even with sterile gloves while doing aseptic procedures. While inserting the central lines using ANTT, the tip of the line and the patient's skin after applying antiseptics are not touched even with sterile gloves. And using ANTT while infusing medication via central lines, the hubs are disinfected using alcohol wipes and the tips should not be touched even with sterile gloves. ANTT will significantly reduce the contamination of CV line tips and hubs.

9. How does USS guided insertion of CV lines reduce CLABSIs?

USS guided insertion minimizes the tissue damage and it eliminates the chance of tricuspid valve damage by the guide wire.

10. How can staff training on insertion and maintenance of CV lines be arranged and monitored?

Continuous professional education is an essential component which is frequently neglected in Sri Lanka. Simulation training on insertion of CV lines should be a compulsory component of training of medical officers attached to the ICUs and the Anaesthetists should monitor their competence. Infection control team should arrange training of nursing staff attached to the ICUs and should regularly monitor the staff competence and compliance of ANTT while handling CV lines. Quality management unit should monitor the staff training programs.

11. How hand hygiene compliance be monitored?

According to the WHO's estimation, a HCW in an intensive care unit will have around 20 moments of hand hygiene per hour of patient contact and around 12 hours of patient contact a day. For each moment 2ml of alcohol rub should be used. It reveals one HCW should use $20 \times 12 \times 2 \text{ ml} = 480 \text{ ml}$ of alcohol hand rub per day. That means an ICU should use around 500ml of alcohol hand rub for each patient per day. Therefore, usage of alcohol hand rub is an effective indirect measure of hand hygiene compliance. This is better than current hand hygiene auditing where one infection control nursing officer physically be present in the ICUs for just 20 min a day for direct monitoring of hand hygiene. Most of the hospitals reveal more than 80% compliance while the consumption of alcohol hand rub is grossly inadequate. This discrepancy should be addressed.

12. Why is transparent dressing better than that of traditional dressing of site of insertion?

Infection at the site of insertion can lead to CLABSIs. Early detection and management of site infections are important. Transparent dressing will allow to examine the site of insertion without touching the line and adjacent skin and therefore facilitates

early detection of site infections. The ICUs should request adequate number of transparent plasters and the administration should ensure timely supply.

13. How can surface cleaning and disinfection be ensured?

Disinfecting all the touchable surfaces at least twice daily will reduce the amount of bacterial colonization and thereby CLABSIs. Applying standard chlorine preparations are cheaper, however there are practical difficulties such as corrosive nature of chlorine which damages metal surfaces and tiled floors if applied in higher concentrations. Commercially available disinfectant wipes are effective but costly alternatives. Supply of these wipes to selective units will be a useful option.

14. Why should we limit the visitors to the ICUs?

CLABSIs are hospital acquired not community acquired infections. Therefore, role of the visitors in causing CLABSIs is

minimal. However, limiting the visitors to ICUs is important as to mitigate other ethical issues.

15. Why should we limit the number of healthcare workers visiting ICUs?

Healthcare workers including specialty teams visit ICUs to review patients. Each team loves to bring their whole crew. ICUs should follow a strict policy of limiting the number of HCWs present in the unit at a time. Because HCWs carry hospital acquired pathogens from one unit to the other and limiting their number will reduce the chance of transmission of organisms.

As it's a teaching hospital, ICUs should accommodate medical as well as nursing students as part of their teaching and learning process. However, ICUs should limit the number of students present at a time.



Easter suicidal bombing in Sri Lanka- the analysis of victim management at teaching hospital, Batticaloa.

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Introduction

Explosions can produce unique patterns of injury seldom seen outside combat. When they do occur, they have the potential to inflict multisystem life-threatening injuries on many persons simultaneously. The injury patterns following such events are a product of the composition and amount of the materials involved, the surrounding environment, delivery method (if a bomb), the distance between the victim and the blast, and any intervening protective barriers or environmental hazards. Because explosions are relatively infrequent, blast-related injuries can present unique triage, diagnostic, and management challenges to providers of emergency care.

After the tragedy of Tsunami in 2004 and since the end of Sri Lanka's bloody civil war a decade ago, the multi-ethnic city Batticaloa was hit by a Man-made National Disaster on April 21st. Terrorist suicidal explosion happened brutally on an Easter Sunday at 9.05am at Protestant Zion Church when nearly 300 worshippers were attending the prayer. The church is situated just 800m distance from the Teaching Hospital Batticaloa.

Because the blast was at the bottle neck pathway to church, rescuers faced a big challenge in rescuing the casualties from the site. They ultimately broke the rear parapet wall to scoop them up to hospital. This made the variation in arrival time although the incident happened nearby. Casualties were brought to A&E, mostly by private vehicles and also by hospital ambulances. Even rescuers were running into the A&E carrying victimized

children on their hands. There were 22 dead bodies, directly brought to hospital mortuary, with body parts of suicide bomber discovered later.

In the Batticaloa hospital, the majority of emergency medical service (EMS) systems are organized and coordinated at the local level. Only Few health professionals have experience with explosive related injuries in the past, this results in an incredibly diverse hospital emergency medical care system that is often markedly different in operational and clinical approaches among jurisdictions.

At the time immediate Notification to staff was through various formal & informal channels. Even Director was directly informing to command staff & other major stakeholders. In the meantime, incident became viral on social media, and in no time almost all the local health staff gathered at A&E. Even doctors who were on New Year vacation in their hometown Batticaloa joined the regular staff to offer support.

Triage Officer was the On-call Consultant Surgeon and the Clinical Commander was the Senior Consultant Anaesthetist. There were nearly 15 multidisciplinary specialists and another 15 PG trainees contributing for the management of casualties. Director was on the floor trying to coordinate the health response for this unexpected tragedy to her level best. There were plenty of health staff available at A&E, at least one medical officer, two nurses and two health assistants caring for each patient.

Literature review

Pathophysiology

Primary blast injuries are due to pressure differentials (Table 1). Alternating over pressure and under pressure imposed upon tissues of heterogeneous densities (particularly air filled organs) impart local and systemic effects. The tympanic membrane (TM) is particularly sensitive, rupturing at pressure differentials as little as 5 pounds per square inch (psi). Higher pressure differentials (56-76 psi) provoke disruption of lung architecture owing to the compression and expansion of alveoli, resulting in shear of the alveolar capillary barrier. Breakdown of pulmonary structural integrity can lead to air emboli, pneumothorax, contusion, hemothorax, pulmonary hemorrhage, mediastinal air, and subcutaneous air. As alveoli become flooded with fluid and cellular debris, it results in "blast lung" with difficulty in maintaining oxygenation secondary to ventilation perfusion mismatch. In the bowel, contusion, ischemia, overt necrosis, and perforation can occur. Bowel injured by pressure waves, although rare, has the potential for delayed presentation. Mild traumatic brain injury (mTBI) can be associated with persistent and often debilitating symptoms. Laboratory studies would suggest that this is due to free radical release and neuronal cell death.

Secondary blast injury occurs when fragments of the device or debris picked up by the blast wave is accelerated toward the victim, causing penetrating wounds. These are more common than primary injuries. Injuries to the extremities dominate in survivors though associated vascular injury can be quickly lethal. There is far greater degree of soft tissue involvement as there can be a large amount of fragmentation, and it is also typically delivered with high energy. The passage of these projectiles through human tissues can be associated with cavitation 20-25 times the size of the fragment, and local pressures can exceed 100 psi. This promotes wounding of areas away from the primary injury site and these wounds can present in a delayed fashion. Although the association of cavitation and remote injury is disputed, the large burden of fragmentation can also result in wounding removed from the site of initial injury.

Crush syndrome can occur as skeletal muscle is compressed, releasing myoglobin, urates, phosphates, and potassium along with cellular death. Untreated, renal dysfunction with cardiac arrest can also follow. Traumatic asphyxiation results from entrapment of the thorax and inability to ventilate. Entrapment can also lead to compartment syndromes even after the victim is removed from the rubble. The initial pressure wave can forcefully dislocate victims, and they can be thrown against fixed objects or the ground, causing a myriad of blunt injuries.

The thermal effects of blast exposure and other environmental factors cause quaternary injuries. This includes burns (due to chemicals or heat) and inhalation injuries.

Others would define quinary injuries to describe effects directly related to particular additives to a device such as infectious or radioactive substances ("dirty bombs").

TABLE 1. Mechanisms of Injury after Blast Exposure

Nomenclature	Mechanism	Injuries
Primary	Alternating Overpressure and Under Pressure on Tissues of Heterogenous Densities (*Air Filled*)	Tympanic Membrane Rupture, "Blast Lung," Hollow Viscous Injury, mTBI
Secondary	Fragmentation or Airborne Debris Accelerated Upon Victim	Multiple Penetrating Injuries, Extensive Tissue Devastations, Traumatic Amputations
Tertiary	Structural Collapse, Violent Displacement, Entrapment	Multiple Blunt Injuries, Traumatic Asphyxiation, Crush Syndromes
Quaternary	Thermal, Other	Burns (Thermal, Chemical), Inhalation Injury, Other

Results

The travel distance from the scene of blast, Zion church, Batticaloa to teaching hospital Batticaloa is 800m. This hospital, which is a tertiary care centre, was the only hospital which received the victims of the Easter bombing. Private vehicles were used initially to transport the victims; however, it was supported by ambulances. A proper field triage was not reported at the scene. The initial triage was done at the accident and emergency unit of teaching hospital Batticaloa. Triage officers were the on-call consultant surgeon and the senior consultant anaesthetist. Anaesthesia & critical care, general surgery, emergency unit and radiology teams were involved in the initial management, whereas the other specialities: ENT, plastic & neuro surgery teams also joined in. Blood bank was fully operational to cater the requirements of mass casualties.

As an immediate response to the blast, all the operating theatre were opened for trauma surgeries. The patients who received intensive care treatment were mobilized and rearranged. Medical intensive care unit and coronary care unit were prepared to receive patients who need critical care treatment.

We received total of 75 admissions from the incident. 63 on the incident day, 6 on next day and rest thereafter. Until 30th April, we received victims with minor injuries and who recognized the injuries later. Out of this 75, 63% were females (47) and 37% males (28). 25% were children & teenagers (19), 59% were young adults & middle aged (44) and 16% were elderly (12). Six bedded Resus Unit, twenty bedded Treatment Unit and sixty bedded Short Stay Unit at A&E building were completely occupied. As the number of casualties were manageable at A&E block, in patients of the hospital were not evacuated.

First medical contact was within 10 minutes of arrival for 70% of victims. 92% of casualties attended within 15 minutes of arrival. 31% of the first contacts were Consultants, 20% were Senior Registrars, 14% were Registrars and 35% were grade medical officers.

Several units were involved in providing post resuscitation & postoperative care of these victims. Majority (44%) were cared

in Short stay units (SSU) and 27% in Casualty Receiving Wards. 15% of casualties were managed in Critical Care Units. Nearly 5% of victims were discharged on the same day after initial care and another 5% was gone missing. 2 patients were transferred out immediately for neurosurgical interventions, one to Teaching hospital, Kandy with spinal injuries and other one to teaching hospital, Anuradhapura with head injuries. Another 5 were transferred later for specialized care, 2 to NHSL Burns Unit, 2 to LRH for plastic & eye interventions and 1 to Chest Hospital Welisara for thoracic intervention.

In addition to 22 public deaths from the scene, additional 4 died at resuscitation room, due to inevitable injuries to head, chest and abdomen failing resuscitation. Out of this 4, three were children which was heart-breaking to our staff. Later, 3 patients cared in ICUs at teaching hospital, Batticaloa succumbed to death due to various complications. The one who was transferred to TH Kandy also died after a long period of postop. ICU care. The last victim died at North Colombo teaching hospital, Ragama was a 20y old girl, She was treated in SICU at teaching hospital, Batticaloa for 41 days and the rest of 80 days at ICUs in Chest hospital, Welisara and North Colombo teaching hospital, Ragama. So, the total death toll reached to 31 up to date. Only 3 patients i.e. 4% of all casualties, had a hospital stay of more than 30 days. One at THB, one at LRH and another one at Welisara.

Regarding the site of injuries, majority had limb injuries (57%) followed by head & neck injuries (32%), chest (15%), abdomen & pelvis (12%) and spine injuries (1%). Majority of the injuries were penetrating shrapnel injuries and flash burns.

40 trauma surgeries were performed in 5 theatres & 3 anesthetic room spaces, stretching nearly about 20 hours operating time. Out of them 90% were primary surgeries (36) and 18% were major surgeries (7) including laparotomies & thoracotomies.

Discussion

The Easter terror bombing on 21st of April 2019, targeting several churches and hotels in Sri Lanka had caused various degree of physical and mental trauma to the victims. It was an unexpected tragedy causing death, permanent disability and other injuries involving all parts of the body. Death and disability of family members had triggered a series of long-term problems related to insecurity, finance, psychological sequelae etc.

Blast occurred at Zion church, Batticaloa, is a part of this serial bombings. It had caused same consequences. The victims from this church bomb were managed solely by the teaching hospital, Batticaloa.

Bomb blast generates a shock wave which disrupts the internal organs and tissues. It causes penetrating injuries due to the sharp or blunt objects. Flash burns also occur because of the fire that erupts at the time of the blast. These are the common causes of injuries in an explosion^{1,2,3,4}. The extent of injuries is intensified if it's in a closed compact space compared to an outdoor blast^{5,6}. Zion church was packed with church members at the

time of bombing as it is a special day in church, the Easter. The suicidal explosion occurred at the bottle neck entrance of a closed building had caused intense injuries.

The travel distance from the scene to hospital is only 800m, but there was a delay in arrival to the hospital. 19% of victims arrived within 15min and 44.4% arrived within 30min from the incident. This is could be due to improper field triage, overcrowded public etc. However, the 1st medical contact was within 10min for 70% of victims. For 51% of the victims, the 1st contact medical professionals were consultants and senior registrars in surgery and anesthesia. Medical staff from teaching hospital Batticaloa and those who were on holiday to Batticaloa joined immediately to cater the needs of the victims.

The injury pattern of this suicidal bombing revealed that the commonest injury was trauma to limbs involving soft tissues and fractures. It accounted for 57% of the injuries. This is consistent with the common blast injury pattern where minor, non-life-threatening injuries make the majority⁷. In 32% of victims, the site of injury was head and neck. However, in addition to the traumatic brain injury, the shrapnel in this region were also included. There is an overlap in the percentage due to the polytrauma involving multiple sites. Burns and tympanic membrane damage were also in significant numbers.

The incident happened on a Sunday where routine surgeries are not performed. This enabled the theatre staff to proceed with trauma surgeries in all operating rooms. There were 7 major surgeries including laparotomies and a thoracotomy. Majority of the surgeries were wound debridement.

All treated victims were given with diagnosis cards irrespective of their magnitude of the injury. A detailed diagnosis sheets were given for the patients who were treated in intensive care units and those who were transferred out.

According to the Institute teaching Hospital Batticaloa, EMS systems are challenged by the following key issues: insufficient coordination, response time disparities, inconsistent quality of care, lack of disaster readiness, divided professional identity, and limited evidence base for the profession. Even though it faces these challenges, the hospital medical care system will play a pivotal role in blast event medical management by identification and transport of patients with potentially significant injuries to appropriate trauma centers, and direction of less injured patients to other medical facilities.

The Teaching hospital care systems play a critical role in managing the emergency medical response to this kind of mass casualty event. The quality of hospital emergency medical response will affect the quality of all subsequent clinical care activities, and it may directly affect patient mortality and morbidity rates. The complexity and scope of a mass casualty event caused by an explosion requires that hospital emergency medical care systems faced the following issues. Recognition of specific hazards associated with a terrorist bombing, such as secondary devices, environmental hazards (e.g., toxins, fires), structural instability,

Identification patients with significant blast related injuries, Effective communication with acute care medical resources and emergency management resources and Expedient patient triage to match available resources with patient needs.

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Polyglandular autoimmune syndrome type II (Schmidt syndrome) presented with Addisonian crisis ; A case report

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Abstract :

Polyglandular autoimmune (PGA) syndromes are collections of multiple endocrine gland insufficiencies. This is due to immune system attack against human own gland. There are mainly three types. Patients commonly present below 30 years of age. We report a case of PGA type II also known as Schmidt syndrome which was presented with atypical symptoms and signs including Addisonian crisis.

Keywords:

Polyglandular autoimmune, hypothyroidism, dilated cardiomyopathy, Carpenter syndrome, Schimidth's syndrome

Introduction

PGA-I is associated with candidiasis, hypoparathyroidism, and adrenal failure. Auto immune Polyglandular type I is diagnosed with two out of three. [1,2]

Polyglandular autoimmune syndrome type II (PGA-II) is the most common of the immune endocrinopathy syndromes. It must consists of autoimmune Addison disease in combination with thyroid autoimmune diseases and/or type 1 DM. This disorder may associated with Primary hypogonadism, myasthenia gravis, and celiac disease, Parkinson, Vitiligo, alopecia, ITP or sero-negative arthritis. [3,4] In autoimmune Polyglandular syndrome type III (PGA-III) it is typically affects middle age women. In this disorder, adrenal cortex is not involved. [5] This disease characterized by auto immune thyroiditis along with other auto immune diseases such as type I DM pernicious anaemia, vitiligo, alopecia, myasthenia gravis and Sjogren syndrome. [6] Genetic factors variation HLA -II gene and environmental factors involved in this syndrome. It is may be inherited autosomal dominantly.

Polyglandular autoimmune syndrome type 1 (PGA-1) is a rare disorder, with sporadic autosomal recessive inheritance. It is mostly presented with candidiasis, hypoparathyroidism, and Addison's disease and diagnosed with the mutation in the AIRE

(autoimmune regulator) gene.[6] Polyglandular autoimmune (PGA) syndrome type I, usually occurs in children aged 3-5 years or in early adolescence, but it can present in early part of the third decade of life too.[1]

PGA-II occurs usually around the third and fourth decades of life. Middle-aged women have shown an increased prevalence of PGA-II. This disorder associated with HLA-DR3 and/or HLA-DR4 haplotypes, and the pattern of inheritance is autosomal dominant with variable expressivity. [4]

The most frequent clinical combination association is Addison disease and Hashimoto thyroiditis (Schmidt syndrome). The complete tri-glandular syndrome involved such as Addison disease, hypothyroidism, and type I DM is called as Carpenter syndrome. [5]

Symptoms may presents according to the glandular involvement. Type I diabetes present with Polyuria, polydipsia, polyphagia, unexplained weight loss, intermittent blurred vision, and lethargy etc. Hashimoto thyroiditis usually presents with nonspecific and include cold intolerance, fatigue, somnolence, poor memory, constipation, menorrhagia, myalgias, and hoarseness etc. Graves's disease may present with heat intolerance, weight loss, weakness, palpitations, oligomenorrhea, and anxiety. Addison disease present with Anorexia, nausea, vomiting, weight loss, weakness, and fatigue. [3]

Serum autoantibodies screen test is useful in screening asymptomatic family members who may develop autoimmune

endocrine disease in the future. The screening panel includes autoantibodies to the following:

1. 21-hydroxylase- anti body against Addison disease.
2. 17-hydroxylase- antibody forms in ovarian failure.
3. Thyroid peroxidase (TPO) - they are diagnostic of Hashimoto's thyroiditis.
4. Thyroid-stimulating immunoglobulins (TSI) in patients with signs of hyperthyroidism are diagnostic of Graves's disease.
5. Glutamic acid decarboxylase-65 and islet cells - The antibodies are used to screen for type 1 diabetes mellitus.
6. Antitissue transglutaminase antibodies - These are used for diagnosis celiac disease. Other antibody include immunoglobulin-A (IgA) endomysial antibodies and antigliadin.
7. Parietal cell and anti-intrinsic factor antibodies - These are used to screen for pernicious anemia.[3]

In autoimmune Polyglandular syndrome mainstay of treatment is primarily hormonal replacement therapy according to the organ involvement.

History

Forty year old previously diagnosed hypothyroidism patient with defaulted treatment presented with aggravated hypothyroidism features such lack of energy, fatigability, depression, loss of libido, generalized body pain, cold intolerance and constipation for 2 weeks.

She complained mild shortness of breath with mild cough. No chest pain, No nausea, No vomiting, No dark pigmentation in the body. There were hypo-pigmented patches on left forearm for six months. According to the patient after the delivery of the last child, she had developed heavy vaginal bleeding. She is having regular menstrual period. Initially she was haemodynamically stable and her blood tests including hemoglobin, electrolytes, renal and liver profiles were normal. Then patient was started on Levo-thyroxine 100 micrograms daily. After that, she developed nausea, vomiting, dizziness, vertigo, excessive sleepiness, drowsy. Her blood pressure was 70/ 40 mmHg. There, she was treated with and IV fluids and inotropes. Her morning cortisol was 103 nmol/l. Her serum, serum Sodium 120 mmol/l, 24 hour urinary Sodium excretion 107 mmol/dL, Blood culture no growth, ECG T inversion in LII, LIII, AVF, V3-V6, ANA - Negative, 2D ECHO - Dilated cardiomyopathy. Short synacthen test was positive.

Then she was treated with the IV hydrocortisone 100mg 6 hourly for 2 days. Patient was clinically improved. Inotropes support was tailed off. Five days later patient was discharged with oral hydrocortisone, Fludrocortisone, Thyroxine.

Discussion

There is 40 years old female known hypothyroidism with defaulted treatment came with hypothyroidism features such lack of energy, fatigability, depression, loss of libido, generalized body pain, cold intolerance and constipation for 2 weeks. This

patient due to the hypothyroidism and defaulted treatment may leads to dilated cardiomyopathy as the complication. [1] Due to hypothyroidism, contraction is reduced, increase peripheral vascular resistance leads to increase diastolic pressure and decrease pulse pressure.[2,3]In hypothyroidism, gene expression defect in contracting system of the heart also leads to dilated cardiomyopathy. [4] If dilated cardiomyopathy was due to hypothyroidism, it can be reversible by starting thyroxine treatment. [3]

But after starting thyroxine treatment, patient became unstable with blood pressure was dropped patient complained vertigo and dizziness with excess drowsy and sleepiness. [5]T his patient is having already vitiligo and hypothyroidism, There is high chance for multi endocrine disorder with Addisonian crisis.

After giving the IV hydrocortisone, patient clinically improved Blood pressure became stable. So if any patient with hypothyroidism clinically ill, we have to consider poly endocrine involvement. In Addison disease patient without the glucocorticoid, giving the thyroxine may worsen the Addison disease. [3] Because thyroxine increase liver metabolite of glucocorticoid, So add thyroxine without placing glucocorticoid may leads to Addisonian crisis.

After giving hydrocortisone, patient became clinically, stable. Sometimes after giving hydrocortisone thyroid function also improved. This patient during her 4th child delivery, developed bleeding. But she is having regular menstrual period so, this is not favouring pan-hypopituitarism and Sheehan syndrome. This patient present with hyponatremia, low morning cortisol level and positive short synacthen test, most favouring for Addison disease. This patient is having hypothyroidism, Addison disease, alopecia, vitiligo and arthritis belongs to poly glandular type II syndrome.[4,5]

In poly glandular type II disease, if 3 components involved such as Addison disease, hypothyroidism or hyperthyroidism, type I DM, it is called as Carpenter syndrome.[4]

In poly glandular type II disease, if 2 components involved such as Addison disease, hypothyroidism, it is called schimidth's syndrome. [5] She is having Addison disease, hypothyroidism with alopecia and sero-negative arthritis. Therefore she was diagnosed as Schimidth's syndrome.[13]

At this time her blood sugar is under control. She is having regular menstrual period. But in the future patient may be progressively affected other components such as type I DM, hypogonadism, auto immune hepatitis, celiac disease and pernicious anaemia also.

So this patient must follow regular clinic and investigate endocrine function regular interval also help to early identification and prevent further complication.

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Blue cures blue ; Dapsone Poisoning with methemoglobinemia & haemolysis

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Abstract :

Methemoglobinemia is a potentially life-threatening manifestation of Dapsone poisoning characterized by functional anemia and tissue hypoxia. With the increasing use of dapsone for various indications, acute poisoning (either accidental ingestion or suicidal) is increasingly recognized. We report a case of twenty year old female presented to the emergency room with alleged history of consumption of about 150 tablets of Dapsone (about five hours after ingestion) and managed successfully. Methylene blue is the most effective antidote for acquired methemoglobinemia. When methylene blue is not available, alternative treatments such as ascorbic acid and hyperbaric oxygen can be useful. Prompt diagnosis of Dapsone induced methemoglobinemia and aggressive management of these patients results in good outcome.

Keywords:

Dapsone poisoning, cyanosis, methaemoglobin, haemolysis, methylene blue.

Introduction

Since its first introduction in 1943, Dapsone (DDS-diaminodiphenylsulphone) has been effectively used for leprosy and for treatment of many other diseases.[1,2] Dapsone was traditionally used as an anti-leprosy drug and later, its use has been expanded to conditions such as acne vulgaris, dermatitis herpetiformis, thrombocytopenic purpura, and pemphigoid. With the increasing use of dapsone for various indications, acute poisoning (either accidental ingestion or suicidal) is increasingly recognized. [1,3] Methemoglobinemia, is an important and potentially life-threatening manifestation of dapsone poisoning. [1,3] Here, we report a case of methemoglobinemia secondary to dapsone poisoning. Since the incidence of dapsone-induced methemoglobinemia is rare, our aim here is to enhance awareness about this life-threatening adverse event.

Case history

A twenty year old female from Kormmathural, Batticaloa presented to the emergency room with alleged history of

consumption of about 100 tablets of 50 mg Dapsone (about five hours after ingestion). This drug was used by her mother for Immune thrombocytopenia. (ITP) She was complaining of severe headache, dizziness nausea and vomiting several episodes after consumption of tablets. There were no significant past medical history. On examination there was central as well as peripheral cyanosis. She was dyspnoic, respiratory rate was 22 breaths per minute and an oxygen saturation of 85% on air, temperature was 98.1°F, pulse was 130 beats per minute, her blood pressure was 130/80 mm Hg. On auscultation of chest there was bilaterally equal air entry.

Her ABG showed respiratory alkalosis with metabolic acidosis with pH 7.51, PCO₂ 22, PO₂ 88, HCO₃⁻ 15.3 & FiO₂ 112 (with O₂). There was a discrepancy between saturation and ABG analysis. Hemoglobin was 11.6 g/dl, total count was 10.2 per mm³ and platelet count was 144,000 per mm³. Renal functions were normal.

Liver function test showed elevated AST 19, ALT 23, Total Bilirubin 61.5 μmol/l with high indirect Bilirubin 51.9 μmol/l and LDH of 958 IU/L. At the time of admission the Bilirubin levels were within normal range (39.1 μmol/l), it continued to increase initially (highest 61.5 μmol/l). Blood picture showed normochromic normocytic anemia with bite and blister cells and mild

thrombocytopenia, favouring dapsone induced haemolysis. Methaemoglobin level was not done but by clinical evidence suggested it was to be more than 30%.

Immediately she was managed with prop up, high flow O2 via mask. Apart from the supportive measures, the patient received injection Methylene blue (1% solution) 2 mg/kg, in three divided doses for one week. Ascorbic acid 1000 mg three times daily was also given. Activated charcoal has been given every 6 hourly four 48 hours to prevent entero-hepatic reabsorption of dapsone. Even though with all these measures the patient clinically not improved, she was dyspnoic and cyanosed with saturation 87%. Exchange transfusion was arranged and finally she gradually improved after 10 days.

Discussion

Methemoglobin is an aberrant form of hemoglobin in which the ferrous (Fe²⁺) atom is oxidized to a ferric (Fe³⁺) atom resulting in an increase in its oxygen affinity but a decrease in its oxygen binding capacity.[6] Increase in the concentration of methemoglobin in the blood (>45%) causes dizziness, fatigue, headache, tachycardia, and weakness.[2] Further increase in concentration causes acidosis, cardiac arrhythmia, dyspnea, seizures, and eventually it can lead to coma if the methemoglobin level approaches 70%.[2]

Dapsone is absorbed through the gut, undergoes entero-hepatic circulation and is metabolized by the liver through the oxidation reactions of N-acetylation and N-hydroxylation. [2,6,7] Hydroxylated amine metabolites produced in the oxidation reactions is recognized as potent oxidants and has been hypothesized to cause dapsone's adverse effects, including hemolytic anemia and potentially life threatening methemoglobinemia.[2] Arterial blood gas analysis paired with oxygen saturation is the mainstay for making a correct diagnosis of methemoglobinemia (normal to elevated levels of PaO₂ with low oxyhemoglobin saturation).[2] Peripheral and central cyanosis becomes evident when there is a minimum methemoglobin level of 15% in the blood.[7] Although in present case we did not perform, the quantification of MethHb which can be performed by spectrophotometer is highly sensitive.[8]

The mainstay of treatment is supportive measures including ventilatory support. [2] Methylene blue is oxidized into leuco-methylene blue and acts as an artificial electron acceptor to methemoglobin, resulting in methemoglobin's conversion back to hemoglobin. [7] When there are symptoms of dyspnea or a methemoglobin level of at least 30% patients should receive methylene blue intravenously at 1 to 2 mg per kilogram of body weight over a 5- minute period. The reduction of MethHb by methylene blue is dependent upon NADPH generated by G6PD. G6PD converts glucose-6-phosphate to 6-phosphogluconate, generating 1 mole of NADPH, as the initial step in the hexose monophosphate shunt. The failure of this reaction to occur would

be rate limiting in the production of NADPH and in the reduction of methylene blue. G6PD-deficient individuals do not generate enough NADPH to efficiently reduce methylene blue to leukomethylene blue, which is needed for activation of the NADPH-dependent MethHb reductase system. As a result, methylene blue may not only be ineffective but also potentially dangerous since it has got oxidant potential that induces hemolysis in G6PD deficient patients. Therefore, methylene blue is not the ideal mode of treatment of methemoglobinemia in G6PD-deficiency as it can worsen the condition of the patient by increasing hemolysis.[9]

Table 1: Clinical manifestations of methemoglobinemia*

MethHb level (%)	Signs and symptoms
< 3	none
3 - 15	Pale, gray or blue color of the skin
15 - 30	Cyanosis, chocolate-brown color
30 - 50	Breathlessness, headache, dizziness, syncopal attacks
50 - 70	CNS depression, coma
> 70	Death

*Adapted from Goldfrank's Toxicologic Emergencies. MethHb=Methemoglobin, CNS=Central nervous system

It was emphasized in literature that multiple dose activated charcoal increased the elimination of dapsone. Repetitive charcoal is recommended to interrupt the entero-hepatic circulation of dapsone. In our case, the agent responsible for toxicity was taken in a high dose and underwent entero-hepatic circulation; therefore, the cure could be provided with repetitive methylene blue administrations.[10]

Ascorbic acid is an effective alternative for treatment, if methylene blue is not available. It is a strong reducing agent that takes part in various oxidative-reductive reactions. It is proved to be effective in treating MethHb when given at the dose of 300 mg/kg IV. In our case, ascorbic acid is given as 1000 mg thrice daily from day 2. [10] Exchange transfusion & hyperbaric oxygen may also be considered for severe refractory cases like our patient.

Conclusion

Dapsone-induced methemoglobinemia is a rare, but life-threatening adverse event. A key to the diagnosis of methemoglobinemia is cyanosis with low SPO₂ and normal PO₂ on arterial blood gas analysis. Treatment should be initiated immediately with IV methylene blue that converts MethHb to normal hemoglobin, thus increasing the oxygen-carrying capacity of erythrocytes. Prompt diagnosis and treatment is lifesaving.

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Advanced Carcinoma of penis - A case report

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Abstract :

Carcinoma of penis is an uncommon tumour but significantly higher incidence in under developed countries. Unfortunately delay in diagnosis may lead to advanced disease and cause management issues. Most tumours of the penis are low grade Tumours. The surgery is the gold standard method in the penile cancer although chemo-radiation has yield local control rates. The scope of surgery varies from wide excision to total penectomy.

Keywords

Carcinoma of penis, Lymph nodes, Amputation of penis

Introduction

Carcinoma of penis is an uncommon tumour but significantly higher incidence in under developed countries (1). Unfortunately delay in diagnosis may lead to advanced disease and cause management issues. Most tumours of the penis are low grade Tumours (1). The primary treatment of penile cancer depends on the size, location, extent of disease and Lymph node involvement. The surgery is the gold standard method in the penile cancer although chemo-radiation has yield local control rates (1,2,3). The scope of surgery varies from wide excision to total penectomy. Circumcision performed for prepuce restricted lesions. The successful local control achieved by partial penectomy depends on division of penis at least 2cm proximal to gross tumour extent.

Case history

67 year old man admitted to the casualty ward with difficulty in passing urine for two weeks duration and Growth at the Glans Penis for seven months durations. The growth increased in size rapidly over eight month period. He also noticed urine leaking from the root of the penis one month back.

He denied any extramarital sexual contact in the past. There was no history of urethral discharge. He didn't have tight prepuce or any changes in the prepuce region. He has no comorbid

disease in the past. Examination of External Genitalia revealed fungating friable cauliflower growth of penis extending to the shaft of the penis. The urethral meatus was obliterated. The whole shaft became indurated and hard. There were two water cane fistulas one at the root of the penis and other one is at scrotal region. There were palpable firm solid inguinal nodes measuring 1.5cm-2.5cm in both inguinal region. Abdomen and systemic examination findings were normal. The supra pubic catheter was introduced for the urinary diversion.

The biopsy and FNAC obtained from Glans penis and Inguinal Lymph nodes respectively, revealed squamous cell carcinoma. The ultrasonic examination revealed normal findings. The Contrast C.T was planned due to lack of M.R.I. facilities in this institution. The contrast C.T revealed large soft tissue mass lesion involving the entire penis and mass extending into the left scrotal wall but root of the penile region appear normal with normal CT abdomen and no significant lymph node enlargement.



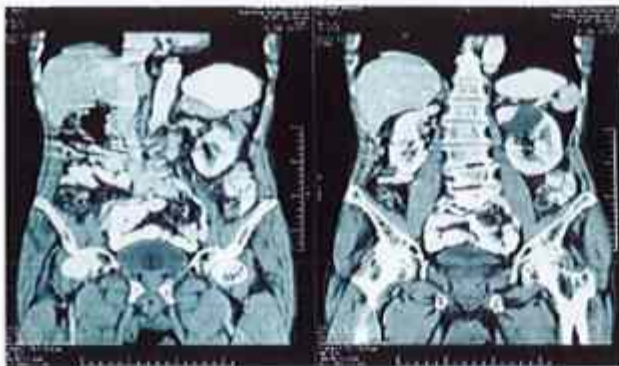


Fig. 1 & 2 CECT of external genitalia, abdomen and pelvis

As per the decision taken during M.D.T meeting concluded the management option of this patient was Neoadjuvant chemo radiation followed by total amputation of penis, bilateral orchiectomy, perineal urethrostomy and bilateral Lymph node dissection. The postoperative period was uneventful and supra pubic catheter was removed after two weeks from the day of surgery. The whole specimen histopathology report revealed squamous cell carcinoma. The patient was then transferred to oncology unit for further treatment.

Discussion

The most common way of presentation of penile carcinoma is as Lump or ulcerated growth (82%) (1),(3). The other presentation include Erythematous lesion (17%), phimosis, bleeding, Haematuria, per urethral discharge and dysuria. 20% of case with penile carcinoma have locally advanced disease with Inguinal Nodal metastatic disease at the time of presentation (1,2,4) This patient present with firm palpable bilateral Inguinal Lymph nodes. The remaining cases with lymph node Enlargement is caused by inflammation or more often secondary infection of primary tumour (1,2,4). The diagnosis is usually made by biopsy and histopathology.

The imaging studies include ultrasonography, computerized Tomography, Position Emission Tomography and Magnetic Resonance Investigate the Loco regional lymph node involvement and extent of the disease (1,3,4).

FNAC Showed 93% sensitivity and 91% specificity with a false negative rate of 20% to 30% (1). The CT plays a role in staging of the disease. Recently SPECT-CT imaging has been used to accurately investigate the patients with clinically Lymph node negative penile cancer (1). PET/CT is considered as such the current body of evidence does not support a role of stand along diagnostic procedure in penile cancer until larger trials need for further information (3,4). Magnetic Resonance imaging in small series of study to detect Lymph Node metastasis in penile cancer has shown 100% sensitivity and 97% specificity for detecting occult Lymph node metastases (1).

The management of penile cancer depends on the site, extent, loco regional nodal state and metastasis of the disease. Treatment of the primary penile cancer lesion aim is to remove

the tumour completely depend on the site and extent of the lesion. The options available are circumcision, penile preservation (glansectomy or partial Amputation) and total penectomy. The intra operative of surgical margins by frozen section is recommended (2). This patient underwent total penectomy because of the extensive lesion. The penis conservative surgery improves the quality of life but local recurrence was 8.9% with a five year disease specific survival rate of 91.7% (2). Any local recurrence rate is much low in radical surgery (2). The radiation treatment of primary tumour (T1- T2 lesion <4cm diameter) has a good results in selected patients.

The inguinal nodes negative patients by FNAC or diagnostic sentinel node biopsy need close follow-up (2). Node specific cases of inguinal lymph nodes need radical inguinal lymph node dissection (ILND) which is curative in approximately 80% of men with limited to one or two metastatic inguinal nodes (5 year disease specific survival rate - 75%). This patient also underwent for bilateral radical ILND for node positivity. The current guideline recommends the men with two or more positive pelvic lymph node should undergo for PLND according to the side (2,3). It has not been reported that neo-adjuvant or adjuvant radiotherapy improve the oncological outcome in node positive penile cancer (2,3). Palliative chemo radiation in advanced cases or relapsed disease is considered (2).

Conclusion

The locations of penile cancerous lesion contribute the tradition surgical intervention of penile cancer. It has severe impact on patient psychosocial domains, quality of life and functional aspect. The advent new approaches such as DSLNB and intra operative frozen section improve the quality of life and better prognosis in these patients.

Learning points

Presentation of carcinoma of the Penis with a locally advanced state is not uncommon. Thus all these patients need radiological and histological evaluation.

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Rapid envenomation following a saw-scaled viper (*Echis carinatus*) bite into a superficial vein causing intracerebral hemorrhage

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Abstract :

Saw Scaled Viper (SSV) is a venomous snake mostly found in the dry zone and coastal areas of Sri Lanka. It is often described as aggressive and irritable. [1] SSV envenom is characterized by coagulopathy with minor bleeding manifestations. [1,2] The SSV sub species in Sri Lanka is rarely known to cause life threatening events. [3,4,5] But in our case, A 33 year old priest came with an SSV bite which had directly injected venom into a superficial vein. The patient rapidly developed Intra cerebral hemorrhage (ICH), Myocardial Infarction (STEMI), venom induced consumptive coagulopathy (VICC) which finally led to multi organ failure.

Introduction

Of the 93 species of land and sea snakes in Sri Lanka, five of the land snakes and almost all of the sea snakes are considered potentially deadly. Saw Scaled Viper (SSV) is considered as one of the potentially deadly snakes. [6]

A venomous snake can inject venom into subcutaneous tissues, muscle, a vein, or an artery. Lymphatic circulation collects the venom and draws it into the systemic blood flow. Direct vascular puncture results in rapid onset of systemic toxicity and it is difficult to manage, even when antivenom is readily available. No first aid measure, other than supportive care and advanced life support, would be of benefit in intravascular venom injections. [7]

Symptoms and signs will vary according to the type of snake responsible for bite and the amount of venom injected. The patient in this report rapidly developed Venom-induced consumption coagulopathy (VICC) which was not reverted with repeated administration of Anti-snake venom serum (AVS) and plasma products including coagulant factors and Fresh Frozen Plasma. The rapid deterioration of the clinical condition was due to a direct venous injection of the venom. The cerebral hemorrhage may be caused by VICC and hemorrhage is due to direct endothelial damage. [8]

In Sri Lanka, the most number of deaths are from bites of Russell's viper, Cobra and Krait while SSV systemic envenomation has rarely reported to cause fatal hemorrhagic manifestations. [4] This case report brings awareness of fatal complications such as ICH which can occur with Sri Lankan SSV.

Case History

32 year old priest came to the hospital around 7.30 a.m with a history of a snake bite on the right wrist; there was fang marks and but no local swelling. He was fully conscious upon admission, giving a history of a bite by a snake hanging on a light post around 2 meters in height, on a paddy field. He described the physical appearances of a Saw-scaled viper (SSV).

At 8.10 a.m. nearly 70 minutes post bite he suddenly developed a right sided focal fit with tonic-clonic movements with rolled-up eyes, froth from the mouth and urinary incontinence. Twenty minutes whole blood clotting time was prolonged and the working diagnosis was VICC. After that the blood pressure was un-recordable, conscious level was lost, there was a feeble pulse and also low oxygen saturation. Cardio-pulmonary resuscitation was able to restore the circulation but the conscious level and respiratory movement was not improved. During the

endo-tracheal intubation, moderate amount of blood clots in the oral cavity and air way was noted. After a while, he developed a ventricular tachycardia (VT) with an undetectable pulse. Ventricular tachycardia was reverted with an electric shock. At this time the ECG showed ST elevation in the inferior leads.

10 vials of Anti-snake venom (ASV) were given with the cover of intravenous Hydrocortisone, intravenous Chlorpheniramine and subcutaneous 1:1000, 0.25 ml Adrenaline. Computer Tomography of the brain showed an intracerebral hemorrhage, while the chest x-ray detected a right-sided lung haemorrhage. Prothrombin time/International Normalized Ratio was 6.0. Second episode of VT was again managed with electric shock. Later, he developed a generalized tonic clonic seizure. Intravenous Phenytoin was able to control the seizure. Endo-tracheal tube bleeding, rectal bleeding and hematuria were developed later. Patient then developed VICC. Patient was transfused with Fresh Frozen Plasma, Cryo-Precipitate, platelets and factor VII. Four cycles of Anti snake venom serum was also given.

Patient needed inotropic support on the second day where he developed multi organ failure. Renal function also deteriorated without a urine output. Serum creatinine was 10.6 mg/dl. Patient was put on Continuous Renal Replacement Therapy (CRRT). Unfortunately, the patient passed on the 4th day.

Discussion

In this presentation, a 34 year old priest came to the hospital early in the morning with a snake bite on the right radial region of the wrist. Snake bite had happened in a paddy field around 7 a.m. Saw Scaled Viper (SSV) and Russel's viper are two venomous snakes favouring the incident because those two generally inhabit vegetation and paddy fields. [1, 9]

The patient was switching off the lights on a light post around 2 meters in height from the ground. No one saw the snake other than him and he did not bring the dead snake to the hospital. The incident happened on December 2018 in the rainy season in the early morning. SSV is mostly active in the rainy season, in a humid environment in early mornings and early nights. They can climb the bushes, shrubs and walls around 2 meters in height from the ground. [10,11] But the Russel's viper bite is the other way around, as they are active in the dry zone and paddy cultivated area and are not known to climb heights. [12,13]

Fang marks on the right wrist on a superficial vein was observed that resulted direct puncture of the vein in the right wrist which caused rapid systematic envenomation. It caused venom induced consumption coagulopathy. Saw scaled viper venom contains proteinous and non proteinous toxins which interfere with coagulation cascade leading to VICC. [14,15] SSV sub-spices in India and other parts of Asia, causing intra cranial hemorrhage and coagulopathy are commoner than Russel's viper but very rarely reported in Sri Lanka. [3,6] With VICC, development of ICH, lung alveolar hemorrhage, hypotension, multi organ failure and cardiac arrest are possible. Intracranial hemorrhage may be

the reason for ST elevation in the ECG because cardio toxicity of SSV is not known before. CPR may have resulted in the development of ventricular tachycardia as well. Due to VICC, patient developed multi organ failure, cardiac, renal, liver functions also deteriorated. Cardio toxicity favours Russels viper bite in this case because

- i. In Sri Lanka, significant mortality developing following an SSV bite is rare.
- ii. Russels viper bite has been known to cause direct venom effect of the heart which can lead to arrhythmias.[16] ST elevation MI due to coronary spams and infarction with severe vasospasms with toxins on endothelium and sarfatoxins.[17, 18]
- iii. Russels viper bite had been reported to cause MI in patients who already has atherosclerosis which may be worsened by vaso spasm due to Russel's viper bite.

In Sri Lanka, ST elevation MI following Russel's viper bite was reported, but no cases of any significant cardiotoxin due to SSV bite have been reported before elsewhere as well. [19, 20] But cardio toxicity following direct vessel injection of SSV venom might be of a hypothetical value to be tested at laboratory level.

Conclusion

SSV sub spices in Sri Lanka causing life threatening hemorrhage has not been previously reported although it happened in the other parts of Asia. [3,6] Through this case report, we would like to alarm the possibility of high risk of death with SSV envenomation and this is a rarest occasion where venom of a snake directly punctured into a vein. [21,22]

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Spigelian Hernia: A rare ventral Hernia

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Abstract :

Spigelian hernia is a rare ventral abdominal wall hernia occurring through the fascial defect lateral to the rectus abdominis muscle. The diagnosis is made thorough clinical examination with radiological confirmations. The operative repair is the treatment of choice due to the risk of incarceration and strangulation. Here we report this rare case of spigelian hernia in a 14year old boy presented with right lower abdominal pain.

Introduction

Spigelian hernias are uncommon and constitute 1-2% of all abdominal wall hernias (1). They occur through the fascia along the semilunar line, which corresponds to lateral border of rectus abdominis muscle. Most of these occur below the level of umbilicus. The hernia sac is usually interparietal and passes through the defect in the internal oblique and transverse abdominis muscles and lies deep to the intact external oblique aponeurosis. They can be repaired either open or laparoscopic techniques.

Case history

A 14 year school boy presented with 5 weeks history of repeated episodes of right lower quadrant abdominal pain associated with a visible bulge in the right lower abdomen in several occasions.



Figure 1: Right lower abdominal bulging at linea semilunaris

He denies any history of bowel or bladder alteration. No associated history of fever, nausea, vomiting or abdominal distension. The swelling appears on coughing or straining and subsides on rest. Physical examinations were within normal limits except for the presence of a bulge while on coughing at the lateral border of rectus muscle in right lower abdomen.

A preoperative diagnosis of spigelian hernia was made on the basis of clinical examination. After an extensive discussion, parents gave the consent for open hernia repair. The hernia was explored under general anesthesia with muscle relaxants. An oblique right lower quadrant skin incision was made, passing through the linea semilunaris over the bulging area. The superficial dissection was done with diathermy and external oblique aponeurosis was split along the fibers. The defect and sac were lateral to the lateral border of rectus muscle. The hernia sac contained greater omentum and covered by pre peritoneal fatty layer.



Figure 2: The hernial sac contains greater omentum.

The defect measuring 3cm in length was identified and repaired. Polypropylene mesh repair was done in sublay technique. Anterior rectus sheath and skin were closed with absorbable sutures. Post operatively patient had an uneventful recovery. He was discharged in post operative day 1.

Discussion

Spigelian hernia is a rare group of hernia occurs through the defect in the anterior abdominal wall at the lateral border of rectus muscle. The fascia lateral to the rectus muscle often called as spigelian fascia. The widest part of this fascia, called spigelian hernia belt, where 85 - 90% of the hernia occur and lies between 0 - 6cm cephalad to the interspinous plane (2). The herniation is very rare above the level of umbilicus because semilunar line is supported anteriorly very strongly by external oblique aponeurosis and posteriorly by muscular fibers of transverse abdominis. The contents of the hernia is commonly greater omentum. Rare contents of the sac include small intestine, part of colon, appendix or ovary (2).

The peak incidence is between 4th and 7th decade of life and occurs commonly on the right side. The patients usually present with abdominal pain, intermittent swelling and features of intestinal obstruction. Preoperative clinical diagnosis most of the time is difficult due to abdominal obesity and vague symptoms.

Plain abdominal X-ray is not helpful unless those who presenting with the features of intestinal obstruction. Ultrasound abdomen is the first line of investigation. It has sensitivity and positive predictive value of 90% and 100% respectively (3). CT scan with contrast is the most reliable investigation for difficult cases.

Surgical repair is the treatment of choice to prevent the subsequent development of complications. It can be repaired open technique or laparoscopically depend on the available facilities and surgical experience.

Conclusion

Spigelian hernias are uncommon and clinical diagnosis needs a high index of suspicion. Strangulation and incarceration are the complications and occur in about a quarter. Radiological investigation, especially CT scan helps for the equivocal cases. Surgery is the choice of treatment and open approach is often practiced where facilities and expertise for laparoscopy are not available. Tension free repair with synthetic mesh is recommended.

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A Case of recurrent Multicentric pleomorphic adenoma

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Abstract:

Pleomorphic adenoma is the most common salivary gland tumour and accounts for high rate of recurrence. It is a tumour of variable capsulation characterized microscopically by architectural rather than cellular pleomorphism. Mixed tumours of the parotid gland are rare in childhood and recurrence of this tumour is infrequent. This case report is about a recurrent neoplasm of parotid in which biopsy revealed a recurrent multicentric pleomorphic adenoma.

Introduction

Pleomorphic adenoma is the most common salivary gland tumour and accounts for high rate of recurrence(4). It is a tumour of variable capsulation characterized microscopically by architectural rather than cellular pleomorphism. Epithelial and modified myoepithelial elements intermingle most commonly with tissue of mucoid, myxoid or chondroid appearance. Mixed tumours of the parotid gland are rare in childhood and recurrence of this tumour is infrequent. High rate of recurrence with some histological subtypes such as hypocellular variant had been reported in literature (1).Female sex and young age at initial treatment are also risk factors(1). Also the first surgical treatment, tumour enucleation or parotidectomy has been implicated as a cause of recurrence(1) This case report is about a recurrent neoplasm of parotid in which biopsy revealed a recurrent multicentric pleomorphic adenoma.

Case history

A 25 year old girl presented to our surgical clinic with a history of right sided recurrent parotid lump for 1 year duration. She had already undergone excision of parotid lump twice in our unit 10 and 7 years before this presentation. The specimen of superficial parotidectomy revealed pleomorphic adenoma in

both occasions. According these reports in both occasions the dissection margins were difficult to be commented on. She was subsequently followed up in our clinic.

There was no pain, discomfort, malaise, facial asymmetry or any history of trauma, and her past medical history was unremarkable. Examination revealed an irregular lobulated lump, roughly measuring about 3*2 cm and the facial nerve was not involved clinically.

After discussion with consultant radiologist, contrast enhanced CT was done. It's revealed non enhancing multiple nodular lesions seen in right parotid gland. It was negative for any associated Lymph node enlargement. An excision biopsy was recommended.

She underwent superficial parotidectomy with preservation of facial nerve. The neoplasm was found to be involving in the tail of the parotid gland. The mass was located in subcutaneous layer and surrounded by fibrous tissue. Multiple nodular lesions were also seen in the superficial part of the parotid gland and the tumor was completely excised with margins of normal glandular tissue. No macroscopically infiltrative nature of the tumor was noted during the surgery.

Histological assessment

Macroscopically the outer surface of the tumor appeared to be irregular and whitish. Cut sections revealed multiple nodular appearances in the gland.

Microscopically the sections of recurrent parotid tumour revealed a multinodular lesion widely dispersed in adipose tissue and within parotid salivary tissue. All the nodules were well defined and surrounded by a thin fibrous capsule. These composed of cords and ductules of myoepithelial and duct epithelial cells, melted in chondromyxoid stroma. There was no cellular atypia, mitotic activity or necrosis. The features were compatible with recurrent multifocal pleomorphic adenoma. The same lesions were present at the tumour margins. There were no features to suggest malignant transformation. Four adjacent reactive lymph nodes were present.

Discussion

This is an example of a recurrent multifocal parotid pleomorphic adenoma that recurred years after the initial surgery. Recurrence is usually due to inadequate clearance of the tumour during resection. This depends on the surgical skill, extend of the tumour in relation to extracranial course of facial nerve and infiltration of tumour to adjacent tissue. Multifocal nature of the tumour challenges the adequate clearance during the surgery as in this patient even though this entity is rare. The presence of pseudopodes and multinodularity are the histological factors associated with recurrences in pleomorphic adenoma(1). In this case the dissection margins were not commented in the previous histopathological reports. There are two possibilities for recurrence in this patient. Lesions could be arising independently by multifocal pathogenesis (primary multifocal pleomorphic adenoma). Or, the tumour nodules could be parasitic nodules that have become detached from a main nodule. Pleomorphic adenomas can be multifocal, particularly when they recur after surgery or when there has been trauma. Small nodular protrusions extending from the tumour surface or tumor spillage and seeding at the time of surgery are presumable responsible for these tumour recurrences (2). Primary multifocal pleomorphic adenomas have been reported in literature. This is a highly unusual condition and little is known about the etiology and pathogenesis and clonal relationship has only been studied only in few patients (3). According to the final histopathological report the primary multifocal pleomorphic adenoma is the highest possibility in this patient even though it's a rare subtype.

Considering the treatment modality simple enucleation of pleomorphic adenoma is associated with high recurrence rates,

between 8% and 45%, which is reduced to less than 5% with superficial parotidectomy and further down to 0.4% with total parotidectomy (4). For this reason, superficial parotidectomy or total parotidectomy is generally preferred over enucleation. Recurrent tumors usually appear near the surgical scar from previous resection. Recurrence rates do not vary according to age or gender. One strategy to reduce the rate of recurrence is to re-evaluate surgical margins using intraoperative frozen sections which is not available in our unit.

Radiation therapy may be an option for postoperative treatment of recurrent pleomorphic adenoma, especially for cases in which multinodular masses were present or multiple recurrences have occurred (4). The facial nerves must be sacrificed, or complete excision is practically difficult(4).

Conclusion

The key point to learn in this patient is that even though recurrence of parotid pleomorphic adenoma is commonly due to seeding during surgery, primary multifocal pleomorphic adenoma may also rarely contribute to recurrence of tumour.

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High level of clinical suspicion need in eczematous rash of areola and nipple for early detection of Paget's disease of the breast. - A case report

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Abstract :

Paget's disease is a rare presentation of breast carcinoma. It's identified as a scaly, itchy, erythematous patch in areolar and nipple. The patients presented with above mentioned lesions of nipple are frequently misdiagnosed as dermatitis or eczema. High level of clinical suspicion and biopsy from lesion of nipple will help to arrive at the diagnosis.

Keywords

Paget's disease, Mastectomy

Introduction

Paget's disease is a rare presentation of breast carcinoma. It's identified as a scaly, itchy, erythematous patch in areolar and nipple (1). The patients presented with above mentioned lesions of nipple are frequently misdiagnosed as dermatitis or eczema (2, 4). High level of clinical suspicion and biopsy from lesion of nipple will help to arrive at the diagnosis.

Case history

47 year married woman presented with itchy, scaly and cracked nipple of breast for three months. She didn't notice any lumps in her both breast. There was no family history of breast cancer, ovarian cancer, colorectal cancer or prostatic carcinoma. She had three children and had breast fed all her children. She was not on oral contraceptives or exposed to radiation in the past. She has no other co-morbid disease and was not on any medication. There was a 1.5cm round, scaly, crusty and cracked lesion in her right nipple. There was no erythematous appearance or ulcer in the nipple or areolar. There was no palpable mass or lesion in both breast, both axilla and neck. The general and systemic examinations were within normal limit. Ultrasonography of breast, Axilla, Neck and abdomen revealed no significant pathology.

The biopsy of the nipple suggested most probably Paget's disease and advised for re-biopsy. Mammography didn't reveal any significant lesion, M.R.I was performed and revealed thickening of right nipple and adjacent skin of peri-areolar region of the right breast compatible with known inflammation. The re-biopsy from nipple and areolar revealed carcinoma in-situ with Paget's disease.

After further discussion with M.D.T and the patient, the simple mastectomy option was given to the patient. The patient elected to proceed with right side simple mastectomy. The simple mastectomy was performed and the patient recovered well after surgery. (The ultrasonography, mammogram and M.R.I didn't reveal any significant nodal enlargement in axilla or neck and the biopsy revealed carcinoma in-situ.

Final surgical pathology of right Mastectomy specimen revealed Paget disease with solid type in-situ component and there is no evidence of residual Paget disease or in-situ carcinoma. The patient was referred for oncology follow-up and confirmed to have no evidence of recurrent or metastasis.

Discussion

Paget disease of breast is a rare malignant lesion of breast. The incidence among all primary breast cancer is about 3% (1,4). The underlying lesion is identified in majority of Paget disease (60-65%) (4) but no underlying lesion is identified in 30-33% of Paget disease of breast according to studies (1,4,6). It's

common in elderly and extremely uncommon in young women. The age group varies from 24 year to 84 year with a mean age of 55 year (5). Our patient's age is 47 year. The classical clinical problem is that Paget disease of breast can be misdiagnosed as eczema or dermatitis. This delayed the diagnosis in this patient. The prognosis of Paget's disease of breast depends on

1. Presence of invasive cancer
2. Presence of positive lymph node (1,2,5)

If there is no underlying breast malignancy or lymph node spread the survival rate is 92-94% (1,2,5). Our patient presented with a scaly cracked lesion of nipple and areolar but not a classical presentation of Paget disease of breast. She has no palpable mass or lesion on physical examination.

This case demonstrates the importance of maintaining high level of suspicion and histological examination even in unusual clinical pictures.

Conclusion

Early and accurate diagnosis is an important factor in Paget's disease of breast. Hence early accurate diagnosis will help the patient and enable organ preserving surgery especially when the lesion is confined to the dermis of nipple. The early diagnosis also plays a major role in the diagnosis. (Liu et al., 2015)

Learning points

1. Paget's disease is a rare presentation of Breast Carcinoma
2. A scaly, itchy erythematous patch in the areola should always warrant a biopsy
3. Multidisciplinary approach is necessary for further management especially in patients with no other features suggestive of malignancy.

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Lupus Nephritis Patient Presents With Convulsion Due To Hypomagnesemia Secondary To Renal Loss

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Abstract :

Convulsion in a systemic lupus erythematosus (SLE) patient is multifactorial and finding the cause is always a challenge for a clinician. Electrolyte imbalance is not promptly thought as a cause in these patients. This is a case report of a patient with lupus nephritis which is a type of inflammatory condition of kidneys in SLE, presents with convulsion due to hypomagnesaemia secondary to renal tubular wasting.

Introduction

Seizures are one of the most serious neuro-psychiatric manifestations of SLE. It can occur at any time in the course of SLE and even before the diagnosis of SLE has been made or other manifestations of the disease are present[1]. Seizures are generally well controlled with the use of anticonvulsants but it has negative impact on the overall long-term prognosis and quality of life of patients with SLE. Factors that are associated with shorter time to the first seizure in SLE patient include African-American ethnicity, younger age, disease activity, damage early in the course of the disease, renal involvement, particularly WHO class IV glomerulonephritis, psychosis, positive antiphospholipid antibodies as well as the treatment with glucocorticoids and cyclophosphamide. But hydroxychloroquine use delays the time to first seizure occurrence[2]. Hypomagnesemia is an uncommon cause of seizure which can be overlooked in the emergency department. Hypomagnesemia can be due to genetic disorder or secondary causes such as gastrointestinal absorption defect or excessive renal wasting. Here we report a case of a 37 year old woman with WHO class IV lupus nephritis who presents with generalized tonic clonic type seizure secondary to hypomagnesemia as a result of excessive renal wasting of magnesium.

Case history

A 37 year old woman who was diagnosed three months back to have class IV lupus nephritis and diabetes mellitus presented to emergency department with generalized tonic clonic type seizure which lasted for 15 minutes. She was lethargic on the day of admission but she had no history of fever prior to admission. She was on high dose of steroid, mycophenolate mofetil and intermediate acting insulin. Her drug compliance was good. She didn't have arthralgia, joint swelling or skin rashes. She had no past history of convulsions. She had two uncomplicated pregnancies as well.

On examination she was drowsy but GCS was 15/15 with bilateral normal pupils. She was afebrile but mildly dehydrated. Tongue bite was noted but she didn't have features of meningism. Her neurological examination was normal. Skin rashes was not noted. Her blood pressure was 135/85 mmHg, pulse rate was 84 bpm, spO₂ was 99%. Other cardiovascular, respiratory system and abdominal examination was normal.

On admission capillary blood sugar was 279 mg/dl and arterial blood gas revealed compensated metabolic acidosis with pH of 7.40 and bicarbonate of 17.9 mmol/L. Laboratory investigations showed normal complete blood count, moderately elevated inflammatory markers (ESR -28, CRP-12). Non contrast CT-brain

as well as EEG were normal. Cerebrospinal fluid analysis was normal and culture was negative for infection. Blood culture was negative. Significant reduction in serum magnesium, phosphorus and moderate hypokalemia were noted. Serum magnesium was 1.0 mg/dl (NL 1.8-2.4), phosphorus 1.8 mg/dl (NL 2.5-4.9), potassium was 2.8 mmol/l (NL 3.5-5.0), sodium was 141 mmol/l, corrected calcium was 8.9 mg/dl (NL 8.4-10.2). Apart from low serum albumin of 2.9 g/dl (NL > 3.5) liver function test and renal function test were normal. ECG showed long QTc of 480 ms. Urine protein/creatinine was in microalbuminuria range. Urine was negative for ketone bodies.

Other investigations which were done in purpose of analyzing the cause for convulsions include negative antiphospholipid antibodies, normal C3, C4 level, urine fractional magnesium excretion which was 300% of normal, urine potassium/creatinine ratio of 2.5 meq/mmol (NL < 1.5), normal parathyroid hormone, normal creatinine phosphokinase level.

Discussion:

This is a case report about a patient with class IV lupus nephritis presenting with convulsion to the emergency department. Aetiology of convulsion in a SLE patient considered multifactorial. Antiphospholipid antibodies (IgM/IgG) and lupus anticoagulant have been described to convey an increased risk of neurological and thrombotic disorders[3][4]. This patient's non contrast CT-brain didn't show any features of thrombosis. Her antibody profile for antiphospholipid syndrome was also negative. Cerebral lupus was unlikely as this patient didn't have any clinical features to suggest SLE flare up. Normal serum C3, C4 levels and normal EEG are in favour of excluding cerebral lupus in our patient. Cerebrospinal fluid (CSF) for antineuronal nuclear antibodies which could have been done in this patient but this test has low sensitivity and specificity. This test has only some value in confirming cerebral lupus.

Central nervous system infection was excluded by absence of meningism, normal CSF report as well as negative CSF culture. Improvement of clinical condition without antibiotic treatment and normal EEG also in favour of excluding central nervous system infection as a cause for her convulsion. Cerebral insult secondary to hyperosmolar hyperglycemic state was unlikely because her random blood sugar was not too high on admission and cerebral oedema was not seen in the non contrast CT brain. Normal renal function tests and normal EEG in this patient exclude uremic encephalopathy as cause for convulsion.

Electrolyte imbalance is the most probable cause for convulsion in this patient. Seizures are especially common in patients with sodium disorders, hypocalcemia, and hypomagnesemia[5][6]. Unlike other electrolyte alterations, hypokalemia or hyperkalemia rarely causes symptoms in the CNS and seizures do not occur[5]. Magnesium and potassium were replaced intravenously during the initial days of admission but antiepileptics were not given. Repeated serum magnesium was

1.7 mg/dl which was significantly reaching the near normal level. But repeated potassium level showed only minimal rise by 0.1 mmol/l and phosphate level showed further drop by 0.2 mg/dl. Since hypomagnesemia is identified as the cause for convulsion in this patient magnesium replacement continued during the ward stay. Although primary cause for hypomagnesemia is gastrointestinal loss, in this patient renal wasting of magnesium due to tubular injury was identified as the cause. Immunosuppressant dose was increased in purpose of preventing further tubular damage due to lupus nephritis. Magnesium and potassium supplements were given and advised on diet rich in dairy products to maintain the phosphorus level. Patient didn't develop further seizures and levels of magnesium and potassium reached normal level after one month duration.

Conclusion

SLE patient can develop convulsion due to several causes. Though cerebral lupus, central nervous system infections and intracerebral vessel thrombosis are well known causes of convulsion in these patients, electrolytes imbalance need to be considered as well. Hypomagnesemia is one of the electrolyte abnormalities which cause convulsion, particularly in lupus nephritis patients due to renal tubular loss. We recommend prompt detection and correction of hypomagnesemia to control convulsion and optimization of immunosuppressants along with magnesium supplement to prevent further convulsions in these patients.

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Imported Malaria With Pancytopenia Caused By Plasmodium Vivax

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Abstract :

Malaria is an infectious disease caused by Plasmodium parasites, mainly *P. vivax*, *P. falciparum*, *P. ovalae*, *P. malariae* and *P. knowlesi* which are spread through Anopheles mosquitoes. Disease is not endemic in Sri Lanka currently, but can be re-introduced from other countries, mostly from India, Africa and Asia. The main sign of the disease is fever with shivers repeated periodically. There is highly effective chemoprophylaxis available and treatment, which should be given quickly. (1)

We describe a young man investigated for pyrexia of unknown origin with pancytopenia who had a history of visit to India 1 year back. He was diagnosed to plasmodium vivax infection with positive thin & thick blood film and rapid diagnostic test. As he had the visit to India 1 year back, malaria was not considered initially in diagnosis. But as patient developed typical tertian fever pattern it has given a clue towards the diagnosis. So as imported malaria cases were found in Sri Lanka recently, all the clinicians must have high index of suspicion for malaria in fever patient with travel history to endemic areas. Early detection and prompt treatment will prevent complications.

Key words

imported malaria, plasmodium vivax, tertian fever, pancytopenia, travel history,

Introduction

Malaria is an infectious disease caused by Plasmodium parasites, mainly *P. vivax*, *P. falciparum*, *P. ovalae*, *P. malariae* and *P. knowlesi*. The mosquitoes of the genus Anopheles are the major vectors of infection, where the parasite undergoes a sexual stage of its life cycle. Malaria has been one of the most devastating diseases to have affected Sri Lankans in the past. During the long documented history of its occurrence in Sri Lanka, several major epidemics have been experienced, the deadliest of these being the epidemic of 1934-1935 during which several millions of individuals contracted the disease and approximately 80,000 of them died. During the past decade, the malaria situation of the country has dramatically changed, with no indigenous malaria cases being reported since October 2012. With progressively

increasing incidence of imported malaria cases in recent years, early diagnosis and treatment of such cases have become the highest priority for prevention of re-introduction. Most of these infections have been acquired in India, Pakistan, South East Asian and African countries.

The main symptom of malaria is periodic fever, however the widespread use of antimalarial prophylaxis and antibiotics in recent years caused that non-typical course of fever occurs increasingly. Malaria often causes nonspecific symptoms (e.g. vomiting, diarrhoea, dyspnoea, and muscle and joint pain). The course of disease caused by *P. falciparum* (tropical malaria) is usually different - the symptoms are more severe and, in addition, there are also signs of multiple organ failure such as hypoglycaemia, acidosis, jaundice, disseminated intravascular coagulation syndrome or acute respiratory distress syndrome (4). The chemoprophylaxis does not eliminate the risk of infection completely, but reduces it significantly, and if the disease develops, the course is milder and increases patient's chance to recover

(6). The diagnosis of malaria is based on epidemiological history, signs, symptoms and laboratory tests (malaria rapid test which detects Plasmodium antigens such as specific lactate dehydrogenase, blood microscopy for a thin smear or thick drop, PCR test). After the diagnosis has been established, causative treatment should be started. Currently, it is based on several antimalarials with different mechanisms of action.

Case Report

A 48-year-old gentlemen from Arayampathy with no significant past medical history, has been admitted to the hospital with the history of fever for 3 weeks duration. His fever was initially started as evening pyrexia, with sweating and no chills & rigors, associated with arthralgia, myalgia, & lethargy. He was treated as viral fever at medical ward and discharged. But his fever spikes were not settled; he developed frequent fever spikes with high grade fever with chills & rigors, headache, nausea & vomiting. He also had generalized body aches, tiredness & lethargy. He has significant loss of appetite & loss of weight. (Usual weight 51kg to 44 kg now). There is no history of rashes, altered bowel habit, urinary symptoms and no other significant system involvement.

He is an electrician, father of 3 children, worked in Saudi Arabia in a ship yard from 2014 - 2018 and also had a history of visit to India with family, last year June.

On admission, the patient's temperature 102°F developed typical tertian pattern (Tab. 1) of fever. He was significantly pale, dehydrated, and had tachycardia 120 bpm, Blood Pressure 80/50. Laboratory tests showed pancytopenia (Tab. 2), blood picture favored anemia of chronic disease with viral infection, liver and renal functions tests were normal and ultrasonically no organomegaly. Blood film for malraila parasite came as positive while we were treating the patient as pyrexia of unknown origin with i.v antibiotics for which the patient had poor response. Malaria examinations (the rapid immunochromatographic test which detects antigens of Plasmodium and thick drop blood test) allowed diagnosing an infection (Tab. 3).

Finally the diagnosis was made as uncomplicated plasmodium vivax malaria. Treatment was immediately started with course of chloroquine tablets total 25mg/kg for 3days followed by primaquine tablets (0.25mg/kg per day) for 14 days. Patient was symptoms free and blood film was negative for parasite following few days of treatment. Pancytopenia also improved within a week.

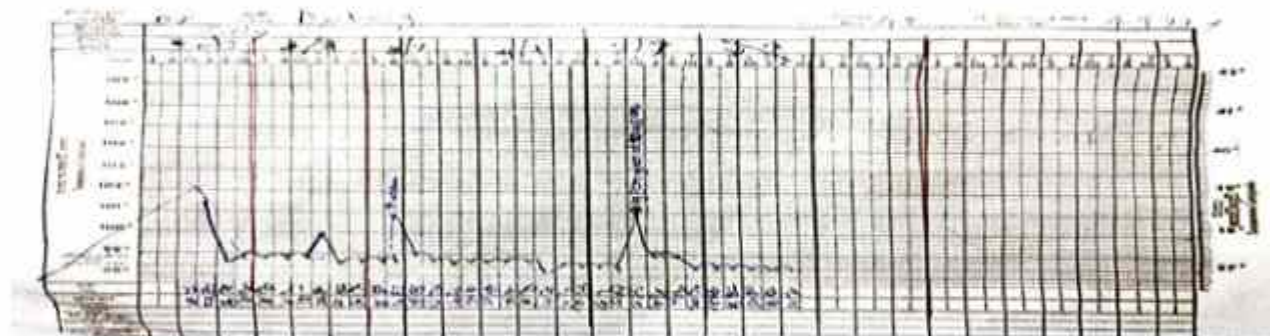


Table 1: tertian fever pattern

TEST	RESULT	UNITS	REFERENCE INTERVAL
WBC	3.48	10 ³ u/L	4 - 10
Hb	9.2	g/dl	11 - 15
Plt	79	10 ³ u/L	150 - 450
RBC	3.45	10 ⁶ u/L	3.5 - 5.0
CRP	121	mg/l	
ESR	123	mm/hr	
Parasite density	4039/ uil		
RDT	positive		

Table 2

Most of these infections have been acquired in India, Pakistan, South East Asian and African countries.

Currently, a low level of clinical suspicion in the backdrop of a very low disease burden has led to a significant delay in diagnosis of malaria cases. As a result, there were several patients who presented to the health care institutions with uncomplicated fever progressing to develop severe malaria while being at the hospital.

Like our patient there are so many patients initially treated as viral fever and discharged from the ward as they are having thrombocytopenia or pancytopenia which is considered as dengue fever. Thrombocytopenia has been a frequent finding among patients with malaria reported in the recent years, yet a diagnosis of malaria has not been considered as a result of them being misdiagnosed as having dengue. This had led to a delayed malaria diagnosis resulting in adverse sequelae.

Though our patient has history of visit to India 1 year back, malaria was not considered as a differential during his first

Discussion

With no indigenous malaria cases being reported since October 2012, Sri Lanka is currently in the malaria elimination and prevention of re-introduction phase. Recently with progressively increasing incidence of imported malaria cases in recent years, early diagnosis and treatment of such cases have become the highest priority for prevention of re-introduction.

hospital stay. Following the infective bite by Anopheles mosquito, the incubation period goes by before the first symptoms appear, which is usually from 7 to 30 days. The shorter periods are observed in *P.falciparum* and the longer ones with *P.malariae*. Antimalarial drugs taken for prophylaxis by travelers can delay the appearance of malaria symptoms by weeks or months, long after the traveler has left the malaria-endemic area. This can happen particularly with *P.vivax* and *P.ovale* both of which can produce dormant liver stage parasites; the liver stages may reactivate and cause diseases months after the infective mosquito bite. (7) Such long delays between exposure and development of symptoms can result in misdiagnosis or delayed diagnosis. So it's a good take home message that patient can develop Malarial infection even one year after the travel to endemic areas because of dormant liver forms of parasites.

Our patient had typical tertian fever & pancytopenia without any organ involvement. Early treatment prevented him developing severe malaria & serious complications. Therefore while investigating any patients with pyrexia of unknown origin malaria also should be considered & excluded to prevent serious complications and early treatment. Elimination of imported malaria will be mainly depending on early detection of cases, community awareness, mosquito control and chemoprophylaxis.

Conclusion

Malaria should be excluded in every patient returning from the endemic area who reports symptoms, even not directly indicating malaria, because immediate treatment significantly increases patient's chance to recover. It is necessary to educate

travelling patients constantly both by methods reducing the risk of mosquitoes bite, and correctly used chemoprophylaxis. High index of clinical suspicion should be there when managing fever patient with travel history to endemic area within 1 year. Early detection & prompt treatment will prevent patients developing severe malaria & serious complications.

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Chronic Uterine Inversion Secondary to Submucous Fibroid.

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Abstract :

Chronic uterine inversion is a rare gynaecological condition. It is associated with submucosal fibroid. Even though the clinical diagnosis is very challenging the diagnosis can be arrived with the aid of ultrasonography. The surgical management requires careful planning and multidisciplinary team work for optimum results.

Key words: Uterine inversion; Haultain's method

Introduction

Uterine inversion refers to a descent of the uterine fundus through the cervix, so that the uterus is turned inside out [1]. It can be acute or chronic. Acute uterine inversion causes severe pain and haemorrhage and almost always occurs in mismanaged third stage of the labour. The chronic uterine inversion is insidious and characterized by pelvic discomfort, vaginal discharge, irregular vaginal bleeding and anaemia [2]. However, chronic non - puerperal uterine inversion (CNPI) can be due to uterine neoplasm and endometrial polyp. Further, prolapsed fibroid is the most common condition associated with it [3]. CNPI is a very rare condition and on average a gynecologist may not see a case in their entire life.

Case history

A 35-year-old woman visited to gynaecology outpatient clinic with complaints of mass protruding per vaginam for 1½ years duration. She had one full-term normal delivery 4 years ago.

Her general examination was in satisfactory condition. Systemic examination was within normal limits. On per vaginal

examination, a large globular mass about the size of 10×13 cm, was felt projecting through the vaginal introitus. Uterus could not be felt separately from the mass.

Ultrasonography of the pelvis could not visualize the uterus in the pelvis which confirmed the clinical suspicion of uterine inversion. Pre operatively bilateral ureters were stented to avoid ureteric damage during surgery. At laparotomy a cup shaped depression was seen with pulling in of the adnexal structures (figure 1). An attempt was made to pull the uterus upwards with a volsellum while upward pressure was given from the vaginal side. As it was unsuccessful due to strong cervical ring, the posterior rim of the cup was incised through both the thickness of the inverted wall (Fig. 3). Still it was not possible to reduce the mass into the abdomen. Therefore, mass through the vagina was surgically removed. Then the entire uterus was replaced easily. There was a difficulty to separately identify the uterine fundus from the fibroid during the surgery.

The incision on the uterus was sutured with interrupted no 1-0 vicryl and haemostasis achieved. Her postoperative period was uneventful and patient was discharged on the 5th day. Patient was seen two months following the surgery and had no post-operative complications and her menstruation was resumed normally.



Fig. 1. Laparotomy view of the uterus before correction of inversion. Note the characteristic dimpling and in-turning of the Fallopian tubes, ovarian ligaments and round ligaments.

Discussion

The incidence of the CNPUI is uncertain. It is usually precipitated by tumors sited at the fundus of the uterus which exert traction force to cause the inversion, although some cases have been reported with no association with tumors. It affects mostly women with intrauterine tumour like submucosal leiomyoma while other causes are leiomyosarcoma, endometrial carcinoma, rhabdomyosarcoma, malignant mixed mullerian tumour and endometrial stromal carcinoma [4].

The uterine fibroid is the most common causes in 78.8%-85% of cases [5].

The major factors that contribute to its occurrence are: tumor attachment site, thickness of the tumor pedicle, tumor size, thin uterine wall and dilatation of the cervix [6].

The cases of non-puerperal uterine inversion commonly present after 45years of age. Only few cases of non-puerperal inversions have been reported in young women.

The diagnosis is easier with complete inversion when a bluish-red mass is identified from the vulva with a constricting ring of the cervix superiorly [7]

The clinical examination of chronic inversion could reveal a mass protruding through the vagina without definite margins of a cervix and the absence of the uterine body during bimanual or rectal examinations. Occasionally, ostia of the Fallopian tubes may be identifiable on its endometrial surface.

The ultrasonographic examination shows an indentation of the fundal area and a depressed longitudinal groove extending from the uterus to the center of the inverted uterus.[6]. Magnetic Resonance Imaging (MRI) demonstrates a "U" shaped uterine cavity, a thickened and in-verted uterine fundus on sagittal section, and a "bull's eye" configuration on an axial image. [6].

Many abdominal and vaginal approaches of surgical methods have been described to treat CNPUI. Huntington and Haultain are commonly used abdominal approaches. Huntington procedure involves grasping the round ligaments and the uterus

below the area of inversion and slowly pulling up repeatedly until the uterus is reinverted [8]. Haultain procedure is where vagino-cervical ring is incised posteriorly and carried up the posterior wall of the uterus until it can be reinverted to its normal anatomy. Then the uterine incision can be repaired or followed by hysterectomy [9].

Kustner and Spinelli procedures are the commonly used vaginal approaches. Kustner procedure is entering the pouch of Douglas vaginally and splitting the posterior aspect of the uterus and the cervix, and reinverting the uterus. In Spinelli operation incision is made on the anterior aspect of the cervix and then the uterus is reinverted. After both the procedures the uterine incision needs to be repaired after repositioning, if the fertility is wished or otherwise can be proceeded for routine vaginal hysterectomy.

Subtotal vaginal hysterectomy or vaginal hysterectomy without reinversion of the uterus has been described to treat CNPUI. Mayadeo et al. in 2003 described a case of incomplete lateral inversion of the uterus, diagnosed with laparoscopy, and treated with vaginal hysterectomy without reinversion of the uterus [10].

Simms et al. who treated a post-menopausal chronic inversion with performed subtotal vaginal hysterectomy, differed removal of the cervix, considering the risks associated with distorted anatomy [5]. Mwinyoglee et al. reported a CNPUI which was treated with vaginal hysterectomy without reinverting the uterus. They had to use USG to locate the bladder at the cervico-fornical region before making the incision. In addition, they had to bisect the corpus to access the top pedicles [11].

Conclusions

Chronic uterine inversion is a rare gynaecological condition that is difficult to diagnose even for the experienced gynecologists. A high index of suspicion is needed for its proper diagnosis. Differential diagnosis of chronic uterine inversion should always be kept in mind even if patient present with the lump through the vagina.

Acknowledgment;

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A case of recurrent intussusception in a young child.

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Abstract :

The early diagnosis of intussusception is a challenge for clinicians. This index case presented with severe abdominal pain associated watery diarrhea in each time at the age of 6 month, 9 month and three and half years. It was ultimately diagnosed as intussusception by ultrasonography and successfully treated with hydrostatic reduction. It is planned for interval laparoscopic fixation.

Key words: Uterine inversion; Haultain's method

Introduction

Intussusception is a common serious pediatric emergency and common in infants (1). Unless we diagnose early, Complications such as gangrene of bowel, perforation, and peritonitis are inevitable. The mortality rate is less than 1% in children due to intussusception. If left untreated, however, this condition is consistently fatal in 2-5 days (2). Recurrence of intussusception is uncommon. We report a case who had been presented with recurrence of intussusception from the age of 6 months and last presentation was at the age of three and half year.

Case history

A three and half year old male child presented to pediatric ward with the history of loose stool fever and vomiting for one day duration and abdominal pain, and refusal of feeds for 6 hours. He passed bloody stool once with the last admission. The pain was intermittent and severe, and it lasts two to three minutes. In between pain, child looked well. He had similar events in the past twice at the age of 6 months and 9 months with abdominal pain and diarrhea. Otherwise the history was unremarkable.

On examination, he was lethargy, ill and crying at the time of pain. He was afebrile and hydration was poor. Abdomen showed normal finding and per-rectal examination exposed empty rectum. Rest of the system examination was clinically normal.

Full blood count, C-reactive protein and urine analysis were normal. Ultrasound scan revealed a mass contain bowel within bowel, appearance like target sign, seen in the right hypochondria. It favored ileocolic intussusception. Few mesenteric lymph nodes were noted within it. So intussusception was confirmed. It was treated with ultrasound guided hydrostatic reduction. Child was kept observation for 48 hours. Diagnostic laparoscopy excluded duplication cyst, meckel diverticulum and polyp. It is planned for interval laparoscopic fixation as it was ilioocolic intussusception in each time with no obvious underlying cause.

Discussion

Intussusception is a one of the grave pediatric emergency and the most common important cause of acute abdomen in infancy(3). Definition of Recurrent intussusception is an occurrence of intussusception of a bowel loop in a patient with a prior

resolution of intussusception, either spontaneously or with an intervention (4). Although it classically occurs in children between 2 months and 2 years (1). There were several incidence with the introduction of complementary food at the age of 6 months (5). Our index case also presented with the introduction of weaning food at the age of 6 months. The roots of intussusception are not fully identified. The majority of cases in young children are idiopathic although it is known that some viral and bacterial infections of the intestine may possibly induce intussusception in infancy (2).

The clinical triad of abdominal pain, currant-jelly stools or hematochezia and palpable abdominal mass are present in less than 50%. There may be some nonspecific abdominal symptoms such as vomiting, and the absence of passage of blood via the rectum. As child is nonverbal in the young age, the diagnosis of intussusception is missed in almost 50% of cases (3). Our index case had fever, vomiting, abdominal pain and bloody stool once, but typical triad of clinical features were not there. As he had fever and few lymph nodes in the ultrasonography, probably, the cause would be viral in origin.

Ultrasonography (US) has a high sensitivity for the diagnosis of intussusception (98%–100%). The routine ultrasonography (US) in the diagnosis of intussusception would be a reliable investigation and also it gives the high level of patient comfort and safety. A potential limitation is that US is not available 24 hours a day at all institutions (3). In addition, plain X-ray abdomen would be helpful in late presentation to exclude perforation and peritonitis. As he had the past history of similar diagnosis, and also parents brought the child early, we have diagnosed easily with US.

Furthermore, there are some common conditions which presents with similar ways such as infantile colic, gastroenteritis or less common conditions such as appendicitis and complicated Meckel diverticulum(5). So accurate diagnosis is important to prevent complications.

Nonsurgical treatment of intussusception is possible in most cases, especially at institutions with the backup radiologist and pediatric surgeon with the US facilities. Use of US guidance excludes the disadvantage of radiation exposure, and facilitate a greater number of attempts at hydrostatic reduction (6,7). Our case was successfully reduced with hydrostatic reduction,

Complicated with perforation and failure of hydrostatic reduction leads to surgical reduction of intussusception. There is a role laparoscopic fixation of bowel in the case of idiopathic cases to prevent further recurrence(6) and also to identify the risk factors such as duplication cyst, meckel diverticulum and, polyp which need an appropriate surgical correction(8,9).

Acknowledgment

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Limited Exchange Transfusion Can Be Very Beneficial in Sickle Cell Anemia with Acute Chest Syndrome

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Abstract :

Sickle cell disease (SCD) is a genetic disorder affecting red blood cells with a high prevalence in Africa and a high morbidity and mortality [1]. Among complications of SCD, the acute chest syndrome (ACS) is a potentially life-threatening one. The management of ACS aims to support the patient through supplemental oxygen therapy, appropriate analgesics, hydration, and antibiotics. We report on a critically ill young woman with SCD and ACS who was successfully managed by manual partial exchange trans- fusion in our hospital.

Introduction

Sickle cell disease (SCD) is a genetic disorder affecting red blood cells with a high prevalence in Africa and a high morbidity and mortality [1]. Among complications of SCD, the acute chest syndrome (ACS) is a potentially life-threatening one; it presents with chest pain, fever, cough, dyspnoea; tachypnoea, decreased peripheral oxygen saturation, and pulmonary infiltrate on chest X-ray. ACS is a frequent cause of hospitalization in SCD patients [2], and it may account for up to 25% of SCD-related deaths. The mortality risk due to ACS has been estimated to be four times higher in adults compared to children [3, 4].

The management of ACS aims to support the patient through supplemental oxygen therapy, appropriate analgesics, hydration, and antibiotics [5]. Blood transfusion is indicated in order to increase the oxygen carrying capacity of the blood and reduce the proportion of red cells susceptible to sickling [6, 7]. However, a simple (so-called "top-up") transfusion may also immediately cause increased blood viscosity and an increased circulating blood volume, which may increase the load on the heart. Furthermore, recurrent blood transfusions contribute to iron overload [8]. For these reasons, exchange blood transfusion or red cell exchange (RCE) has been introduced and is recommended for the management of ACS [3, 5].

RCE is now a day's frequently carried out by the use of automated equipment; however, it can be also carried out manually with minimal equipment [9]. We report on a critically ill young woman with SCD and ACS who was successfully managed by manual partial exchange trans- fusion in our hospital.

Case history

Prashalini 22 year old girl, a resident of batticaloa, she was a known patient of sickle cell- β^+ thalassemia trait, Never received blood transfusions in the past, She was recently diagnosed hypothyroidism , was on hematology clinic regular follow up. She was admitted to the medical ward with with B/L knee joint pain and B/L ankle joint pain with intermittent fever for one week. On physical examination, the patient was febrile (38°C), pale, mildly jaundiced, febrile, with multiple joint tenderness was present. Investigation revealed WBC 14*10³, Hb-8.4g/dl, plt-452, LFT-Total bilirubin-27.4micromol/L, CRP-7mg/dl

After 2 days of admission she became tachypnoeic, and dyspnoeic without lower limb edema. Her respiratory rate was 50breaths/min, pulse rate was 124bpm, and blood pressure was 124/64mmHg. Her oxygen saturation (SPO₂) was 65% on room air and 100% on oxygen 5l with face mask. On chest examination, there were reduced breath sounds in the right base. She had no

palpable enlargement of the liver or spleen. Hemoglobin was 84g/L, leukocytosis of $19.8 \times 10^9/L$, with a predominance of neutrophils ($12.4 \times 10^9/L$), and platelets of $535 \times 10^9/L$ (Table 1). From previous records, her steady-state hemoglobin was about 70g/L. Urine culture revealed no bacterial growth after 24 hours; renal function tests and serum electrolytes were within the normal range. A chest X-ray showed right-sided consolidation, bilateral mid-zone changes, and cardiomegaly. A diagnosis of ACS was made, but pneumonia and septicemia could not be excluded. The patient was placed on oxygen supplementation and was started on IV ceftriaxone 2g bd, IV normal saline (NS) 3L/24 hours, and oral ibuprofen and folic acid were continued. Pain was managed by sub cut morphine 5mg 4 hourly. Two units of packed red cells were cross matched and transfused.

After 5 day, the difficulty in breathing worsened, and the patient was transferred to a Medical Intensive Care Unit (MICU). Oral clarythromycine 500mg bd was added to her therapy as a cover for possible atypical pneumonia. Despite improvement in the right basal consolidation on the repeat chest X-ray, the bilateral mid-zone changes were still present and there was no improvement in her clinical condition.

On day 2 in the MICU, the patient's condition deteriorated: her RR was 72/min with PR of 124b/min and SPO₂ ranging between 60 and 66% on room air, patient intubated and ventilated. The patient was prepared for manual RCE. Prior to exchange transfusion, the hemoglobin was 95g/L and HbS quantified by HPLC was 78.2%, as a result of the recent blood transfusion, RCE was performed in two steps 20 hours apart. In the first step, a total of 500ml of the patient's blood was replaced by 250ml of normal saline (NS) and 250ml of packed red cells. In the second step, 450ml of the patient's blood was exchanged with 250ml of NS and 200ml of packed red cells. In total, 1400ml were replaced, corresponding to an estimated 40% of the total blood volume.

Post Exchange transfusion Hb-10.4g/dl achieved, DAT negative, D-Dimer negative, NCCT brain normal, Post Exchange HPLC revealed HbS 18%, 5th day of MICU stay patient was clinically improved and extubated following days patient fever continued with adequate antibiotic cover with dropping Haemoglobin, clinical diagnosis made hyper hemolysis syndrome started IV immunoglobulin 2g/kg give for day followed oral prednisolone 1mg/kg/day gradually improved fever settled, hemoglobin maintained 10-11g/dl without transfusion, she was discharge after 3weeks of admission with the plan off clinic review and tail off regime of predniselone.

Family workup of the patient revealed that his father and mother had sickle cell and beta thalassemia trait, respectively.

Discussion

Blood transfusions in SCD can be simple or exchange transfusions. RCE procedure has seen an increased popularity in these last 1-2 decades,[5] since it allows for an effective treatment for both acute sickling crisis unresponsive to conventional therapies

and a prophylactic treatment for high-risk patients. With RCE, hematocrit and HbS can be adjusted rapidly and simultaneously, allowing for intervention in an emergency and eliminating the risks associated with alterations in viscosity and patient's blood volume.[6]

For acute complications of SCD, the goal of transfusion therapy is to reduce the posttransfusion HbS level to <30%; for chronic complications, the goal is to maintain the pretransfusion HbS level at <30%-50% while maintaining the Hb level at ~10 g/dL. Rapid lowering of HbS levels can only be achieved by acute RCE.[7] Exchange can either be performed manually or by automated cell separators. Erythrocytapheresis can be performed using different cell separators, operating either with continuous or discontinuous flow.[6] The former is preferable when low-weight patients (namely pediatric patients) are treated, since these devices allow for a lower extracorporeal blood volume throughout the procedure. With the ready availability of automated cell separators and ease in technical performance, erythrocytapheresis is being used increasingly to treat acute and chronic complications of RBC disorders, particularly in patients with SCD. As opposed to automated RCE, manual RCE is labor intensive, prolonged, and perhaps less safe and efficient than erythrocytapheresis, but can be beneficial for cases with monetary, infrastructural, or venous access constraints.[7]

The two primary goals of transfusion are to correct the low oxygen-carrying capacity caused by severe anemia and to improve microvascular perfusion by decreasing the proportion of sickle red cells in the circulation.[6] RCE is recognized as the most rapid method for lowering HbS levels, offering potential advantages over simple transfusions, but its role in the treatment of SCD, except in severe crises, is still controversial and it is not widely adopted. Advantages of RCE are that the exchange prevents the removed sickle cells from participating in new vaso-occlusive events, reduces hemolytic complications, provides with added oxygen-carrying capacity, reduces iron accumulation, better control of blood volume and viscosity.[5]

In SCD, both transfusion methods (simple transfusion and erythrocytapheresis) offer similar benefits in maintaining target HbS levels for long-term transfusion therapy. Although simple transfusion is available worldwide and is simple to perform, erythrocytapheresis is not universally available, requires experienced personnel to perform, and may require a central venous catheter/port. The distinctive benefits of chronic erythrocytapheresis are prevention of iron overload. In addition, erythrocytapheresis may avoid the risk of circulatory volume alterations and hemodynamic distress and, thus, is a safer procedure than other methods of transfusion. However, to date, the advantages and efficacy of RCE have not been substantially documented through clinical trials, especially compared with simple transfusion or manual versus automated RCE.[7] All SCD patients undergoing major surgery are prepared in advance with transfusion to correct their anemia to a Hb of approximately 10 g/dl and HbS percentage to approximately 30%.[2,3,]

Conclusion

This patient with SCD met the clinical and radiological criteria for the diagnosis of ACS, and on RCE, she had full clinical resolution of ACS. This result was obtained in spite of the fact that only 1.4l of blood was exchanged, instead of the recommended amount of 1.4L [7]. Furthermore, after RCE, her HbS level was 18% and did not reach the commonly recommended <30%. With respect to the clinical benefit observed in this patient, we offer the following comments:

- 1) Although the patient had marked tachypnoea and marked desaturation, she did tolerate a 6-day wait before the exchange; a patient with a more severe ACS might not have survived that long. Therefore, we classify her ACS as moderately severe.
- 2) In spite of this delay, we have no doubt that RCE was crucial in resolving the patient's ACS; indeed, her respiratory rate started decreasing and her blood oxygen saturation started improving soon after the RCE was started. Both parameters were completely normal by the time RCE was over.
- 3) In spite of the eventual good outcome, RCE should have been carried out earlier, before the clinical state became life threatening.
- 4) Although we are not aware of any evidence basis for the currently recommended volume of RCE or target HbS percentage, we are not suggesting a change to current recommendations. However, we note that too many times RCE is not carried out due to "lack of equipment" or shortage of blood. This case illustrates that the equipment needed is minimal and that even 2 blood units together with equivalent amount of normal saline may be sufficient to produce a dramatic clinical benefit.

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Acute pancreatitis and hypertensive crisis in pregnancy diagnosed as primary hyperparathyroidism

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Abstract :

The occurrence of acute pancreatitis in primary hyperparathyroidism during pregnancy is less common. It can lead to significant complications to both mother and fetus. Physiological changes of pregnancy may attenuate the signs and symptoms of primary hyperparathyroidism. Common risk factors for the development of acute pancreatitis during pregnancy include gallstones, hyperlipidemia, alcohol consumption, and genetic variants. Our patient had another rare cause of pancreatitis, due to hyperparathyroidism induced hypercalcemia.

Key words

Primary hyperparathyroidism, Hypercalcemia, Acute pancreatitis, Pregnancy.

Introduction

Primary hyperparathyroidism (PHPT) is a disorder of calcium homeostasis caused by inappropriate overproduction of parathyroid hormone. It is the third most common endocrine disorder and the most common cause of hypercalcemia. It is a rare condition during gestation, occurring in less than 1% of all during pregnancy with less than 200 cases have been reported since the condition was first recognized in 1930 by Hunter.⁽¹⁾ It can lead to significant complications to both mother and fetus. Physiological changes of pregnancy may attenuate the signs and symptoms of primary hyperparathyroidism.

Acute pancreatitis during pregnancy is rarely encountered and can occur during any trimester, with an incidence ranging from 0.02 to 0.1%.⁽²⁾ It has the potential to cause significant maternal mortality and fetal loss. Common risk factors for the development of acute pancreatitis during pregnancy include gallstones, hyperlipidemia, alcohol consumption, and genetic variants. Our patient had another rare cause of pancreatitis, due to hyperparathyroidism induced hypercalcemia.

Case history

A 21 year old primi gravid mother with back ground history of polyuria, polydipsia, renal stone, recurrent urinary tract infection, hyperemesis gravidarum and gestational hypertension, presented with severe epigastric pain at 33 weeks of gestation. On examination she had blood pressure of 160/90 mm Hg with bilateral pleural effusion and tenderness over epigastrium. Her serum amylase level was very high 1600U/L. Laboratory evaluation revealed hypokalemia (2.6mEq/L), hyponatremia (135mEq/L), hypercalcemia (13.5mg/dl), hypophosphatemia (2mg/dL), hypomagnesemia (1.4mg/dl) and elevated intact parathyroid hormone level of 176pg/ml. Arterial blood gas revealed normal anion gap metabolic acidosis with urinary pH 7.6. Ultrasound scan of abdomen revealed swollen hypoechoic pancreas, left renal calculus and nephrocalcinosis. Neck imaging revealed no pathological parathyroid tissue. She was diagnosed with primary hyperparathyroidism complicating with acute pancreatitis, pregnancy associated hypertensive crisis, nephrocalcinosis and distal renal tubular acidosis. She was stabilized and a healthy baby girl was delivered at 35-week gestation by caesarean section with an unremarkable neonatal course. She was referred for SPECT scan and followed by parathyroidectomy.

Presentation

A 21 year old primigravid woman was admitted at 33 weeks of gestation with severe abdominal pain confined to the upper abdomen, which was sharp and persisting pain radiate to her back and associated with severe vomiting and nausea. She also had a six month history of polyuria, polydipsia and non specific abdominal pain. She had urinary tract infection and treated twice with oral antibiotics during pregnancy. Passage of a stone during urination was noted once. She also had poor sleep with increased lethargy and mood changes. Her pregnancy had been complicated by prolonged hyperemesis gravidarum and was found to be with high blood pressure on her monthly antenatal visit at 30 weeks of gestation. She did not take any other medications or herbal supplements except antacids for peptic ulcer disease. She did not have any family members with renal stones.

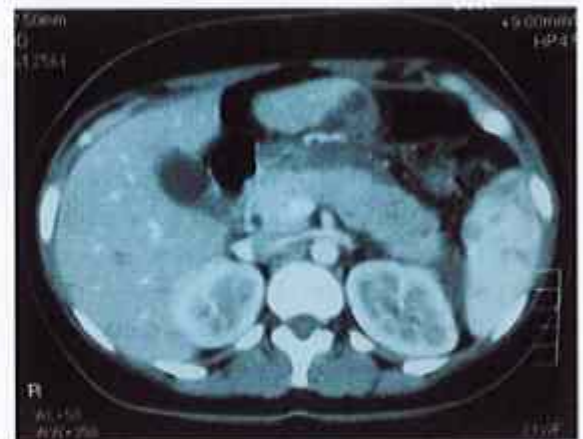
On admission, her temperature was 37.60c, pale, dehydrated, pulse rate was 96 beats per minute, blood pressure was 160/90mmHg, respiratory rate was 24 breaths per minute and oxygen saturation was 94% on air. She had severe epigastric tenderness with guarding. Respiratory examination indicated decreased breath sounds in the bilateral lower hemi thorax with stony dullness on percussion. The findings on neurological examination were unremarkable. Uterus was 32weeks gravid size, relaxed and fetal heart rate was at 140/min.

Laboratory tests done on the day of admission showed a white blood count of 18670/mm³, haemoglobin 9.5g/dl, and a platelet count of 318000, Serum amylase 1600 IU/l and C-reactive protein 52mg/dl. Urine analysis did not reveal any pyuria. Fasting blood sugar and renal function tests were within normal limits. Laboratory evaluation revealed hypokalemia (2.6mmol/l) hyponatremia (135mmol/l), hypercalcemia (13.5mg/dl), hypophosphatemia (2mg/dl) and hypomagnesemia (1.4mg/dl). Hyperparathyroidism was suspected, which was confirmed by an elevated intact parathyroid hormone level of 176pg/ml (normal range 10-59pg/ml). Arterial blood gas parameters were the following: pH 7.35, carbon dioxide partial pressure 36mmHg, bicarbonate level 20mg/dl, and base excess -4.2mEq/l. Her urine biochemistry revealed urine osmolality 250mOsm/l, urinary pH 7.6, urinary calcium 9.6mg/dl, urinary creatinine 26mg/dl, urinary calcium creatinine ratio 1.04, urinary sodium 34mmol and urinary potassium 26.1 mmol/l on spot sample.

Ultrasound scan of abdomen revealed swollen hypochoic pancreas, 1.1cm calculus seen at lower edge of left kidney and nephrocalcinosis changes. It showed a single live intrauterine fetus of 32 Weeks \pm 2 days of gestational age with normal cardiac activity and adequate liquor. Computed tomography scan of the abdomen was not done because of her pregnant state. Neck imaging revealed no pathological parathyroid tissue. Except for the lack of localization imaging, all other features in our patient were consistent with hypercalcemia due to primary hyperparathyroidism complicated with acute pancreatitis, nephrolithiasis, nephrocalcinosis, distal renal tubular acidosis

and pregnancy associated hypertensive crisis.

Patient was shifted to the intensive care unit and managed by nil orally, intravenous fluids, antibiotics, analgesics, antipyretics with strict fetal monitoring. The patient was started on IV labetalol for blood pressure control with methyldopa. Given the gestational age, it was decided to treat her hyperparathyroidism with intravenous fluids and loop diuretic. The serum calcium level ranged from 13.5 to 10.5 mg/dl after hydration. The patient improved with this treatment and was shifted back to the antenatal ward after 5 days. Meanwhile, she was treated with dexamethasone to promote fetal lung maturity. At the 35th week gestation, after calcium levels began to rise again, she underwent emergency cesarean section. A female newborn was delivered with low birth weight of 2kg and Apgar score of 9 at both 1 and 5 minutes. Blood tests showed normal serum values of calcium with altered coagulopathy. She had received one dose of pamidronate and discharged with losartan. Four weeks after delivery, her calcium levels was 11.8 mg/dl, phosphorus level was 2mg/dl. CT showed early changes of nephrocalcinosis in both kidneys with 1 cm calculus in lower edge of left kidney and tetrofosmin parathyroid scan was normal. Patient was referred for SPECT scan and followed by parathyroidectomy.



CT scan - early changes of nephrocalcinosis



99m Tc-Tetrofosmin Parathyroid scan

Discussion

Most cases of hypercalcemia diagnosed during pregnancy are due to primary hyperparathyroidism. Hypercalcemia in pregnancy can be secondary to familial benign hypocalciuric hypercalcemia, milk alkali syndrome, parathyroid-related-protein (PTHrP) mediated hypercalcemia, neuroendocrine tumor of the pancreas, humoral hypercalcemia of pregnancy. Hypercalcemia with elevated parathyroid hormone levels were seen in primary hyperparathyroidism and familial benign hypocalciuric hypercalcemia. Urinary calcium excretion is inappropriately low in familial benign hypocalciuric hypercalcemia and it can be reasonably differentiated from primary hyperparathyroidism based on the calcium to creatinine clearance ratio of less than 0.01 with 85% sensitivity and 88% specificity.(3)

The most common cause of primary hyperparathyroidism in pregnancy is parathyroid adenoma, accounts for 85% of cases, followed by 10% from primary parathyroid hyperplasia, 3% from multiple adenomas and 2% from parathyroid cancer.(4) It can also be seen in association with multiple endocrine neoplasia. Hypercalcemia can lead to a variety of clinical symptoms depending on the degree of serum elevation. Mild hypercalcemia, defined as less than 12mg/dl, produces vague symptoms which are similar to common complaints of pregnancy such as fatigue, constipation, nausea, vomiting and depression. Once hypercalcemia becomes more severe, with levels above 13 mg/dl, the resultant symptoms are much less commonly seen in normal pregnancy. These include renal manifestations, central nervous system abnormalities, osteoporosis, cardiac dysrhythmia, acute pancreatitis, peptic ulcer disease, pseudogout and muscle atrophy. Hypercalcemic crisis has been reported with calcium levels above 14 mg/dl leading to uremia, coma and maternal death. Maternal hypercalcemia can directly affect the fetus as well. It leads to an increase in net placental calcium transport, which can excessively suppress the fetal parathyroid glands and lead to neonatal hypocalcemia. It can lead to tetany with respiratory failure in new born.

Renal manifestations of severe primary hyperparathyroidism include nephrolithiasis, nephrocalcinosis and renal functional abnormalities that range from impaired concentrating ability to end-stage renal failure. Kidney stones are reported in only 10% to 25% of patients with primary hyperparathyroidism. Hypercalciuria and alkaline urine are considered risk factors that lead to stone formation. Longer disease duration can predispose to medullary nephrocalcinosis and lead to distal renal tubular acidosis. Hypokalemia, metabolic acidosis and high urinary pH (>5.3) were favoured diagnosis of type 1 renal tubular acidosis. Central nervous system abnormalities include mental status changes, lethargy, depression and confusion. Hypertension is frequently observed in patients with primary hyperparathyroidism. In our patient, hypertension was not well controlled and hypertension persists even after delivery.

Pancreatitis is a rare condition in pregnancy. Our patient had acute pancreatitis due to hyperparathyroidism induced hypercalcemia. The association of primary hyperparathyroidism and pancreatitis can take many forms. So, Jacob et al. proposed a classification of this association which can be in 4 forms; PHPT revealed by acute pancreatitis, PHPT revealed by recurrent acute pancreatitis without chronic pancreatitis, PHPT revealed by a chronic pancreatitis with or without pancreatic calcifications, or PHPT complicated by acute pancreatitis in the postoperative period(5). Persistent hypercalcemia might increase the level of calcium concentration in pancreatic juice and activates trypsinogen to trypsin causing pancreatic ductal and parenchymal damage leading to pancreatitis.(6) Hypercalcemia causes formation of protein plugs in the pancreatic duct which obstruct the flow and leading to pancreatitis. Hypercalcemia also causes vasoconstriction and pancreatic duct narrowing. Some believe that parathyroid hormone itself acts as a toxin causing local thromboendarteritis. (7) All these might result in pancreatic tissue necrosis.

Treatment in patients with pancreatitis on Primary hyperparathyroidism first rests on a management of the acute pancreatitis which can be fatal. Early recognition and better supportive treatment of acute pancreatitis are the main stay of the management in the third trimester of pregnancy with delaying the definitive treatment until after delivery. There are several other documented maternal and fetal complications associated with pancreatitis in pregnancy. These include preeclampsia, pre term delivery at neonatal respiratory distress and intrauterine fetal demise.

Physiological changes in calcium homeostasis during pregnancy like maternal blood volume expansion, hypoalbuminemia and calcium transport across the placenta mask the symptoms of hypercalcemia in pregnancy. Asymptomatic presentation and rarity of primary hyperparathyroidism in the child bearing age group makes its diagnosis during pregnancy challenging. Laboratory findings are also masked by pregnancy-induced changes in calcium homeostasis.(8) Ionized calcium is the initial diagnostic test for this disease and if elevated should be followed by parathyroid hormone levels.

Imaging studies are used predominantly to plan the operative approach. Ultrasound scan of neck is the first choice during pregnancy which has a sensitivity of 61%. Sestamibi scintigraphy is contraindicated during pregnancy which has a sensitivity of 86% in diagnosis.(9) Negative sestamibi does not preclude the diagnosis of primary hyperparathyroidism, since it occurs in 12-25 percent of patients with disease.(10) Sestamibi single photon emission computed tomography(SPECT) is a three dimensional sestamibi scan that provide higher resolution imaging with detection of ectopic glands and multiglandular disease. Other imaging modalities been used successfully include four dimensional computed tomography and positron emission tomography.

A multidisciplinary approach is needed to assure optimal outcomes in pregnant women with primary hyperparathyroidism. Management should be carefully individualized to minimize maternal and fetal complications. Patients undergoing conservative medical management must be maintain adequate hydration. Bisphosphonates should not be used in pregnant because they cross the placenta. Calcitonin promotes renal calcium excretion and can be use in refractory hypercalcemia during pregnancy. Calcium-sensing receptor agonist cinacalcet has been used to control extreme hypercalcemia in pregnancy. Dietary calcium should not be restricted as low calcium diet may lead to further increase in parathyroid hormone secretion. Fetal well being should be monitored with serial obstetric ultrasounds and cardiotocographic monitoring. Women with Primary hyperparathyroidism during pregnancy should undergo parathyroidectomy during the second trimester to minimize complications. Standard surgical approach is bilateral neck exploration. Minimally invasive parathyroidectomy is emerging as the procedure of choice with the improvement of imaging modalities and parathyroid hormone level monitoring.

Conclusion

Primary hyperparathyroidism can lead to significant complications for both mother and fetus. Life threatening acute pancreatitis secondary to primary hyperparathyroidism can occur very rarely. The pregnant state presents a challenge to the diagnosis and treatment of primary hyperparathyroidism. High level of suspicion of primary hyperparathyroidism and an early diagnosis can thus greatly benefit and help to prevent complications. Well-organized multidisciplinary management decisions can significantly reduce the morbidity and mortality associated with the disease during pregnancy Parathyroidectomy in the second trimester appears to be a safe as definitive therapy.

Competing interests

The authors declare that there are no competing interests regarding the publication of this paper.

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