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EDITORIAL

Smart Phone among doctors

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Smartphone have become ubiquitous in our day-to-day living. The purpose-built software applications available on these phones have contributed to their emergence in clinical practice and increasing prevalence amongst doctors. Advancements in technology have always had major impacts in medicine. The Smartphone is one of the most omnipresent and energetic trends in communication, in which one's mobile phone can also be used for communicating via email, performing Internet searches, and using specific applications. Smartphone have penetrated the medical world and are beginning to be embraced by the profession. New apps are popping up which can diagnose patients and even allow you to speak to a doctor on demand. The most commonly used apps include those for disease diagnosis, management, drug reference, clinical scoring systems, and medical calculators.

The Smartphone is one of the fastest growing sectors in the technology industry, and its impact in medicine has already been significant. It usage has spread to many settings including that of healthcare with numerous potential and realized benefits. The ability to download custom-built software applications has created a new wealth of clinical resources available to healthcare staff, providing evidence-based decisional tools to reduce medical errors. We found a wide range of apps for the layperson, from wellness apps to apps that allow better communication with health

care providers. The greatest concern is the general lack of regulation and an evidence base for many of these wellness apps. Much like the general information available on websites, the content of many health-related apps is poorly scrutinized for accuracy. From patient monitoring and diagnostics to more efficient medical education and communication, Smartphone serve a vital role in the practice of medicine today. In this editorial, we will the available literature to understand how the Smartphone has changed the field of internal medicine and medical education. We also survey the ways in which the Smartphone is used to better understand how that impact might be achieved.

With respect to patient care and monitoring, we found various ways of using the Smartphone to monitor patients. We identified research attempting to provide evidence that the Smartphone has advantages in this area; however, much of this is still in the preliminary phase. Apps such as iWander for people with dementia (see Patient Care and Monitoring above) could improve quality of life and decrease financial burden. As we enter a new era of rising medical costs exacerbated by a growing elderly population, our health care system is looking for ways to meet the rising demand. It remains to be seen whether the Smartphone can help. There is currently no governing body that regulates medical apps to ensure that their content is correct, up-to date, and reliable. To address these issues, further in-depth focus on vigorous institutional policies is necessary to ensure that patient information is stored and transmitted securely on Smartphone. While the Smartphone's role in medicine and education appears promising and exciting, more high-quality studies are needed to better understand the role it will have in this field. We recommend popular Smartphone applications for physicians that are lacking in evidence and discuss future studies to support their use.

The FDA does not control the content of most apps; only when apps cross the line of providing direct medical advice does the FDA make approval mandatory. An example of this is the diabetes app WellDoc Diabetes Manager System (WellDoc, Inc, Baltimore, MD, USA), which required FDA approval when it started providing medical advice based on input blood glucose levels. Other apps available for free download include symptom checkers, where people can input basic symptoms such as abdominal pain and get a whole list of possible causes, prompting inappropriate self-diagnosis and unneeded anxiety.

The amount of research in the use of the Smartphone in medicine is rapidly growing, but there are very few good- quality studies to answer many questions about its use and the impact it may have. Apps for pharmacology, medical references, and a myriad of other categories are providing physicians with quick and practical medical information that will aid in education and patient care. Communication within hospitals and between patients is improving. Additionally, developing countries now have the potential to access better diagnostic tools in resource-poor regions. However, many obstacles still stand in the way of this progress. The question regarding whether Smartphone in medicine will lead to a healthier population with better patient–doctor relationships remains to be answered. Nevertheless, the Smartphone has a very bright future in the world of medicine, while doctors, engineers, and others alike continue to contribute more ingenuity to this dynamic field. It is our hope that by informing the medical community of the numerous ways in which the Smartphone can be used to benefit health care providers, patients, and their families, the Smartphone may one day be recognized as a diagnostic and therapeutic tool that is as irreplaceable as the stethoscope has been in the practice of medicine.

Today, Smartphone are being manufactured by numerous companies and are one of the fastest growing sectors in the technology industry. Operating systems include Google's Android, Apple's iOS, Research in Motion's BlackBerry, Nokia's Symbian, and the Windows Phone 7 platform; we conclude this editorial with suggested apps for doctors based on anecdotal experience and suggest studies that can

better answer these questions. In conclusion, it is undeniable that the use of personal Smartphone in clinical practice is an established reality, particularly amongst junior doctors. Smartphone apps provide user-friendly and intuitive interfaces for junior doctors to efficiently perform their daily clinical tasks. However, if barriers and benefits to implementing and recommending this technology remain constant among doctors, the early addition of Smartphone education to the medical curriculum and in continuing medical education would be beneficial to current and future healthcare providers.(1,2)(2)

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MANAGEMENT OF OSTEOPOROSIS

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Abstract

Osteoporosis is a disorder characterized by reduced bone strength, diminished bone density, and altered macro-geometry and microscopic architecture. Bone loss related to declining estrogen levels increases fracture risk in postmenopausal women, who make up the majority of osteoporosis cases.Both peak bone mass and subsequent bone loss can be modified by environmental factors, such as nutrition, physical activity, and concomitant diseases and medications. Osteoporosis prevention requires adequate calcium and vitamin D intake, regular physical activity, and avoiding smoking and excessive alcohol ingestion. The present review provides a brief outline of the current evidence in the management of osteoporosis. Novel anti-osteoporotic agents are also briefly presented.

Introduction

Osteoporosis is a condition in which bones lose their strength and are more likely to fracture, usually following a minor trauma or fall. Fractures that occur because of reduced bone strength are described as 'fragility fractures' and many of these will be caused by osteoporosis. One in two women and one in five men over the age of 50 experience fractures, mostly as a result of low bone strength. Although fragility fractures caused by osteoporosis can happen in various parts of the body, the wrists, hips, and spine are the most commonly affected sites. Women are more susceptible to osteoporosis because bone loss becomes more rapid for several years after the menopause when sex hormone levels decrease. Adult bone mass is the integral measurement of the bone mass level achieved at the peak minus the rate and duration of subsequent bone loss. There is clearly a genetic predisposition to attain peak bone mass, which occurs by a person's mid-20s. Bone loss with age and menopause are universal, but rates vary among individuals. A previous vertebral or hip fracture is the most important predictor of fracture risk. Bone density is the best predictor of fracture risk for those without prior adult fractures. Age, weight, certain medications, and family history also help establish a person's risk of osteoporotic fractures.

Impact of osteoporosis

Fractures due to osteoporosis have a serious impact on a person's health, happiness, and quality of life. They can result in chronic pain, long-term disability and death. Despite its serious impact, the disease remains under diagnosed and under treated. Around 80% of people at high risk and who have already had at least one osteoporotic fracture are not identified or treated. Hip fractures result in pain, reduced mobility, disability, and an increasing degree of dependence. People who survive a hip fracture often suffer a loss of physical function and independence. 40% are unable to walk independently and 60% still require assistance a year later. Vertebral fractures can lead to back pain, loss of height, deformity, immobility, increased number of bed days, and even reduced pulmonary function. Vertebral fractures significantly affect the ability of people to carry out activities of daily living. Besides their impact on quality of life, they can result in loss of self-esteem, distorted body image and depression. In many countries, fractures caused by osteoporosis are responsible for more days of hospitalization among women over 45 years of age than most other diseases.

Pathophysiology

Osteoporosis is caused by an inequity of bone resorption and bone remodeling leading to decreased skeletal mass. In most individuals, bone mass peaks in the third decade, after which bone resorption exceeds bone formation. Failure to reach a normal peak bone mass or acceleration of bone loss can lead to osteoporosis.

Diagnosis

Traditional X-rays can't measure bone density, but they can identify spine fractures. Bone mineral density (BMD) has to be measured by more specialized techniques. A number of different types of BMD tests are available, but the most commonly used is DXA (dual-energy X-ray absorptiometry). DXA is a low radiation X-ray capable of detecting quite small percentages of bone loss. It is used to measure spine and hip bone density, and can also measure bone density of the whole skeleton. There are

a number of different types of test options are available however a DXA scan, which is used to measure spine and hip bone density, is the most common technique for assessing the risk of osteoporosis

The World Health Organization (WHO) has defined a number of threshold values (measurements) for osteoporosis. The reference measurement is derived from bone density measurements in a population of healthy young adults (called a T-score). Osteoporosis is diagnosed when a person's BMD is equal to or more than 2.5 standard deviations below this reference measurement. Osteopenia is diagnosed when the measurement is between 1 and 2.5 standard deviations below the young adult reference measurement.

Status	Hip BMD
Normal	T-score of -1 or above
Osteopenia	T-score lower than -1 and greater than -2.5
Osteoporosis	T-score of -2.5 or lower
Severe osteoporosis	T-score of -2.5 or lower, and presence of at least one fragility fracture

Laboratory Evaluation

When a diagnosis of osteoporosis is made, a general laboratory evaluation, including FBC, serum calcium, and tests of renal and hepatic function, is warranted. In those with vertebral fractures and in those with particularly low z scores (-2 or below), an additional evaluation to exclude hyperthyroidism (thyroid-stimulating hormone), hyperparathyroidism (PTH), 25-hydroxyvitamin D, celiac disease (transglutaminase antibody), multiple myeloma (protein electrophoresis and/or immunofixation), and renal hypercalciuria (24 hour urine calcium) should be performed. If clinically suspected, a 24-hour urine cortisol or a dexamethasone suppression test should be performed to exclude Cushing's disease, and a serum tryptase can be considered to look for evidence of mastocytosis.

Risk factors for osteoporosis

A risk factor is anything that increases your chance of getting a disease. Having a risk factor, or even several does not mean that you will have osteoporosis. However, the more risk factors you have, the greater your chance of developing a disease and also, the greater the level of each risk factor, the greater the risk. There are different kinds of risk factors - fixed and modifiable. Some factors, such as age or gender, cannot be changed; whereas, others are linked to personal lifestyle choices, such as smoking, alcohol intake, and diet.

Fixed risks

- Age
- Female gender
- Family history of osteoporosis
- Previous fracture
- Ethnicity
- Menopause/hysterectomy
- Long term glucocorticoid therapy
- Rheumatoid arthritis
- Primary/secondary hypogonadism in men

Modifiable risks

- Alcohol
- Smoking
- Low body mass index
- Poor nutrition
- Vitamin D deficiency
- Eating disorders
- Insufficient exercise
- Low dietary calcium intake
- Frequent falls

Screening for osteoporosis

An easy and painless procedure, an osteoporosis screening requires you to place your foot in an ultrasound device called a bone densitometer. This device then measures the bone mineral density of your heel. The heel is measured because its bone is similar to that found in the hip, where fractures most often occur. Screening tests can help identify people who are most likely to benefit from further bone density testing.

The National Osteoporosis Foundation also recommends bone density testing in the following

- Women age 65 and older and men age 70 and older, regardless of clinical risk factors.
- Younger postmenopausal women and men age 50 to 69 about whom you have concern based on their clinical risk factor profile.
- Women in the menopausal transition if there is

a specific risk factor associated with increased fracture risk such as low body weight, prior low-trauma fracture, or high-risk medication.

- Adults who have a fragility fracture after age 50.
- Adults with a condition (e.g., rheumatoid arthritis) or taking a medication (e.g., glucocorticoids in a daily dose greater than or equal to 5 mg prednisone or equivalent for three months or longer) associated with low bone mass or bone loss.
- Anyone being considered for pharmacologic therapy for osteoporosis.
- Anyone being treated for osteoporosis, to monitor treatment effect.
- Anyone not receiving therapy in whom evidence of bone loss would lead to treatment.

Postmenopausal women discontinuing estrogen should be considered for bone density testing.

Secondary osteoporosis

Osteoporosis that results from having another disease or condition or from the treatment of another condition is called secondary osteoporosis. Secondary osteoporosis results from specific clinical disorders that are potentially reversible. Up to 30% of postmenopausal women and 50% of men with osteoporosis may have an underlying cause. The underlying pathogenesis of secondary osteoporosis is often multifactorial. The most commonly recommended laboratory tests include serum 25-hydroxyvitamin D, calcium, creatinine, and thyroid-stimulating hormone levels.

Common Causes of Secondary Osteoporosis.

- Medical conditions Central nervous system disorders (e.g., epilepsy, multiple sclerosis, Parkinson disease, spinal cord injury, stroke)
- Chronic obstructive pulmonary disease.
- Endocrine/metabolic disorders (adrenal insufficiency, athletic amenorrhea, Cushing syndrome, hemochromatosis, homocystinuria, primary hyperparathyroidism, hyper-prolactinemia, hyperthyroidism, primary or secondary hypogonadism, premature menopause, thyrotoxicosis, type 1 diabetes mellitus)
- Gastrointestinal disorders (celiac disease, gastric bypass, inflammatory bowel disease, malabsorption, pancreatic insufficiency, primary biliary cirrhosis).
- Hematologic disorders (hemophilia, leukemia, and lymphomas, monoclonal gammopathies, multiple myeloma, sickle cell disease, thalassemia).
- Human immunodeficiency virus infection or AIDS Liver disease (severe).
- Nutrition disorders (alcoholism, anorexia nervosa/bulimia, malnutrition, vitamin An excess, vitamin D deficiency).
- Renal insufficiency or renal failure.
- Rheumatoid arthritis , Systemic lupus erythematosus

Drugs increase fracture risk

- Corticosteroids (≥5 mg prednisolone daily or equivalent for ≥3 months).
- Antiepileptics: carbamazepine, phenytoin, phenobar-bitone.
- Hypoglycemics: thiazolidinediones, empagliflozin.
- Selective serotonin reuptake inhibitors.
- Excess thyroxine.
- Aromatase inhibitors.
- Tamoxifen (when used in pre-menopausal women).
- Gonadotropin-releasing hormone.
- Chemotherapy.
- Immunosuppressants: cyclosporine, tacrolimus, methotrexate.
- Lithium.
- Heparin.
- Proton pump inhibitors.
- Aluminium-containing antacids.
- Depot medroxyprogesterone acetate.
- Antipsychotics.

Management

The treatment of osteoporosis involves management of osteoporosis-associated fractures, universal prevention measures, and medical treatment of the underlying disease.

Non-pharmacological treatment

Nonpharmacologic preventive measures include modification of general lifestyle factors, such as increasing weight-bearing and muscle-strengthening exercise, which epidemiologic studies have linked to lower fracture rates, and ensuring optimum calcium and vitamin D intake as an adjunct to active anti fractured therapy[6]. In addition, potentially treatable underlying causes of osteoporosis such as hyperparathyroidism and hyperthyroidism should be ruled out or treated if detected.

Fracture management

Treatment of the patient with osteoporosis involves management of acute fractures and treatment of the underlying disease. Hip fractures almost always require surgical repair if the patient is to become ambulatory again, with procedures including open reduction and internal fixation with pins and plates, hemiarthroplasties or total arthroplasties. For vertebral compression fractures that present with back pain, analgesics, including nonsteroidal anti-inflammatory agents, acetaminophen, and sometimes narcotics, may be needed. Short periods of bed rest may be helpful for pain management, although early mobilization is advantageous to help prevent any further bone loss, as well as complications associated with prolonged bed rest.

Risk reduction

All patients who have osteoporosis or who are at risk should first review the risk factors and modify any that are modifiable, including those that contribute to bone loss as well as those that contribute to a risk of falling. Medications that may cause bone loss should be reviewed to ensure that the drug is truly indicated and is being given in the lowest possible dose. For those on thyroid hormone replacement, TSH levels should be measured to ensure that an adequate, but not excessive, a dose is being used. In patients who are smokers, efforts should be made toward smoking cessation, and alcohol abuse should be evaluated and treated.

Exercise

High physical activity throughout the life span has a beneficial effect on bone mass.Postmenopausal women who initiate weight-bearing exercises (done while standing) and/or muscle-strengthening exercises can help prevent bone loss. Exercise also has benefits on neuromuscular function, coordination, balance, and strength, and thereby can reduce the risk of falling. It is important that exercise be consistent, optimally at least 3 times a week, but any exercise is better than none.

Pharmacological treatment

Patients with the greatest fracture risk are those who have had prior vertebral or hip fractures, and a history of one of these or discovery of a prevalent deformity on x-ray or morphometry should be a mandate for medical therapy. Those patients who have osteoporosis by bone mineral density criteria, with a t score of -2.5 or below, should begin medical treatment. Women in the low-bone-mass range between -1.5 and -2.5 should be treated with medication depending on the number of other risk factors present and the strength of those risk factors as well as the personal motivation of the patient. Risk factors to be considered in making the treatment decision include age, prior history of fracture, family history of fracture, weight, underlying diseases and medications, bone turnover, and current smoking[8].

The currently approved medications include alendronate, risedronate, and raloxifene for prevention and treatment of osteoporosis; teriparatide and nasal calcitonin spray for treatment only and estrogens or combinations of hormones (hormone replacement therapy) for prevention only[9].

Bisphosphonates

Bisphosphonates are the most commonly used agents for osteoporosis. They have been employed for both treatment and prevention. Oral and intravenous options are available. Alendronate is approved for the treatment of osteoporosis in men, in postmenopausal women, and in patients with glucocorticoid-induced osteoporosis. It has been shown to increase spinal and hip mineral density in postmenopausal women. The treatment dose of alendronate is 70 mg/wk, to be taken sitting upright with a large glass of water at least 30 minutes before eating in the morning. Other oral bisphosphonates include risedronate or risedronate delayed-release, given daily, weekly, or monthly. It is also available as a combination product with calcium as risedronate/calcium carbonate. Ibandronate is another bisphosphonate that can be given orally once a month. Zoledronic acid is the most potent bisphosphonate

available. It increases BMD at the spine by 4.3-5.1% and the hip by 3.1-3.5%, as compared with placebo. Zoledronic acid is a once-yearly intravenous infusion approved for the treatment of osteoporosis in men, in postmenopausal women, and in patients with glucocorticoid-induced osteoporosis. The American association of clinical endocrinologists (AACE) recommends that if osteoporosis is mild, clinicians should consider a drug holiday after 4-5 years of bisphosphonate treatment; if fracture risk is high, a drug holiday of 1-2 years may be considered after 10 years of treatment[11].

Selective estrogen receptor modulators (SERMs)

SERMs are considered to provide the beneficial effects of estrogen without the potentially adverse outcomes. Raloxifene is a SERM indicated for the treatment and prevention of osteoporosis in postmenopausal women. The usual dose is 60 mg given orally daily. It can also be given in combination with calcium and vitamin D. Raloxifene is commonly associated with increased vasomotor symptoms[12]. It is associated with an increased risk of venous thromboembolism and a decreased risk of invasive breast cancer.

Parathyroid hormone
Teriparatide

Teriparatide is a recombinant human parathyroid hormone PTH, that acts as an anabolic agent for the treatment of osteoporosis[10]. It is indicated for the treatment of women with postmenopausal osteoporosis who are at high risk of fracture, who have been intolerant of previous osteoporosis therapy, or in whom osteoporosis treatment has failed to increase bone mass. Before treatment with teriparatide, levels of serum calcium, PTH, and 25(OH)D need to be monitored. Teriparatide cannot be given for more than 2 years[13].

Abaloparatide

Another PTH analogue, abaloparatide , was approved by the FDA in April 2017. Approval was based on results of 18 months from the Abaloparatide Comparator Trial in Vertebral Endpoints (ACTIVE) trial.

Calcitonin

Calcitonin nasal spray is an antiresorptive agent approved for the treatment of postmenopausal osteoporosis. It has been shown to decrease the occurrence of vertebral compression fractures only. Calcitonin is no longer widely used for a treatment of osteoporosis. Nevertheless, it remains an option for patients who are not candidates for other available osteoporosis treatments.

Denosumab

Denosumab is a humanized monoclonal antibody directed against the receptor activator of the nuclear factor-kappa B ligand (RANKL), which is a key mediator of the resorptive phase of bone remodeling. Denosumab also increases bone mass in men at high risk for fracture who are receiving androgen deprivation therapy for nonmetastatic prostate cancer. In these patients, denosumab also reduces the incidence of vertebral fractures

Hormone replacement therapy

Hormone replacement therapy (HRT) was once considered a first-line therapy for the prevention and treatment of osteoporosis in women. Although HRT is not currently recommended for the treatment of osteoporosis, it is important to mention because many osteoporosis patients in a typical practice still use it for controlling postmenopausal symptoms.

Vertebroplasty and Kyphoplasty

The goals of surgical treatment of osteoporotic fractures include rapid mobilization and return to normal function and activities. Traditional operative management of vertebral compression fractures is uncommon and is usually reserved for gross spinal deformity or for threatened or existing neurologic impairment.

Conclusion

Osteoporosis is a highly prevalent condition characterized by decreases in bone mass and microar -chitectural alterations. The detection of osteoporosis relies on the quantification of bone mineral density via imaging techniques such as dual-energy X-ray absorptiometry. However, therapeutic decision-making should be based on a comprehensive fracture risk assessment. Cost-effectiveness analyses support early detection and treatment of high-risk patients with antiresorptive medications such as bisphosphonates.

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Review Article

ACANTHOSIS NIGRICANS (AN)

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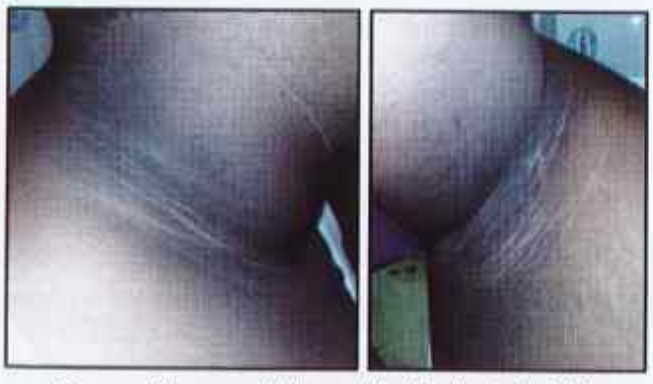
Introduction

Acanthosis nigricans is a skin condition charac -terised by dark, thick, velvety areas of intertriginous surfaces and they are less common in extensor surfaces. Most affected areas are neck, axillae, antecubital and popliteal fossae and groin folds[1].

Acanthosis nigricans is a Latin word (Acanthosis denotes 'Thick' and Nigricans denotes 'Dark')

Acanthosis nigricans can be classified into benign and malignant types. Benign AN include obesity associated, syndromic, drug induced, acral, unilateral, autoimmune, familial and mixed varieties. Malignant AN are those associated with underlying malignancies. Even though AN is solely diagnosed clinically, histopathological input might be needed for difficult cases. Further investigations are aimed at diagnosing the underlying cause.

An increasing prevalence of AN is observed due to rising number of obese and diabetic patients[1]. AN is not a disease but a manifestation (Symptom) of another condition. The goal of treatment is to correct the underlying cause. But specific treatment options can be considered for cosmetic improvement. Obesity associated AN showed significant improvement by changing to healthy life style by means of physical activity and healthy diet, which ultimately led to weight reduction.



Figures: 11 year old boy with AN. Neck & Axilla

Background

The AN was first described by two Germans, Unna and Pollitzer, in 1889. In 1976 Kahn et al published their study, describing the association between AN and insulin resistance. In 2000, the American Diabetes Association accepted AN as a risk factor for the develo -pment childhood diabetes.

Epidemiology

The exact incidence of AN is unknown and there is no sex predilection to its manifestation. The prevalence of AN varies largely according to age, race and degree of associated condition such as obesity and endocrinopathy. As the obesity and diabetes are on the rise worldwide, there is rise in the prevalence of AN also[2]. Even though AN is seen in all races, it's more common in people with darker skin.

In an unselected population of 1412 children, the changes of AN were present in 7.1%. AN can occur at any age, but malignant type is usually seen above 40 years. Malignant AN is less common. In one study only 2 of 12 000 cancer patients had Acanthosis Nigricans.

AN is more frequently associated with adenocarcinoma of gastro intestinal tract (70-90%) than other malignancies.

Causes and classification

AN can be classified broadly as benign (such as obesity related, syndromic, drug induced etc.) and malignancy associated AN. Some types of AN are described below.

1. ***Obesity associated AN***

   It is the most common type of AN. It is also the most common dermatologic manifestation of obesity and insulin resistance and labeled as pseudo acanthosis nigricans[2].

   Degrees of lesions are positively related to the weight of the individual. Therefore weight

reduction seems to have a good response in regression of the lesion. Although Insulin resistance is often present in these patients, it is not a universal finding[2].

2. ***Syndromic AN***

Many syndromes can be associated with AN. Type A syndrome and type B syndrome are special examples.

Type A syndrome

HAIR-AN syndrome – presents with hyperadro -genaemia, insulin resistance and AN. This syndrome is familial and specifically affecting young women. It may be associated with polycystic ovaries. High level of plasma testosterone is a common feature.

Type B syndrome

It occurs in women with uncontrolled DM, ovarian hyperadrogenesis or an autoimmune disease such as SLE, scleroderma, sjogren syndrome. Antibodies to the insulin receptor may be present.

3. ***Drug – induced AN***

AN may be induced by several medications but it is uncommon. Drugs include nicotinic acid, insulin, pituitary extract, systemic corticosteroids, oestrogen, Oral contraceptive pills, triazinate, fucidic acid, diethyl stilbestrol and melanocyte stimulating hormone preparation. Nicotinic acid is most common association recognized. Lesions usually regress following discontinuation of the offending medication.

4. ***Acral AN***

This occurs in otherwise healthy patients, most commonly in dark-skinned individuals (African American or sub – Saharan descent). The lesions are more prominent over the dorsal aspect of the hand and feet with knuckle hyperpigmentation.

5. ***Unilateral AN***

It is a rare form and also named as nevoid AN. It is inherited as an autosomal dominant trait. Unilateral nevoid AN is not related to endocrinopathy.

6. ***Familial AN***

It is rare a genodermatosis and transmitted in an autosomal dominant manner. The lesions appear during early childhood and progress until puberty. After that it stabilizes or regresses.

7. ***Autoimmune AN***

It occurs due to development of circulating antibodies to insulin receptors in autoimmune diseases such as SLE.

8. ***Malignant AN***

In contrast to AN associated with endocrine dysfunction, malignant AN is usually rapid in onset and associated with weight loss, where the former is more insidious in onset and patients are often obese[3].

AN has been reported with many types of cancers, but the most common underlying malignancy is adenocarcinoma of gastrointestinal tract, especially a gastric adenocarcinoma. In a study of 94 cases of malignant AN, 61% were secondary to gastric neoplasm. Manifestation of Malignant AN can either precede, occur simultaneously or follow the diagnosis of underlying cancer[3].

9. ***Mixed type***

A patient with one of the above types of AN may develop a new lesion of a different etiology.

Pathophysiology

AN manifests due to stimulation of epidermal keratinocytes and dermal fibroblast proliferation.

Benign AN

Insulin crosses dermoepidermal junction to reach keratinocytes at higher concentrations and binds with IGF-1 receptors promoting proliferation of keratinocytes and fibroblasts. These new cells contain more melanin.

High insulin concentrations also indirectly increase the free IGF-1 levels. IGF-1 activity is regulated by insulin/insulin-like growth factor binding proteins (IGF BP). IGFBP-1 & IGFBP-2, which decrease in obese patients with hyperinsulinaemia. This leads to increase level of IGF-1, which stimulate the cell growth and differentiation. Perspiration or friction may also have contributory effect, because AN is seen commonly in body folds where sweating is high[1].

Malignant AN

The stimulating factor for AN is secreted either directly by tumour or in response to the tumour. Transforming growth factor α (TGF α) is one of the growth factors secreted by tumours. It is structurally similar to epidermal growth factor (EGF). When TGF α in excess binds with EGF receptors, there will be excess growth of epidermal tissues leading to AN.

Presentation / Clinical features

Patients usually present with asymptomatic dark and thick areas of skin especially over skin folds. Initially skin gets grey-brown / grey-black pigmentation with dryness and roughness that is palpably thickened. Skin tags (Acro-chordons) may develop at the latter part and they often found in and around the affected areas. The lesions are symmetrically distributed and any part of the body can get affected. Neck is the most common site affected than axillae in children. Occasionally lesions of AN may present on mucous membranes of the oral cavity, nose, pharynx and oesophagus. AN associated with endocrine dysfunction is more insidious in onset, less wide spread and the patients are often obese.

Malignancy associated AN is usually rapid in onset and may be accompanied by skin tags, multiple seborrheic keratosis or tripe palms and weight loss is common.

In malignant AN one third of patients can present with skin changes before any symptoms or signs of cancer. One third may present with AN & malignancy simultaneously and the remaining one third may develop AN after diagnosis of cancer.

Except for the rapid onset the lesions of malignant AN are clinically indistinguisible from a benign one.

Differential diagnosis

Following conditions can mimic AN,

- Confluent and reticulated papillomatosis
- Intertriginous granular parakeratosis
- Dowling-Degos disease
- Atopic dermatitis
- Melanocytic nevi (Giant)
- Ichthyosis hystrix
- Becker Melanosis

Diagnosis

Diagnosis is mainly clinical. Histopathological confirmation is only needed in difficult or doubtful cases.

Patients with AN especially benign childhood AN, are at risk of obesity, hypertension, hyperinsulinaemia, insulin resistance and type 2 diabetes mellitus. Other investigations, such as Fasting Blood Sugar, insulin level, lipid profile[4], Ultrasound Scan of the abdomen are needed to diagnose and monitor the underlying conditions. Any suspicion of underlying malignancy needs targeted investigations for confirmation or exclusion. Association of AN with malignancy might help for early detection of underlying GIT cancer and further it can also help to monitor recurrence and metastasis[5].

Treatment

The goal of therapy is to correct the underlying disease condition. Treatment of the underlying condition may restore some of the normal colour and texture of the affected skin[6].

Targeted treatment of AN itself is only a cosmetic purpose.

Topical medications.

- Keratolytics
  - Topical tretinoin, ammonium lactate or combination.
  - Triple combination of tretinoin, hydroquinone and fluocinalone acetonide nocte and sun cream during day time.
- Calcipotriol, urea & salicylic acid Oral agents.
- Ectretinate, isotrtinoin, metformin & rosigli tazone.

Dermablasion and long-pulsed alexandrite laser therapy may also be used to reduce the bulk of the lesion[8].

Surgical removal of the tumour is the mainstay of treatment for malignant AN if possible.

Based on underlying etiology, multi-disciplinary evaluation may be needed, involving,

1. Primary care physician
2. Endocrinologist
3. Dermatologist
4. Oncologist
5. Clinical nutritionist

Conclusion

The benign type of AN is more common than malignant type. AN is the most common dermatological manifestation of obesity and insulin resistance. As the obesity and insulin resistance are in the rise, prevalence of AN also going up. Because of its relation to obesity and insulin resistance, it can be used as a positive predictor to identify future diabetic patients. With proper intervention on these population (such as weight reduction and glycaemic control by the means of low calorie diet and increase physical activity), we can prevent or postponed the onset of diabetes mellitus. Weight loss may cause the AN to regress almost completely.

Patients as well as health care givers should be educated that AN is not a skin disease but rather a sign of an underlying problem which may have more serious consequences than the symptom per se.

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IRON DEFICIENCY ANAEMIA IN PREGNANCY

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INTRODUCTION

Iron deficiency (ID) is the commonest nutritional deficiency among pregnant women worldwide. It affects more than 2 billion people worldwide. It is also acommon medical condition seen in everyday clinical practice. Iron-deficiency anemia (IDA) remains the top cause of anemia. Therefore the diagnosis and treatment of IDA should be improved.[1]

The WHO estimates IDA to affect approximately 42% of pregnant women and it has been found to be commoner in the non-industrialised countries mainly from Asia.A reliable estimate of the prevalence of IDA in Sri Lanka is unknown .However currently it is estimated that 29% of the pregnant population are anemic.

Several cellular functions such as respiration, energy prod -uction,DNAsynthesis and cell proliferation require iron.[2]

There is no excretory way to iron from the body and excess iron is toxic to human body. Therefore the human body has evolved to conserveiron in several ways, including the recycling of iron after the breakdown of redcells and the retention of iron in the absence of an excretion mechanism. However,since excess levels of iron can be toxic, its absorption is limited to 1 to 2 mgdaily, and most of the iron required daily (about 25 mg per day) is met byrecycling of macrophages that phagocytose senescent erythrocytes. The latter twomechanisms of recycling and absorption, are controlled by the hormone hepcidin.It maintains iron within normal ranges, avoiding both iron deficiency and excess.

Iron deficiency refers to the reduction of iron stores that precedes overt irondeficiencyanemia or persists without progression.Iron-deficiency anemia is amore severe condition in which low levels of iron are associated with anemia andthe presence of microcytic hypochromic red cells.When the delivery of iron to erythroidprecursors is defective it is called Iron-restricted erythropoiesis

IRON METABOLISM

Distribution of Iron

In the balanced state, 1 to 2 mg of iron is absorbed andlost fromthe body each day. Most of the iron in the body is incorporated into hemoglobin in erythroid precursors and mature red cells. Each erythrocyte contains a billion atoms of iron; at normal rates of turnover this concentration corresponds to the incorporation of 2×10^{20} atoms of iron per day.[3]

Consequently, anemia is the cardinal sign of iron deficiency. An approximately 10 to 15 percent of the iron is present in muscle fibers in the form of myoglobin and rest incorporated with enzymes and cytochromes. Further,Iron is also stored in parenchymal cells of the liver and reticuloendothelial macrophages. The dietary iron is readily taken upby the liver when it exceeds the binding capacity of plasma transferrin.

The reticuloendothelial macrophages breaks haemoglobin from senescent erythrocytes and reload ferric iron into transferrin for delivery to cells. This process is indispensable; the erythron alone has a daily requirement of about 20 mg of iron, but only 1 to 2 mg of iron normally enters the body each day through the intestine.[4]

Iron Absorption

All iron absorption occur gastro duodenaljunction. The low pH of gastric effluent helps dissolve ingested iron and provides a proton-rich milieu. This facilitates enzymatic reduction of ferric iron to its ferrous form by a brush borderferrireductase. Therefore enteric coated iron should not be used as it is less efficiently absorbed.[5]

Iron Transport across the Intestinal Epithelium

Iron must cross apical membranes (DMT1) via apical transporter and basal membrane via basolateral transporter. Each transmembrane transporter is coupled to an enzyme that changes the oxidation state of iron. The basolateral transporter requires hephaestin, a ceruloplasmin-like molecule, for the transfer of iron to the plasma.The hephaestin is believed to be a form of ferroxidase. Iron within enterocytes is stored as ferritin. Divalent metal transporter 1 (DMT1) is a protein that

However, no mutations in the gene encoding DMT1 have been found in humans.[37]Such patients couldhave defects in other iron-transport steps.

CLINICAL EFFECTS OF IRON DEFICIENCY IN PREGNANCY

Iron deficiency anaemia has major effects on maternal morbidity and mortality, fetal and infant development and pregnancy outcomes.

Maternal morbidity and mortality

Iron deficiency impairs immune function with increased susceptibility or severity of infections especially puerperal sepsis poor work capacity and performance and disturbances of postpartum cognition and emotions There are evidences that when haemoglobin level is 8.9 g/dl maternal death risk doubles.[38]

Iron deficiency also has effects on pregnancy outcome. It is associated with preterm delivery,[39]low birth weight [40], possibly placental abruption and increased postpartum blood loss.[41]However more study isrequired to ascertaina causal relationship with pregnancy and fetal outcomes.

Effects on the fetus and infant.

The fetus is relatively protected from the effects of iron deficiency by upregulation of placental iron transport proteins.[42] However iron deficiency in pregnancy increases the risk of iron deficiency in the first 3 months of life.[43]Iron deficiency anaemia can cause impaired psychomotor and/ or mental development,negatively contribute to infant and social emotional behaviour[44]and have an association with adult onset diseases.[45]

DIAGNOSIS OF IRON DEFICIENCY ANAEMIA

Laboratory tests

Full blood count, Blood film for picture

A full blood count report display Haemoglobin, mean cell volume (MCV), mean cell haemoglobin (MCH), and mean cell haemoglobin concentration (MCHC).The Haemoglobin level shows degree of anaemia.Further, a blood film may confirm presence of microcytic hypochromic red cells and characteristic 'pencil cells'. However, microcytic, hypochromic indices may also occur in haemoglobinopathies. In addition, for milder cases of iron deficiency, the MCV may not have fallen below the normal range. Other tests either assess iron stores or the adequacy of iron supply to the tissues.

Serum ferritin

When iron store is depleted, it is reflected by low level of serum ferritin .It is not influenced by recent iron intake. In pregnancy due to expansion of plasma volume and mobilisation of iron from the store for increased erythropoiesis the serum ferritin level changes. Its concentration initially rises, followed by a progressive fall by 32 weeks to about 50% pre-pregnancy levels. The levels increase again mildly in the third trimester (Asif et al, 2007). However,it is generally considered the best test to assess iron-deficiency in pregnancy.

Serum ferritin concentration cut-off point of 30 mg/l is indicates low iron storage (van den Broek et al, 1998) and below 15 mg/l indicates iron depletion in all stages of pregnancy as defined by no stainable bone marrow iron (Hallberg et al, 1993). As it is an acute phase protein, concurrent inflammation can alter the serum ferritin level (Van den Broek et al found that). Therefore concurrent measurement of the C-reactive protein (CRP) may be helpful in interpreting higher levels. The CRP concentration seems to be independent of pregnancy and gestational age, although some studies describe a mild increase.

Serum Iron (Fe) and total iron binding capacity (TIBC)

Recent intake of Fe, infection can alter the level of Serum Fe. An increased total iron-binding capacity is specific for iron deficiency, but it is lowered by inflammation, aging, and poor nutrition, its sensitivity specificity is low.

Zinc protoporphyrin (ZPP)

ZPP increases when iron availability decreases, as zinc, instead of iron, is incorporated into the protoporphyrin ring. This gives an indication of availability of iron to the tissues. Red blood cell ZPP has greater sensitivity and specificity for iron depletion (Schifman et al, 1989) but is rarely performed.

Soluble transferrin receptor (sTfR)

Measurement of sTfR is a sensitive measure of tissue iron supply. (Choi et al, 2000). The transferrin

receptor is a transmembrane protein that transports iron into the cell. It is shed into the circulation from the cell membrane. Therefore the circulating level gives an accurate estimate of iron deficiency. When iron deficiency is established, the sTfR concentration increases in direct proportion to total transferrin receptor concentration. There is little data on its use in pregnancy and expensive test, therefore not routinely performed in clinical practice.

Reticulocyte haemoglobin content and reticulocytes

Both reticulocyte number and reticulocyte haemoglobin concentration are reduced in Iron deficiency. Measurements of reticulocyte showerythropoietic activity in anaemia. But it is not widely available, and there is no data on its usage in pregnancy.

Bone Marrow iron

It is the gold standard for assessment of iron stores. But this is an invasive investigation. As other simple tests are available, it is not widely practice.

Trial of Iron therapy

A trial of oral iron could be used as the first line diagnostic test for normocytic or microcytic anaemia especially in low resource setting. When there is an increase in haemoglobin concentration in 2 weeks iron therapy can be continued otherwise further tests are needed.[26,46,47]

Treatment of iron deficiency anaemia

IDA is the commonest form anaemia in Sri Lanka. Therefore the trial of therapeutic oral iron can be undertaken when anaemia is detected at the field level, and if the Haemoglobin improves, the treatment can be continued longer.The reticulocyte count should rise in 1 week, and the haemoglobin level starts rising by the second week of therapy. The rate of rise of haemoglobin is one g/dl/week. So therapy must be continued until haemoglobin level becomes normal and continued until iron stores are replete.

It takes several months (Three months) of replacement therapy to replenish body iron stores. Therapeutic dose of 100-200mg of elemental iron is needed daily.[22]

The corresponding amounts of about 60 mg of elemental iron are available in 300 mg of ferrous sulphate or 180 mg of ferrous fumarate or 500 mg of ferrous gluconate. There are several oral iron preparations are available in the market. However no one compound appears to be superior to another.[48]

Traditionally, oral ferrous sulfatetwice or thrice a day has been prescribed for the treatment of iron deficiency. This treatment used by the 19th-century French physician Blaud and still considered as safe and effective.[49]

At the same time, it is to note that several trials suggest that lower doses of iron, such as 15 to 20 mg of elemental iron daily, can be as effective as higher doses and have fewer side effects.

WHO Recommendation:

Intermittent oral iron and folic acid supplementation with 120 mg of elemental iron a and 2800 µg (2.8 mg) of folic acid once weekly is recommended for pregnant women to improve maternal and neonatal outcomes if daily iron is not acceptable due to side effects, and in populations with an anaemia prevalence among pregnant women of less than 20%.[50,51]

The reason may be that enterocyte iron absorption appears to be saturable; one dose of iron can block absorption of further doses.[52]When iron is taken with meat protein iron absorption can be increased.[53]

Calcium and fiber can decrease iron absorption, but this can be overcome by taking vitamin C.Therefore a calcium supplement during antenatal period is not to be taken with iron tablets and Vitamin C is additionally prescribed to enhance iron absorption.[54]Tea can reduce iron absorption by 90%.[55]Coffee may also decrease iron absorption but not to the degree that tea does. Iron from heme sources is better absorbed 10 times as high as that of iron from nonheme sources.

Several factors may be contribute if patients who do not have a response to oral iron. First,poor compliance due to gastric side effects. Second, in a patient who has ongoing bleeding (e.g. Hook worm infestation in our country), the iron loss may be too great for oral iron to overcome. Finally, absorption of iron may be decreased owing to celiac disease or bowel surgery.

PARENTERAL IRON THERAPY

It is considered if patients who cannot tolerate, patients with malabsorption, severe iron deficiency who are not compliant with oral treatment,those with profound iron deficits may benefit from parenteral iron therapy.

Parenteral iron is preferred in patients with IDA in advanced pregnancy closer to the delivery. Parenteral iron is used in the treatment of postpartum anaemia, and such treatment reduces the need for postpartum blood transfusion, which is a more costly intervention with higher risks.[56]

Although parenteral iron has the ability to replenish the depleted iron stores more rapidly, the rate of improving the haemoglobin status is similar to that with oral iron. Since overtreatment with iron can cause harm by toxicity, ID should be confirmed prior to parenteral iron therapy and the correct dose calculated, considering the patient's body weight and the iron deficit. Chronic liver disease and ongoing systemic infections are considered contraindications and parenteral iron is better to be avoided in the first trimester.

Different preparations are available including, Iron hydroxide dextran complex, Iron sucrose complex,Iron isomaltoside and Iron carboxymaltose.

Iron hydroxide dextran complex (Iron dextran)

Iron dextran isadministered intravenously. A test dose is given before the replacement dose. It is generally safe and effective but there are rare cases of anaphylaxis. Although the manufacturer recommends administering a maximum of 100 mg per day, most clinicians practicegiving an entire replacement dose at one time.[57]Intravenous iron dextran is taken up rapidly by reticuloendothelial macrophages, where it can be processed and loaded to transferrin without toxic effects. The correct dose can easily be calculated with the use of a calculator provided by the manufacturer on the Internet. (http://www.infed.com/calcltor.htm).

Iron sucrose complex

Iron sucrose (Venofer) has lower reaction rates, but they need frequent infusions to fully replete iron stores. It is available in Srilanka

Lowmolecular-weight iron dextran (INFeD)

Low-molecular-weight iron dextran may be the least inexpensive and most convenient option. It is associated with lower reaction rate but allows for higher doses of iron replacement — up to 1000 mg — in a single session.[58,59]

Ferumoxytol (Feraheme) is a superparamagnetic iron oxide coated with carbohydrate has unique complication - severe hypotension which was found in 1.9% of patients.[60]

BLOOD TRANSFUSION

Blood transfusion is less often used to correct IDA in current obstetric practice. It is considered in severe degrees of anaemia (aHb<7mg/l), or when there are signs of decompensation. Blood transfusion would be a better option when woman presents with anaemia close to the delivery especially when it is severe degree.

Excessiveerythropoiesis will ensue following blood transfusion when destruction of transfused blood cells occurs. So receive therapeutic doses of iron should be continued after blood transfusion.

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given within first 24 hours of onset of pulmonary symptoms[3]. Our patient also managed with intravenous methyl prednisolone followed by oral prednisolone.

Another study evaluated the efficacy of cyclophosphamide and plasma exchange in patients with leptospiral pulmonary hemorrhage. Author's findings showed that plasma exchange with immunosuppression improved survival in patients of pulmonary alveolar haemorrhage due to leptospirosis[3].

They recommended the timely initiation of mechanical ventilation with PEEP and high concentration of inspired oxygen for respiratory failure. Extra corporeal membrane oxygenation (ECMO) for resistant hypoxemia in spite of maximal ventilation in adults is not well validated[3].

Conclusion

Most of the leptospirosis patients recover uneventfully. Supportive therapy requires correction of hypotension, hypovolaemia and electrolyte abnormalities. Dialysis and transfusion of blood and blood products may also be required in patients with acute kidney injury. Use of Methylprednisone has been found to be beneficial only if used within 24 hours of onset of pulmonary symptoms. Plasmapharesis and immunosuppression therapy improve survival in pulmonary haemorrhage. It is recommended to manage respiratory failure with timely initiation of mechanical ventilation with PEEP and high concentration of inspired oxygen.

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STROKE DISABILITY IN PATIENTS WHO ARE ADMITTED TO TEACHING HOSPITAL BATTICALOA - AN INTERIM REPORT

Arulmoly K, Jayasinghe VP, Firnaz BM, Mythreye T, Samarasinghe SS, Vinothini K

Abstract

Introduction

Cerebrovascular accident (CVA / Stroke) is a major cause of mortality as well as disability worldwide. Loss of productivity owing to stroke disability is a major concern for a developing country like Sri Lanka. We hope to assess the level of disability in patients admitted to medical wards in Teaching Hospital Batticaloa, at presentation as well as at 1 month and 6-month intervals using the modified Ranking Scale (mRS) as the study tool.

Results and Discussion

We analyzed 61 patients with CVA, including infarction (n=50) and intra – cerebral hemorrhage (n=11) (ICH). The socio demographic data was obtained and analyzed. We were able to demonstrate that there is some degree of improvement of disability with time following ischemic stroke, where as there much less improvement following intra cranial hemorrhage.

Introduction

Cerebrovascular accident (stroke) is an important cause of mortality and morbidity in many developing countries. It is the second most common cause of death after heart attack in Sri Lanka as well as in the world[1]. There are two types of stroke, ischemic and hemorrhagic. Ischemic stroke accounts for about 87 percent of all cases, which occurs as a result of an obstruction within a blood vessel supplying blood to the brain. Hemorrhagic stroke accounts for about 13 percent of stroke cases.

The loss of productivity caused by the disability and the cost involved in the management of stroke could be significant enough to have an impact on the economy ofacountry[1]. About 3% to 5% of the Sri Lankan population is affected by strokes and a significant proportion is at risk. In Sri Lanka, 30% to 45% of males are prone to strokes[4] because of the bad habit of smoking. Those who have high blood pressure, high cholesterol and diabetes are at a risk of stroke if they don't control their comorbidities. Stroke victims'10% die within the first month, 50% are severely disabled and 30% return back to normal after making changes in their lifestyle. Each year one in 110 people in Sri Lanka will suffer a stroke[4]. Even though significant number of stroke patients being admitted to Teaching Hospital Batticaloa per year, still there were no studies have been done to assess the outcome of stroke patients in Batticaloa District. Although definitive treatment of ischemic stroke is thrombolysis, it is practiced in few hospitals in Sri Lanka and which has very limited criteria for implementation, ultimately most of the stroke patients are managed conservatively in which the family support and the physiotherapy plays the major role. Therefore, we would like to conduct a research to assess the outcome of stroke patients by which we can improve the quality of care and life of the patient. In addition, we would like to improve knowledge, attitude and practice of stroke and rehabilitation among healthcare workers and general population.

Objectives:

General objective:

- To assess the disability of stroke patients who are admitted to ward 21&22 of Teaching Hospital Batticaloa.

Specific Objectives:

- To assess the disability on admission immediately after the stroke.
- To assess the disability of the patient one month after the stroke.
- To assess the disability of the patient six months after stroke.
- To assess the stroke concurrence with the premorbid and comorbid conditions such as hypertension, dyslipidemia, diabetes mellitus and smoking.

Methodology:

1.1. *Study design:Qualitative descriptive study*

- Study setting: Ward 21 and 22 of Teaching Hospital Batticaloa.

1.2.. *Study population:*

- The patients admitted to ward 21 & 22 with stroke (limb weakness or any neurological deficit with CT evidence of ischemic or hemorrhagic stroke or with normal CT brain.)

1.3. *Sampling:*

- Convenient sampling method was adapted.
- The number of patients who are going to get admitted to the wards 21 & 22 with stroke for a period of six months.

Inclusion criteria: -Patients admitted to ward 21 & 22 with stroke(limb weakness or any neurological deficit with CT evidence of ischemic or hemorrhagic stroke or with normal CT brain.)

Exclusion criteria: - Patients admitted with limb weakness or any neurological deficit with NCCT brain evidence of other pathologies.

1.4. *Study instruments:*

- MODIFIED RANKIN SCALE
- Interviewer administered questionnaire

1.5. *Data collection:*

- The investigatorsexplained in detail to the patients about the purpose of the study and the consent wastaken. Data collection was done using an Interviewer administered questionnaire to get the details of each patient.
- Thereafter MRS was administered to assess initially the disability of the patient on admission. It was again applied to assess the disability of the patient one-month after admitted with stroke. Again the MRS will be applied assess the disability of the patient six months after admitted with stroke to the ward.

1.6. *Data analysis*

- All statistical analysiswas carried out using SPSS version 16.0 (2007, SPSS for Windows, SPSS Inc., Chicago, IL, USA) package. The results were expressed as mean ±1 standard deviation (SD) and the p value of less than 0.05 considered significant.

1.7. *Ethical consideration*

- Informed written consent was obtained from all participants after explaining the procedures. Non-invasive measurements were done on all patients, as the assessments are routine investigations.
- Any participant would not be harmed mentally, physically or any other means. The feelings and emotions expressed were kept very confidentially. Unethical handling of individuals would not take place during study. The participant could withdraw any time during the study, if not satisfied. The interview and discussion with the participant would be in their mother tongue.
- Ethical approval was obtained from the Ethics Review Committee of the Faculty of Health Care Sciences – Eastern University

Results

61 patients admitted to wards 21 and 22 of Teaching Hospital Batticaloa were included in the interim analysis.

Table 1. Socio – demographic data and risk factors for CVA

Age	Mean 62.8yrs
Sex	
• Male	54.1%
• Female	45.9%
Ethnicity	
• Sinhalese	1.6%
• Tamil	68.9%
• Muslim	29.5%
Pre – existing Risk Factors	
• Diabetes	41%
• Hypertension	60%
• Dyslipidemia	48%
• Ischemic Heart disease	14.8%
• Atrial Fibrillation	1.6%
• Smoking	32.8%
• Family history of CVA	11.5%
• Past history of CVA	18%

Out of the 61 patients, 82% (n=50) had infarction on the CT scan while 18% (n=11) had Intra – Cranial Hemorrhage.

The modified Ranking Scale (mRS) at presentation and at 1-month duration was assessed and the data was analyzed.

Table 2. Modified Ranking Scale at presentation and at 1 month.

MRS Score	% of patients at presentation	% of patients at 1 month
1	3.3 (n=2)	14.8 (n=9)
2	8.2 (n=5)	18 (n=11)
3	18 (n=11)	32.8 (n=20)
4	52.5 (n=32)	14.8 (n=9)
5	18 (n=11)	1.6 (n=1)
6		18 (n=11)

The level of improvement was calculated by improvement of the mRS score, by at least 1 point. Out of the 50 patients with infarction, 74% (n=37) showed some degree of improvement at 1 month, the mean mRS score at presentation was 3.62 and at 1 month was 2.86, this difference was also statistically significant (t<0.05%).

The improvement was noted primarily in patients with infarction, where as in patients with ICH improvement of symptoms was seen in only 18.2% (n=2). This difference of level of improvement between ischemic stroke and ICH was also statistically significant (p<0.05).

There was only 10% of patients with mRSof equal or less than 2 (n=20) at 1 month.

There was no statistically significant correlation between level of improvement and control of diabetes, hypertension, dyslipidemia and prior use of aspirin.

Discussion

This study was aimed at assessing the stroke disability in conservatively managed stroke patients at 1 and 6 months. Our data showed that there is some degree of improvement over 1 month in patients with ischemic CVAs, defined by increment of mRS score by at least 1 point. There are 8 trials worldwide that used mRS as the measure to assess disability in stroke comparing thrombolysis and conservative management where the favorable outcome was defined as mRS equal or less than 2.[19] In our study mRS of 2 or less was only seen in 10% of patients. This brings us to a conclusion that over the period of 1 month although there was some improvement of disability, there was no overall improvement of disability compared to active management with thrombolysis. The perceived improvement may be due to regaining some of the function of the ischemic penumbra.

Conclusion

This is an interim report of an ongoing study to assess the stroke related disability in patients admitted to a medical ward in Teaching Hospital Batticaloa. This shows that there is some degree of improvement of disability with time following ischemic stroke, where as there much less improvement following intra cranial hemorrhage.

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FACIAL REANIMATION - SURGICAL REHABILITION OF PARALYZED FACE

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Abstract

Facial reanimation is a complex surgical procedure to provide motion to the paralyzed face. The causes of facial nerve palsy consist of congenital and acquired defects. Patients suffering from facial paralysis may have difficulty with smiling, eating, drooling of saliva and inability to close the eye depend on the degree of damage. The treatment options are divided into either static or dynamic procedures. In this case series, four patients with facial paralysis who underwent facial reanimation were presented. Two different surgical procedures were performed. Two patients were treated with gracilis free flap as a one stage procedure whereas other two were treated with rotation of temporalis muscle flap with fascia lata. The objective of this article is to review currently available treatment options for paralyzed face. It is difficult to correct long standing facial paralysis. Modern surgical techniques offer various treatment modalities. However, all techniques have advantages and disadvantages. Facial reanimation with gracilis free flap is better choice but time consuming and need surgical expertise. Temporalis muscle transfer is easy and quick method. The surgery is associated with low morbidity compared to free flap muscle transfer. To improve the final outcome, ancillary procedures can be offered.

INTRODUCTION

Facial reanimation is a complex surgical procedure to provide motion to the paralyzed face.[1, 2, 3]The causes of facial nerve palsy consist of congenital and acquired defects. The congenital defects are Moebius syndrome and Hemifacial microsomia. Acquired defects are tumor resection, Bell's palsy and trauma.[4]Patients suffering from facial paralysis may have difficulty with smiling, eating, complaining of drooling of saliva and inability to close the eye depend on the degree of damage.[5]

The treatment modalities are divided into either static or dynamic procedures.[3, 5]Static procedures are suspensory surgeries that hold in position the corners of the mouth or eyelid to improve closure[4,6] Dynamic procedures involve replacing lost mobility of facial muscles with transplanted and innervated muscle. Patients who undergo dynamic reconstructions not only improve the resting tone of their mouth and face, but can animate voluntarily.[3, 7]

If the diagnosis of facial palsy has been made within two months of insult, treatment option will be primary nerve suture or interpositional nerve graft with upper lid weight.[3, 6]

In the case of late reconstruction; 12-18 months after insult, treatment modalities will be considered as following.

1. When facial musculature is present, reinnervation with cross face nerve graft or hypoglossal facial anastomosis and upper lid weight.
2. When facial musculature is absent, reconstruction with microvascular free muscle transfers or temporalis rotational flap.[3,8]

Four patients with facial palsy who underwent facial reanimation surgery were presented here.

Case series

Age/ Sex	Cause	Lesion	Duration of palsy	Treatment
21F	Congenital	Lower motor neuron (Fig: 1 & 2)	Since birth	Rotation of temporalis muscle + upper eye lid gold implant
26F	Congenital	Upper motor neuron. (Fig: 3 & 4)	Since birth	Rotation of temporalis muscle
29F	Schwan -noma	Lower motor neuron. (Fig: 5 & 6)	2 year	Gracilis free flap
31F	Acoustic neuroma	Lower motor neuron. (Fig: 7 & 8)	1 years	Gracilis free flap

All the cases were done under standard general anesthetic procedures. An electromyography (EMG) study was done for all cases prior to surgery.

Surgical procedure of gracilis free flap transfer:

A preauricular incision was made on the paralyzed side of the face with temporal extension and inferiorly in the neck 2cm below the angle of the mandible. Once parotidomassetric fascia is identified dissection proceeds medially towards anterior border of the parotid. The modiolus and orbicularis oris muscle were identified. Facial vessels identified from the lower border of the mandible along its travel. Masseteric nerve was identified through dissecting the masseter muscle in the forward dirrection. The masseteric nerve is identified 7-11mm anterior to the articular tubercle. The nerve was transected as distally as possible.

Gracilis harvest

Surface marking are medial condyle of tibia inferiorly and superior pubic ramus and symphysis superiorly. Incision was placed 1-2 cm medially parallel to the line which connect above mentioned two points. The skin incision was made and dissection continues through subcutaneous fat towards muscle. The gracilis muscle was identified. The adductor artery arises from either deep femoral or medial circumflex femoral artery was traced. The artery was accompanied with two vena commitants. The anterior branch of obturator nerve was identified 1-2cm superior to the vascular pedicle. The vascular pedicle and obturator nerve were harvested with gracilis muscle.

Microvascular anastomosis of the flap

One end of muscle was connected to the oral commisure whereas other end was attached to temporalis fascia. To maintain satisfactory smile, it was decided to do the overcorrection when fixing the muscle. Under the magnification, vascular anastomosis and neurorraphy were done.

Temporalis muscle transfer in facial reanimation

Preauricular incision was extended superiorly up to temporal line and inferiorly 2cm below the angle of the mandible. Incision deepened to parotido masseteric fascia and dissection proceeds above the fascia towards anterior border of parotid. Temporalis muscle was traced up to its insertion at coronoid process. An intraoral coronoidectomy was done. Temporalis muscle was elevated extra orally and temporalis tendon was prepared. Curved S shape incision over lateral thigh and broad sheet of fascia lata harvested. Fascia was attached to end of temporalis muscle to fix the angle of the mouth.

For the purpose of eye closure, 0.9g gold weight was made as thin plate. Skin incision was made over midportion of the supratarsal fold and appropriate size of pocket created. Gold implant was placed and sutured with tarsal plate.

Discussion

Facial paralysis is affecting the quality of life of individual in varying degree from facial asymmetry and facial distortion to functional problem such as lack of lubrication to the eyes.[4]Main objective of facial reanimation is to restore movements to the paralyzed face. The treatment of recent facial paralysis differs from long standing paralysis. In long standing paralysis, facial muscle become irreversibly fibrotic and it will not receive any neural input. But, in recent facial paralysis which can be treated with neural input as early as possible.[4, 9]

When facial nerve is damaged during surgery or by trauma immediate reconstruction should be considered. The simplest and efficient method of repair is direct neurorrhaphy[10].If considerable amount of nerve is missing; an interpositional nerve graft must be placed.

If the lesion is endocranial, neurorrhaphy is impossible because the lesion is too deep. In these circumstances, it is better to choose a motor nerve as a repair material[11].

The motor nerve sources may be categorized into two groups.

1. Strong stimuli producing nerves.eg: hypoglossal, masseteric.

2. Good quality stimuli producing nerves.eg: facial nerve.

When a person shows emotion, the facial nerve only controls the expression via facial expression muscles. Other motor nerves which are connected to facial nerve must be trained to express an emotion. If the motor nerve is masseteric, patient must clench the teeth while smiling. If it is hypoglossal nerve, patient must move the tongue.[4, 12,13]

Another important functional problem is reduction in corneal lubrication. Blinking is an involuntary movement that ensures correct distribution of tear film. Blinking occur 10-19/minute. One of the prime objectives in facial reanimation is recovery of blinking.[5]

When ipsilateral facial nerve is not available, contralateral facial nerve branches can be utilized as cross-face nerve graft. The drawback of this graft is that they deliver small amount of stimulus which is not adequate to produce facial movement.[4]

The masseteric nerve was first used by Spira in1978 and it was popularized by Toronto group to treat developmental paralyzes in Moebius sequence.[4]The masseteric nerve has several advantages over other nerves such as easy harvesting, less donor site morbidity and providing sufficient stimulus for mimetic movement.[12, 13]

In long standing facial paralysis, it is important to provide neural input as well as muscle transfer.[7]Because in these instances, the mimetic musculature is irreversibly atrophic and fibroadiposed. The muscular atrophy is generally varied in between 18 to 24 months. It can be measured by EMG which shows no muscular fibrillation.[5, 13]It is not technically possible to restore all facial expression muscles in long standing paralysis. Restoration of the eye closure and smile are the two important considerations in this cases.[12]

The techniques that have been introduced to reanimate long standing mid facial paralysis may be divided into two groups.[1, 5, 7]

1. Free- flap muscle transplantations.
2. Rotation of temporalis muscle[6] .

Facial reanimation with rotation of temporalis muscle flap.

This technique was first described by Lexer and Eden in 1911.[5]This procedure involves detaching posterior fibres from temporalis fossa to enable anterior transposition of lower tendon of muscle fibres that is attached to the nasolabial sulcus by sutures.

Facial reanimation with free flap muscle transfer.
There are two types of procedures.

1. One stage procedure
   It has low morbidity and rapid recovery.
2. Two stage procedure

It exhibits at first stage cross face nerve graft followed by muscle transfer. This is popular procedure with maximal neural input with great movement. The drawbacks are longer time to get the results and physical, psychological burdens associated with two surgeries[13].

Conclusion

It is difficult to correct long standing facial paralysis completely. However, modern surgical techniques offer various treatment modalities. All techniques have advantages and disadvantages. Facial reanimation with gracilis free flap is better choice but time consuming and need surgical expertise.Temporalis muscle transfer is easy and quick method. This is better option for patients who are demanding for immediate solution. The surgery is associated with low morbidity compared to free flap transposition. The disadvantages are smaller facial movement and loss of spontaneity during smile. To improve the final outcome, ancillary procedures can be offered.

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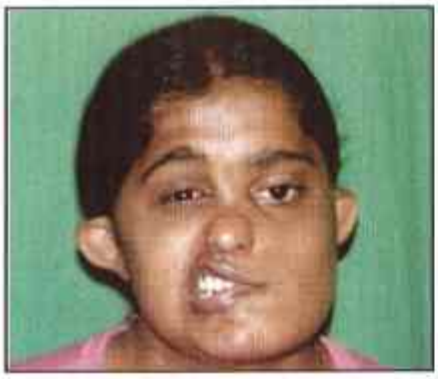

Fig 1: Smile before temporalis muscle transfer

Fig 2: Smile after temporalis muscle transfer

Fig 3: Smile before temporalis muscle transfer

Fig 4: Smile after temporalis muscle transfer

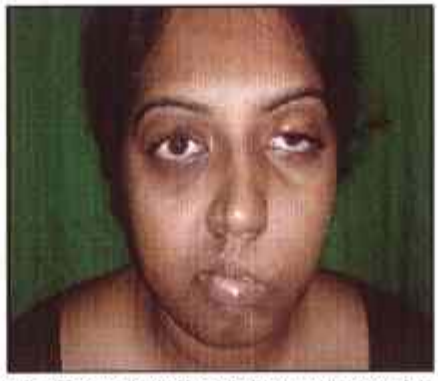

Fig 5: Smile before gracilis muscle transfer

Fig 6: Smile after gracilis muscle transfer

Fig 7 : Smile before gracilis muscle transfer

Fig 8: Smile after gracilis muscle transfer

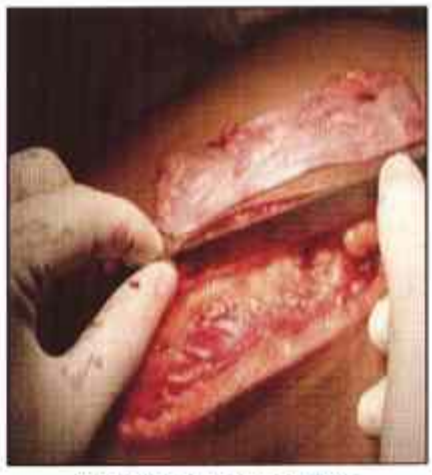

Fig 9 : Harvesting fascia lata

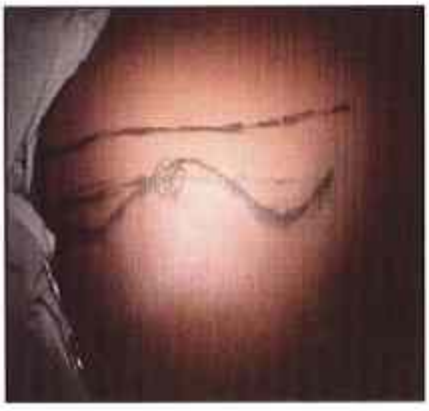

Fig 10:Surface marking of gracilis harvesting

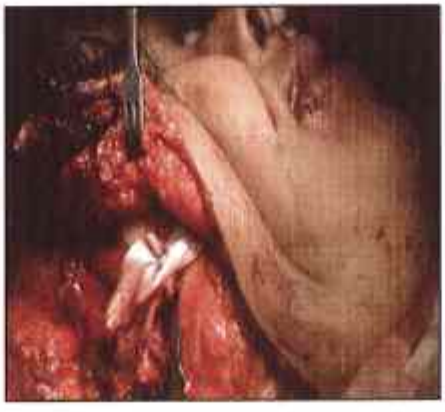

Fig 11:Temporalis muscle attached with fascia lata

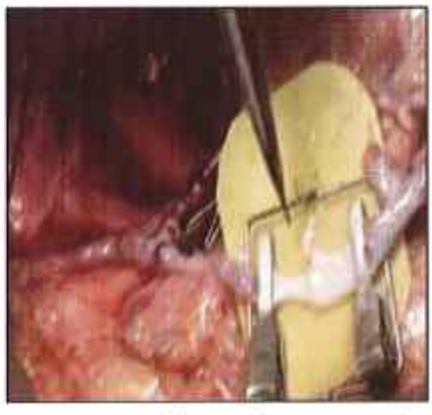

Fig 12: Gracilis vascular anastomosis

ENHANCED RECOVERY AFTER SURGERY FOLLOWING LAPAROSCOPIC ANTERIOR RESECTION AND ABDOMINO-PERINEAL RESECTION

HKGR Anurada[1], R Nirmalasingham[1], WS Ratnaweera[1], V Pinto[2], KB Galketiya[3]

Abstract

Enhanced Recovery After Surgery protocols for colorectal resections involves avoiding mechanical bowel preparation, reduced pre-operative fasting and carbohydrate loading, antibiotic and deep vein thrombosis prophylaxis, avoiding narcotic analgesics, early mobilization and early feeding. The aim of ERAS is early discharge of patients. Laparoscopic resections are expected to facilitate ERAS.

We present twenty four patients who underwent laparoscopic resections for carcinoma of rectum. In them execution of post-operative ERAS protocols were easier allowing early discharge of patients.

Key words-ERAS, Laparoscopic, Colorectal resections

Introduction

Anterior resection and abdomino-perineal resections are the curative resections for carcinoma of the rectum. Anterior resection is possible when the anus and sphincters can be preserved while having a resection margin of 3cm. Surgery may be performed by open or laparoscopic.
For enhanced recovery after surgery (ERAS), protocols were formulated promoting reduced morbidity and early discharge of patients. ERAS involves avoiding mechanical bowel preparation, reduced pre-operative fasting and carbohydrate loading, antibiotic and deep vein thrombosis prophylaxis, avoiding narcotic analgesics, early mobilization and early feeding.(6,7,8)

When the surgery is performed by minimal access it may facilitate to adopt enhanced recovery protocols.(9) This is because with a smaller incision there is less post-operative discomfort which will favor early mobilization. Also with less handling of bowels there will be early return of bowel functions.(1,2,3,4,5)

Objective

To evaluate post-operative recovery following laparoscopic anterior resection and abdomino-perineal resections.

Method

A retrospective analysis of post-operative recovery following laparoscopic anterior resection and abdomino-perineal resections performed from January 2014 to December 2016 was done.

All resections were performed in standard head low and left lateral tilt. Five ports were used. The dissection was with a combination of ultrasonic dissector, bipolar and monopolar diathermy. A high ligation of inferior mesenteric artery was performed in all cases. A total mesorectal rectal resection was performed. In case of anterior resection laparoscopic stapled anastomosis was performed. In three anterior resections where tumour was situated at 4cm from the anal verge a pull through colo-anal anastomosis was done.

The following data were analyzed.

1. Analgesic requirement
2. Mobilization
3. Feeding
4. Complications
5. Discharge from hospital
6. Re admissions within a week of discharge

Results

Thirty two patients underwent the procedure. There were twenty six anterior resections and six abdomino-perineal resections.

On the first post operative day all were managed with epidural analgesia. From second day on wards they were managed with oral diclofenac sodium combined with paracetamol and codeine (panadeine).

All were mobilized out of bed on the first post operative day and started on liquids. Early mobilization was easy as they had minimal pain. Two patients who underwent anterior resection developed mild distension with tenderness in lower abdomen and oral feeding was stopped to recommence on fourth day. In others by the third day semi-solids were begun and normal diet by the fifth day.

One patient after APR developed a wound infection of the perineal wound which settled with suture removal and antibiotics.

Except Five patients all were discharged between fifth to seventh post operative day as shown in the table below.

Post op day of discharge	5	6	7	8	9	10	11
AR	2	6	16	1			1
APR	1	1	1	2		1	

AR-anterior resection APR-abdomino perineal resection
There were no re-admissions within one week from discharge.

Discussion

Minimal access surgery (MAS) will facilitate application of ERAS protocols.(9) This is because MAS is associated with reduced pain allowing early mobilization. (1,2,3,4,5) Non of the patients studied required narcotic analgesics and were mobilized out of bed from first post post operative day. Early mobilization is psychologically satisfying to the patient. It also reduces post-operative complications like chest infections and deep vein thrombosis. With reduced handling of viscera, in MAS, there is less ileus allowing early feeding. Except in two patients, of this study, early feeding was possible.

All of this allows early discharge of patient from hospital. In the case series presented ,27 out of 32 patients (84%) were discharged within seven days from surgery.

Conclusions

Laparoscopic anterior resection and abdomino-perineal resection facilitates enhanced recovery with early discharge of patients.

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MOLECULAR EPIDEMIOLOGY OF THE VIRUSES AND THE SOCIO DEMOGRAPHIC PROFILE OF 1 -24 MONTHS OLD CHILDREN WITH ACUTE BRONCHIOLITIS.

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Abstract

Bronchiolitis is a common cause of viral lower respiratory tract infection (LRTI) in infants. Laboratory diagnosis of viral respiratory infections is generally performed by virus isolation in cell culture and immunofluorescent assays. Reverse transcriptase PCR is now recognized as a sensitive and specific alternative for detection of respiratory RNA viruses. A real-time PCR assay was developed for human respiratory syncytial virus (RSV), parainfluenza virus(PIV), HMPV And H1N1 to test the pathogens. A total of 30 respiratory samples taken over a 1-year period were analyzed by the real-time assay.

The incidence of respiratory viruses detected in these samples was 2 of 30 (7%) by real-time PCR. There were two RSV A viruses detected in 30 samples tested. But the other viruses HMPV, PIV, H1N1 were not detected in the same samples tested. The application of real-time PCR to clinical samplesincreases the sensitivity for respiratory viral diagnosis. In addition,results can be obtained within 6 h, which increases clinical relevance. Therefore, use of this real-time PCR assay would improve patient management and infection control.

Considering the small sample size, it would be advisable to perform a multidisciplinary survey over the country to obtain sufficient data to generalize the results and to help the health care system make suitable decisions regarding viral infection prevention and control, especially for acute bronchiolitis.

Key Words: Bronchiolitis; respiratory viruses; children; epidemiology

Introduction

Bronchiolitis is one of the major acute, inflammatory, infectious diseases of the lower respiratory tract. It is a distressing, potentially life-threatening respiratory illness that affects young children. It is the most common severe LRTI of infancy. Since its recognition as a clinical entity in the 1940s, our understanding of bronchiolitis has grown with respect to the extent of disease, epidemiology, treatment, and longterm effects. Studies have shown a wide variation in how bronchiolitis
is diagnosed and treated[1]. Young infants, especially those aged<3 months are at risk for a more severe disease [6, 7]. RSVhas been shown to cause more severe disease than other virus; the severity of bronchiolitis can also be affected by multiple viral infections or co-infections with bacteria [8]. About 20 % of infants in the United States (US) get bronchiolitis each year and 2–3 % of these children require hospitalization. Children younger than 1 year of age are almost twice as likely to develop bronchiolitis as children between the ages of 1 and 2 years. About 20% of infants in the United States (US) get bronchiolitis each year and 2–3 % of these children require hospitalization. Children younger than 1year of age are almost twice as likely to develop bronchiolitis as children between the ages of 1 and 2 years. Worldwide, the rate of infection in developed countries is similar to those in theUS.

Currently, there is not enough data to determine bronchiolitis rates in underdeveloped countries, but studies suggest that poor nutrition and sub-standard medical care may contribute to the condition in these areas. In tropical regions, bronchiolitis is more common during the rainy season [19]. Sri Lanka is a small island in the Indian Ocean.Climate of Sri Lanka is tropical in nature and consists ofonly wet and dry seasons [20]. Despite the absence of clearly demarcated seasonal variations, respiratory diseases are a leading cause of hospitalization at all ages (10.4% in 2007).Diseases of the respiratory system are among the five leading causes of death in children less than 5 years in Sri Lanka. In the case of bronchiolitis it is around 7%of total respiratory admissions and 13.2% under 2 years [21, 22].

Bronchiolitis is usually a seasonal viral illness, with peak incidence in winter and is caused by RNA viruses RSV (63%) and other viruses such as parainfluenzaviruses (2%), influenza viruses (5%), rhinovirus, corona virus and DNAvirus such as adenoviruses (7%). Recently identified

viruses such as HMPV (3%), HBoV, also contribute to the disease burden [3, 33, 34]. In general, four methods are used for viral identification: virus culture and serology have been the gold standards for the past two decades, while immunofluorescence assay (IFA) for antigen detection and nucleic acid/ Polymerase chain reaction (PCR) based tests are coming to the fore. Culture and serology are more suitable for use in epidemiologic studies and/or patient follow-up because they take more time to generate appropriate results. In contrast, PCR is faster and significantly more sensitive and is poised to take over the field of virus diagnosis [15]. Currently the availability of molecular methods facilitates and improves the identification of infectious organism. In the last two decades, molecular techniques such as PCR and other types of amplification techniques have made tremendous technical improvement. PCR and its variations are increasingly being used in routine laboratories because they are conceptually simple techniques, rapid, potentially very sensitive, specific and can be automated [16].

There are only limited studies done in the past on molecular epidemiology of viruses causing acute bronchiolitis in the Sri Lankan setup. But none of them have utilized by molecular techniques. The aim of this study is to investigate the molecular epidemiology of the causative viruses in 1-24monthsold children suffering from acute bronchiolitis

3. Materials and Methods

This study was a prospective, cross-sectional survey of patients hospitalized with acute bronchiolitis in pediatric professorial unit of NCTH, Ragama between 1 st of August 2015 to the 31st of December 2015. This study was conducted over a period of 12months with the plan of work. Children with symptoms of acute bronchiolitis infections of suspected viral etiology hospitalized in the pediatric professorial department were chosen for collection of data and NPA samples. 30 children with acute bronchiolitis who were admitted to the pediatric professorial unit of CNTH, Ragama were selected as the source of convenient samplesfor this study after considering the inclusion and exclusion criteria. Before implementing the study, a detailed explanation with demonstration was given to the nursing officers who were involved in collecting NPA samples and a case recorded form was constructed and the data was collected in each case. Children aged from 1 month to 2 years of both sexes admitted to ward consecutively with symptoms of bronchiolitis diagnosed according to the SIGN (Scottish Intercollegiate guidelines Network).

Ethical approval to conduct this study was obtained from the Ethical Review Committee of the Faculty of Medicine, University of Kelaniya. The approval for this study was also obtained from the head of the professorial pediatric unit participating in the study. Before collecting patient data and respiratory specimen, children parents or guardians were asked to give their written consent to include the children into the study.

Viruses

Reference control RNA for RSV, PIV, H1N1used for optimizing molecular assays were obtained from National Influenza Centre, Department of Virology, Medical Research Institute, Colombo.

The range of cycle thresholds(Ct)for positive control samples for RSV was 30-35 cycles, 35-40 cycles for PIV, 35-38 cycles for H1N1.

Primers

The following primer sets were selected from previous studies. The referencearticlesmentioned in the table 1. All primers (supplied by Integrated DNA Technologies, IDT, U.S.A.) evaluated with positive control RNA material. Common PCR parameters including thermal cycling parameters, volume of target material and primer concentration were optimized. Unfortunately hMPV positive controls could not be gained from the contacted sources. Clinical samples. From 1st of August 2015 to 31st December 2015, 30 nasopharyngeal aspirates, were received in the laboratory for pcr assay of respiratory viruses.

Table 1: The oligonucleotide primers used for viral PCR

Virus	Primers	Sequence	Target gene	Product	Reference
RSV	RSV-G267-sA RSV-F164-as RSV-G32s RSV-G399-sB PdN6	5' GAT GCA ACA AGC CAG ATC AAG 3' 5' GTT ATG ACA CTG GTA TAC CAA CC 3' 5' GCA ACC ATG TCC AAA CAC AAG 3' 5' AAT ACA AAA TCA GAA ACA CAC C 3' Random primer	G Protein	900bp-RSV-A 760bp-RSV-B 1100bp-Common RSV	[25]
HMPV	hMPV-N-as hMPV-N-fp hMPV-N-rp	5' TGG GAC AAG TGA AAA TGT C 3' 5' GAG TCT CAG TAC ACA ATT AA 3' 5' GCA TTT CCG AGA AGA ACA C 3'	Nucleoprotein Nucleoprotein Nucleoprotein	928 bp	[26]
PIV3	PIV3-1 PIV3-2	5' CCA GGG ATA TAY TAY AAA GGC AAA A 3' 5' CCG GGR CAC CCA GTT GTG 3'	Hemagglutinin-Neuraminidase	83 bp	[67]
Influenza A H_1N_1 pandemic 2009 virus (HA-173)	HKU-SWF HKU-SWR	5' GTGCTGAGCTTTGGGTATGAA 3' 5' AGCTCAGTGTCATCATTTGAA 3'	HA HA	173 bp	[68]

Viral nucleic acid (RNA) extraction

Qiagen viral RNA mini purification kit (cat 52906, Germany) was used for extracting and purifying viral RNA from all the specimens according to manufacturer specifications. Briefly, 140µl of specimens were added to a tube containing 560 µl of lysis buffer with RNA carrier, mixed well by vortex, incubated at room temperature for 10 min; applied all of this solution to the QIAamp mini spin column, centrifuged, washed twice with buffer AW1 and buffer AW2; viral RNA was finally eluted in 60 µl of AVE solution and kept at -20C. The extracted RNA from each specimen was used for all amplification assays for identification of viral etiologies such as RSV, hMPV, PIV3, H1 N1 of acute bronchiolitis.

Storage of RNA

The extracted RNA was stored at -20C in the freezer (Whirlpool of 61 T, Italy) until further processing.

Real time RT-PCR assays specific for each viruses

Two steps real-time RT- PCR was carried out. cDNA synthesis and the real-time PCR was carried out by using SwiftTM Spectrum 48 Real-Time Thermal Cyclers, Singapore.

Respiratory Syncytial Virus

Genus specific PCR for RSV

Genus specific PCR amplification was performed in a final reaction volume of 25.0 µL using the following reaction volumes. The primers used G 32 as the forward and F164 as reverse. 5 µl of 5X PCR buffer, 2.5µl of 25mM MgCl2 ,0.5µl of10mM dNTP, 1µl of forward and reverse primers, 0.25µl of TAQ polymerase (5u/µl),EvaGreenTM Dye 20X(Biotium,C.A,Cat No.31000) 1.25µl and 7.25µl of DEPC water and 5 µl of cDNA were added into a 0.2ml PCR tube An initial step of 95 ºC for two minutes was performed, followed by a total of 45 cycles at 94 ºC for one minute, 65 ºC for 1.5 minute and 72 ºC for 1.5 minute, with final cycle at 72 ºC for seven minutes[65].

Genotype specific Semi nested PCR for RSV strains

The dilutedgenus specific PCR product was used asthe substratefor semi nested PCR and similar reagents volumes applied except the use of different primers.

Semi nested PCR wasconstituted withG267 and F164 primers for RSV A and G399 and F164 for RSV B.

Parainfluenza 3

PCR was conducted in a final reaction volume of 25.0 µL. 5 µl of 5XPCR buffer, 2.5µl of 25mM MgCl2,and 0.5µl of10mM dNTP, 1µl of forward and reverse primers, 0.25µl of TAQ polymerase (5u/µl), Eva Green Dye, 20 x 1.25µl and 7.25µl of DEPC water and 5 µl of cDNA were added into a 0.2ml PCR tube. PCR cycle condition consisted of a 950 C for minutes followed by40 cycles of 950 C for 30 seconds 600 C for 30 seconds, 720 C for 30 seconds followed by 7 minutesat 72ºC[69].

Influenza A H1N1

PCR was conducted in a final reaction volume of 25.0 µL containing the following reagents. . 5 µl of 5X PCR buffer, 2.5µl of 25mM MgCl2, 0.5µl of10mM dNTP, 1µl of forward and reverse primers, 0.25µl of TAQ polymerase (5u/µl), Eva Green 20x1.25µl and 7.25µl of DEPC water and 5 µl of cDNA were added into a 0.2ml PCR tube. PCR cycle condition consisted of950C for 2 minutes followed by 40 cycles of 950C for 30 seconds 560C for 30 seconds, 720C for 45 seconds followed by 7 minutes at 72 ºC[61].

hMPV

It was not possible to obtain reference controls for this virus from anylaboratory in Sri Lanka Therefore,the real time PCR assay was not possible to establish.

Statistical analysis

The Statistical Package for Social Sciences (SPSS) for Windows® release 20.0 (SPSS Inc. - Chicago, IL, USA) was used for the statistical analyses. Values were given as percentages for categorical variables, and as median with range for continuous variables. Categorical variables were compared by using $\chi 2$ test or Fisher's exact test,

Results

Clinical characteristics

The study was carried out in 30 children with clinically diagnosed acute bronchiolitis. The age of the patients ranged from 2 months to 2 years. The mean age was 6.83 months, and the median was 6 months, Male were 17 (57%) and female 13(43%). median duration of hospital stay 3 days (range, 2-5 days) Cough and corysal symptoms were the most experienced symptoms by all children (100%). Rhonchi (96%) and crepitation (72%) are the most detected signs in the clinical examination.80% of the children were needed some sort of observed management while on admission. Nearly half of the children have the family history of atopic diseases. Nebulization was the important treatment modality in the treatment of bronchiolitis (100%). Even the main etiology of the bronchiolitis is viruses, steroids (45%) and antibiotics (59%) were used significantly. Only 3(10%) children with bronchiolitis need O2 support(spO2<92%).

Socio demographic profile

The majority of the patients (86.7%) included in this study were sinhalese which reflect the region majority. The parental education was very good because the 67.6% of the parents educated upto ordinary lavel or more than that according to the country's educational level. Most of the included children in this study were single (1st baby).

With each positive control sample real-time PCR was establi -shed and amplification curves and melting curves were obtained. The highest ct value which was tested in previous studies taken as cut off value for each virus. Beyond this value was taken as negative results. Table 2 [67].

Virus	Highest ct value
RSV	41.56
PIV3	41.12
H_1N_1(Influenza A)	42.51

Table 2: The highest ct value for each virus taken as positive

Beyond this value was taken as negative results. Table 4 [67].

Table 2: The highest ct value for each virus taken as positive

Virus	Highest ct value
RSV	41.56
PIV3	41.12
H_1N_1(Influenza A)	42.51

According to the figure 1 the ct value of RSV A and RSV B was 35.00 and 35.27 respectively.

Of the 30 samples tested, two (02) samples were positive for RSV with the genus specific primers.

The ct value of the positive samples were 36.00 and 36.62 with melting temperature 73°C figure 2.

Positive control ct value of 35.05 and the melting temperature was 73°C

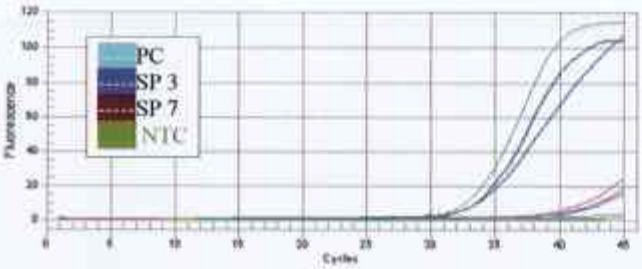

Figure 1 :Real time PCR curve for samples tested with RSV common primers G-32

PC- positive control, SP- sample, NTC-no templatecontrol

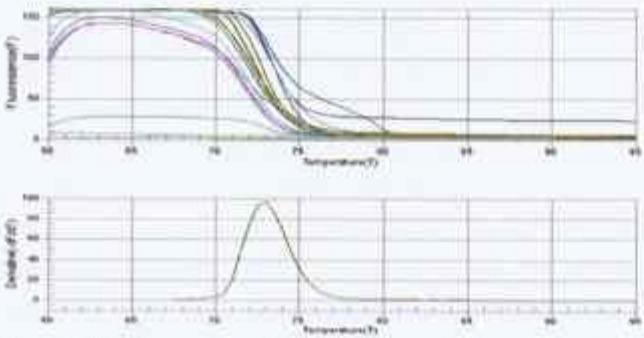

Figure 2: Melting curve analysis of RSV samples

Sub typing of positive samples with RSV was done. Both samples were found positive for RSV A. the ct value of positive control 35.05, and the samples 36.00 and 36.62 (Figure 3)

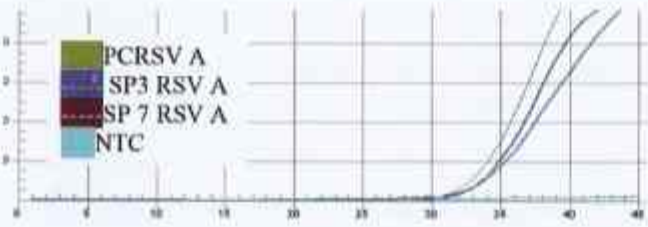

Figure 3: Real time curves for positive samples tested for RSVA subtype

PC- positive control, SP- sample, NTC-no template control

Visualization of real time RT- PCR products of positive controls in gel electrophoresis

The real time RT-PCR products of positive controls of RSV, PIV, and H1N1 were loaded in agarose gel and the following gel picture was obtained(Figure 4).

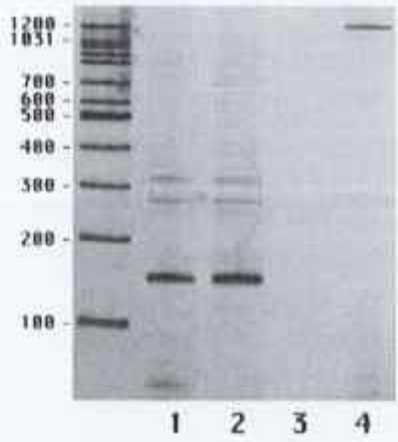

Figure 4: Gel photograph showing the amplification of positive reference controls of the viruses studied. Lane 1: H1N1 (176 bp); Lane 2: H1N1 (176 bp): Lane 3: Parain -fluenza (---- band not visible); Lane 4: RSV genus specific band (1.1 Kb)

Table 3 : Real Time PCR testing – Summary of test results

No of samples tested	RSV		H1N1	Para-influenza	Human metapneumo
	RSV-A	RSV-B			
30	2	0	0	0	Not tested

Discussion

Respiratory viruses have been associated with acute bronchiolitis [1, 2, 3, 4], and detected in various molecular studies on acute bronchiolitis [8, 9, 15]. Recently PCR has been used in laboratory and clinical settings as a diagnostic tool and it is an effective technique to detect genetic material of viruses [16]. PCR is currently used to detect respiratory viruses as it has been recognized to be as sensitive and specific than conventional methods (virus isolation, cell culture and serology) [41]. Hence PCR has increased the ability to detect and quantify individual and multiple viral pathogens in clinical samples [42, 47]. In this study we have used conventional and real time RT- PCR toidentify the respiratory viruses in 1- 24 months old with acute bronchiolitis.

Many factors will affect the performance of the assays in detection of respiratory viruses. Pre analytical factors such as specimen collection, transport, nucleic acid amplification may influence the amplification. Since the all viruses tested is RNA virus that it can degrade easily, and inhibited by inhibitory substances in the samples may reduce the yield (false negative).

In this study the specimens tested by PCR underwent multiple freeze/ thaw cycles, which may have adversely affected assay sensitivity. Only two samples were positive for single virus in this study .The primers selected for this study were very specific for sub types of the viruses such as PIV3, H1N1 rather than PIV or Influenza A. Increasing the sample size might have given more positivity than the current study has detected. We decided to include H1N1 in this project because it was circulating in Colombo and suburbs during that time.

Detection rate is varying for the detection of respiratory viruses by various methods. The detection rate for culture, ELISA, and immunostaining techniques is 25-30% but detection rate for real time PCR has been about 30-40% [41]. The improved detection rate of PCR for specific individual respiratory agents is the greatest advantage in using as a tool for virus identification. Sri Lanka is a tropical country and highest number of respiratory tract infections reported in June [21] or worldwide it is in February. But the sample collection was done during the period of August to December. This may be another reason for less positivity of viruses in our study.

Assuming positivity (Sensitivity) in real-time PCR is depending on the ct value of the sample and melting curve analysis to conform the correct PCR product. Making a standard curve from known positive control by making a serial dilution of concentration of virus is the ideal way to tell which range of ct value will give the positivity of a particular virus. It is impossible to make a serial dilution of concentration without a good reference positive control usually from cell culture. In our study we were unable to get such positive controls. We used the positive samples tested by real time PCR with low concentration of RNA (Table 3) for this study. It was very difficult to achieve a standard curve for each virus. Here in our study the previous study papers were selected to determine the highest ct value for each virus Table 4 [67].

To assess the specificity of the real-time PCR the test should be compared with another test like cell culture, ELISA, and immunostaining techniques. But in our study lack of such facilities and funds to conduct and compare PCR results with other method is another limitation in this study.

Seasonal variations of bronchiolitis and detected viruses

To address relationships between seasonal variations of bronchiolitis and respiratory viruses, we couldn't cover the whole year for the specimen collection. In this study we have got 2 positives of RSV A. RSV is the predominant virus classically associated with bronchiolitis and is responsible for 70%-75% of bronchiolitis, but the prevalence of RSV was found from autumn to winter, while prevalence of HRV was found in all season. In addition, both viruses were detected from autumn to winter. RSV A is considered more common than RSV B worldwide [34].

There are other respiratory viruses like HRV, HBoV also emerging in various parts of the world. Different respiratory pathogens have different traits in response to certain meteorological conditions. But no studies in Sri Lanka to compare the seasonal variation of virus positivity. In tropical regions, children tend to be kept indoors during the rainy season, and the resultant crowding may account for the increased incidence of RSV infection. Another reason that has been suggested is that high humidity may help to prevent the virus from. desiccation and loss of infectivity.

Disease severity of respiratory viral infections

To examine the disease severity in the virus positive and negative groups, clinical and demographic data should be compared. It was unable to compare the severity with statistically significant manner.

But both the children were under 6 months and had severe bronchiolitis who were treated in high dependency unit. The demographic and clinical information is important to put the results into a practical setting and allow them to be applied in clinical practice. In the current study, statistical significance was not achieved for the detection of RSV and its association with clinical severity. The fact

that nearly half of the RSV infections occurred in children under 6 months [1, 6] showed that patients in this age group are the most vulnerable to severe RSV infections.

Co-infection

In bronchiolitis co-infections can occur usually with RSV and HMPV but others also can occur together. There is no direct correlation with viral co-infection on disease severity [9]. In the present study, only RSV A was found. Subgroup B infected children were admitted to the hospital less frequently than subgroup A infected children.

Study limitations

Sample number and sample selection:

Sample number was not calculated according to the formula but it was restricted to 30.Owing to the financial and time constraints it was not possible to extend to that many samples. Because of the small sample and less positivity we could not assess correlation with etiology and clinical significance.

It was not possible to get hMPV RNA or cDNA from other laboratories in the country, therefore the study did not have a reference positive control for hMPV. We were unable to assess the annual prevalence and annual monthly variation in bronchiolitis due to time constraints. Sample collection was done from August to December, laboratory and data analysis were done from September to March. District prevalence and ethnic variation could not be assessed as the study was not conducted in several places. Only four viruses included in this study was due to financial and time constraints.

Conclusion and recommendation

The knowledge gained from this study can use for the detection of virological agents causing acute bronchiolitis. It is noteworthy to include other viruses also to see the pattern of distribution in bronchiolitis. The use of real time RT-PCR in clinical virology practice allows for rapid and accurate detection of conventional and newly discovered viral respiratory pathogens in children. The use of such rapid and reliable molecular tools is needed to provide not only epidemiological and virological data but also the opportunity to understand the emergence of novel influenza virus strains.

Because of the small sample size used in the present study, a multidisciplinary survey should be performed over the country to obtain sufficient data to allow generalization of these results to the population. Confirmation of our findings could help the health care system to make suitable decisions for prevention and control of viral infections, especially in the respiratory tract. Further investigations of the study would include genotyping positive samples from this study and constructing phylogenetic lineage of respiratory viruses in Sri Lanka.

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SOCIODEMOGRAPHIC AND HEALTH CHARACTERISTICS IN CHILDREN WITH LOWER RESPIRATORY TRACT INFECTIONS AT A TERTIARY CARE CENTRE IN SRI LANKA

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Introduction

Acute Lower respiratory tract infections (LRTIs) are one of the most frequent causes of admission to paediatric units leading to high morbidity and mortality in children (1). It is one of the main causes of childhood mortality and morbidity in developing countries (2,3). Several socio demographic factors like poverty and poor household and existence of other known risk factors such as anaemia, malnutrition and congenital heart diseases are associated with LRTIs (4). It causes substantial burden in healthcare system leading to over treatment and increased hospital admissions. We were unable to find much local literature on the above topic after thorough search. Therefore, the aims of this study were to find out sociodemographic and other health chara -cteristics among children with lower respiratory tract infections admitted to a Tertiary Care Centre in Sothern Sri Lanka.

Methods

This was a descriptive cross-sectional study which included children with LRTIs admitted to Paediatric Units at a tertiary care center in Southern Sri Lanka. Study was conducted over the period of 1 January to 31 December in 2013. Data were collected using an interviewer-administered questionnaire and from the hospital records.

The data on sociodemographic background, the presence of known risk factors for LRTIs, and the data on treatment after admission to hospital were gathered. Five social classes (SC) were defined considering parents' educational level, occupation and the monthly family income according to Barker and Hall guidelines (5). Anaemia was defined based on the haemoglobin concentration of 111g/L. Ethical clearance was obtained from local Ethical Review Committee and informed written consent was obtained from the parents and ascent was obtained from the children who were above the age of 8 years. The data were analysed using descriptive statistics and Chi-Square test.

Results

We collected data from 281 children with LRTIs in the age range of one month to 12 years. The duration of hospitalisation varied from one to 16 days. Majority of patients (n=238) were from the same districts and suburb areas where the hospital is situated. Table 1.shows the sociodemographic characteristics and the presence of known risk factors. Five patients (1.8%) had four risk factors, 18(6.4%) had three risk factors, 61(21.7%) had two risk factors and 117(41.6%) had at least one risk factor. Majority of parents were in the social class 3-5.The presence of at least one risk factor was associated with the low socioeconomic category (Pearson Chi-Square = 7.995, df = 1, p = 0.005).

Table 1.Sociodemographic characteristics and presence of known risk factors of LRTIs.

Characteristic	Patient (n=281)
Median age (months)	27
Age < 5 years	206 (73.3 %)
Boys	173
Girls	108
Presence of known risk factors	
Low birth weight (< 2.5 kg)	57(20.2%)
Bronchial asthma	76(27.0%)
Past history of LRTI	26(9.2%)
Preterm birth	12(4.2%)
Anaemia	95(33.8%)
Congenital heart disease	18(6.4%)
Exposure to passive smoking	33(11.7%)
Developmental delays	9 (3.2 %)
Syndromes/other congenital anomalies	5 (1.8 %)
Socioeconomic background	
Social class 1	4 (1.4 %)
Social class 2	32 (11.3 %)
Social class 3	66 (23.4 %)
Social class 4	117 (41.6 %)
Social class 5	62 (22.0 %)

LRTIs = Lower respiratory tract infections

Diagnosis was primarily based on the clinical evidence which was sometime accompanied by the investigations. Serum C-reactive protein was done on 92(32.7%) and it was elevated (more than 6 mg/L) in 57(61.9 %) children. ESR was performed in 47(16.7%) and elevated (ESR> 13mm/ hr) in 35(74.4 %) patients. Blood culture was done and reported as no growth of organism in 67(23.8 %) patients. Full blood count was done in 269(95.7%) and leukocytosis was observed in 158(58.3%). Abnormal findings were revealed in 54 chest X-rays. There were 54(19.0 %) with pneumonia, 11 (3.9 %) with bronchiolitis, 87(30.9 %) with LRTI plus wheezing and 129(45.9 %) with LRTI without wheezing among them. Majority of patients 278(98.9 %) were treated with antibiotics. Total of 13 antibiotics were prescribed among them. Figure 1.shows the distribution of antibiotic use according to the route of administration. Table 2. demonstrates different types of antibiotics used. Among the patients, 47(16.7 %) were given intravenous fluid and oxygen therapy was required by 44 (15.7 %) patients.

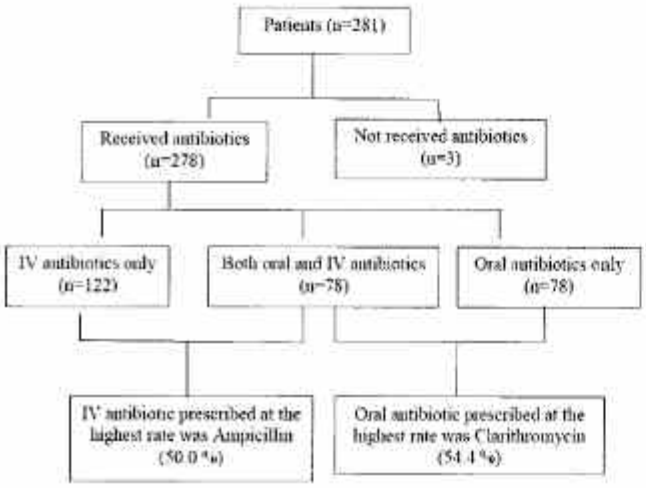

Table 2. Categories of antibiotics used among the patients

Category	Antibiotic	Number (n=278)
Penicillin	IV Ampicillin	100
	Oral Amoxicillin	18
First generation cephalosporin	Oral Cephalexin	1
Second generation cephalosporin	IV Cefuroxime	49
	Oral Cefuroxime	8
Third-generation cephalosporin	IV Cefotaxime	50
	IV Ceftriaxone	7
Carbapenems	IV Meropenem	3
Aminoglycosides	IV Gentamicin	47
Macrolides	IV Amikacin	1
	Oral larithromycin	85
	Oral Erythromycin	17
Other	IV Co-amoxiclavulonicacid	6
	IV Metronidazole	1
	Oral Co-amoxiclavulonicacid	44

Discussion

The proportion of children with known risk factors for LRTIs was high according to our study. Among the known risk factors anaemia, bronchial asthma and low birth weight were reported in highest proportions and these had been identified as risk factors by other studies also (6). Iron deficiency anaemia is known to make the children more susceptible for LRTIs, because of its effects on immune functions (7). Occurrence of LRTIs is multi-factorial which includes personal s and environmental risk factors and most of these are potentially preventable and modifiable (6,8). We found that majority of children were from lower socioeconomic background and lower social categories were associated with the prevalence of risk factors. Recent studies suggested that prevalence of acute respiratory tract infections is higher in low social classes with poor parental literacy and had found a significant association with the social class (9). Furthermore, there are studies that have found out the link between the prevalence of risk factors and the socioeconomic status (10).This could probable be due to various factors such as lack of access to basic health services and public health awareness programmes with the lower socioeconomic background. We found that antibiotic prescription remains very high for these children. The use of old generation antibiotics was less common, while newer broad spectrum drug use had become a preferable option. According to the existing literature, most of these LRTIs and bronchiolitis are due to viral origin, though new broad spectrum antibiotics have been started on majority of patients. However, antibiotics use in these situations neither reduces the complications nor expedite the recovery (11, 12). Exacerbation of asthma is usually associated with viral respiratory tract infections, though most patients have received antibiotics

(13). Guidelines on indications and the type of initial antibiotic which should be started have been laid down by the local authorities, though it had been deviated. Inappropriate use of antibiotics has increased to a considerable degree recently (14). Promotion of available diagnostic algorithms in the institutions, and development of locally-set systems to determine the patients who are likely to get benefitted from antibiotic treatment may reduce the total use of antibiotics. The rational use of antibiotics is important to maintain the quality of healthcare, to reduce the expenditure and to minimise the side effects and the development of antibiotic resistance (15).

Conclusions

Anaemia and bronchial asthma were reported in highest rates among the known risk factors for LRTIs. Majority of the patients were from lower socioeconomic background. The use of antibiotics for lower respiratory tract infections remains high. Further, larger studies are required to elucidate the Sri Lankan status at different settings to confirm the above conclusions.

Limitations

Relatively small sample size and the absence of specific coding on the aetiology are the major limitations of our study.

Conflict of interest

The authors declare that they have no competing interests in this study.

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A RERE PRESENTATION OF GUILLAIN - BARRE SYNDROME : PHARYNGEAL - CERVICAL - BRACHIAL VARIANT

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INTRODUCTION

Guillain-Barre Syndrome is an acute immune mediated polyneuropathy it includes a heterogeneous group of disorders with various subtypes and variants.GBS is clinically characterized by symmetrical, generalized ascending weakness with areflexia due to peripheral nerve involvement. Pharyngeal-cervical-brachial (PCB) variant is a rare form of Guillain–Barré syndrome (GBS) presents with a rapidly progressive oropharyngeal and cervicobrachial weakness with areflexia predominantly of the upper limbs. Lower limbs muscle power is usually preserved or only mildly affected. Here we report a classical case of PCB variant of GBS presenting in a 70 year old.

Case report

A 70-year-old woman known hypertensive on treatment presented with weakness of left upper limb more proximal than distal progressed into weakness of right upper limb and subsequently left lower limb for 4 days duration. Lower limb weakness was less severe compared to the upper limbs. She had difficulty in swallowing for similar duration, with mild respiratory discomfort. There was no preceding diarrhoeal illness but described a respiratory illness one month back.

On examination she was conscious and oriented. Facial power, eye movements and pupillary response were normal. Palatal movements were impaired. There was weakness of neck muscles with a Neck flexion of 3/5. Single breath count was more than 25. Shoulder abduction was 2/5 on the right and 1/5 on the left. Distal upper limb power was 2/5 on the left and 3/5 on the right. Power in the lower limbs was 4/5 .there was global areflexia with no sensory involvement .plantar response was flexor bilaterally and there was no demonstrable fatigability or ataxia.

She was afebrile with a blood pressure of 140/80 and pulse rate of 80 bpm. Cardiovascular, respiratory and abdominal examinations were unremarkable.

Her base line blood investigations were white cell count 12.3x109/1 and platelet count 197x109/l. serum potassium- 4.2 mmolll, sodium -138 mmolll, , fasting blood glucose – 103 mg/dl and creatinine- 57 umol/l. The haemoglobin was 13.9g%, The liver function test was normal. The chest radiograph was unremarkable. Viral panel (EBV,CMV,herpes virus),cultures for salmonella ,shigella and campylobacter jejuni and serological examination for mycoplasma pneumonia were negative. Neurophysiological studies done on day 3 of admission indicated motor conduction abnormalities with conduction blocks and sensory responses were normal suggestive of GBS. Cerebrospinal fluid (CSF) examination on day 08 of illness revealed typical "albumino cytological dissociation" with no white or red blood cells. The protein level was high at 60 mg/dl,with normal glucose. CSF cultures and gram stain were unremarkable.

With clinical and neurophysiological evidence the diagnosis of PCB variant of GBS was made and patient was initiated on intravenous immunoglobulin 2mg/kg over 5 days. Strict monitoring of respiratory functions were assessed with respirometer observing for potential complications. Over the following 10 days with physiotherapy and supportive care she made a remarkable recovery with upper limb power of 4-/5, normal lower limb strength, neck flexion and normal swallowing.

DISCUSSION

Guillan-Barre syndrome is characterized by bilaterally symmetrical ascending paralysis, absent of deep tendon reflexes, sensory loss, cytoalbuminologic dissociation in cerebrospinal fluid and typical findings in nerve conduction studies.

In 1986, Ropper(1) described the first patients who developed rapidly progressive oropharyngeal, neck and shoulder weakness, with relative sparing of the lower limbs in the absence of sensory disturbance and, mimicked

like decending paralysis seen in botulism although relative sparing of the lower limbs were initially thought as the hallmark of the disease some patients were later described to have minimal limb weakness as in our patient.

According to the proposed new criteria for the PCB variant of GBS (3), our patient fulfilled all the features required for the diagnosis she had relatively symmetrical oropharyngeal weakness, neck weakness, arm weakness and arm areflexia. There was absence of ataxia, disturbed consciousness and prominent leg weakness. She had a monophasic illness pattern and interval between onset and nadir of oropharyngeal or arm weakness was around 2 weeks and there was absence of identifiable alternative diagnosis.

In addition to these she had other strongly supportive Features such as, Antecedent infectious symptoms, cerebrospinal fluid analysis showing albumino cytological dissociation, Neuro physiological evidence of neuropathy. GBS is one of the auto immune illness preceded by an infectious illness. Auto antibodies against specific neuronal gangliosides have been implicated in the pathogenesis of different GBS variants. The strongest association for PCB is the presence of IgG anti-GT1a antibodies is thought to be a useful marker in supporting the diagnosis.(2)in our case serological assays were not available in our country.

The presence of additional ophthalmoplegia and ataxia indicates overlap with Fisher syndrome. This is the commonest association with the variant.

Patients with pure PCB were more likely to require intubation than those with overlap syndromes and this correlated with degree of bulbar involvement (2). fortunately our patient didn't need ventilation.

Conclusion

PCB variant of GBS should be remembered in patients with symptoms of bulbar and upper extremity weakness not only for early diagnosis but also to plan the treatment early and follow up the potential complications. Due to the unfamiliarity with PCB variant, clinical picture is often misdiagnosed as brainstem stroke, myasthenia gravis or botulism.

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TROPICAL CHRONIC PANCREATITIS - A CASE REPORT

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Keywords : Chronic Pancreatitis, Tropical Pancreatitis

Abstract

Chronic pancreatitis is a multifactorial disorder, which has pancreatic calcifications as the hallmark of the disease. Tropical pancreatitis is a form of chronic calculus pancreatitis seen predominantly in the tropics. There are large pancreatic head calcifications, which may lead to recurrent obstructive jaundice, and many complications. We present a case of an elderly man presenting with recurrent obstructive jaundice secondary to tropical pancreatitis.

Background

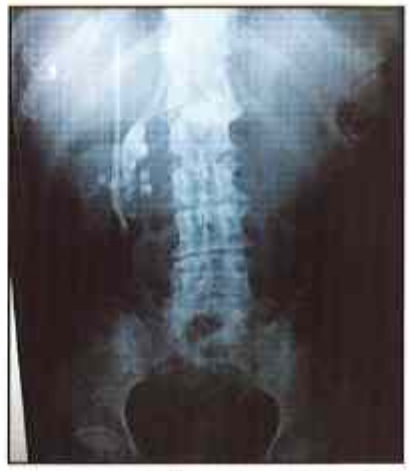
Chronic pancreatitis is a progressive inflammatory and fibrotic disease of the pancreas. An idiopathic chronic pancreatitis occurring predominantly in the young non – alcoholic population in the tropical regions has been reported since 1950s and is termed Tropical Chronic Pancreatitis. [1,2] The hallmarks of the disease are chronic abdominal pain with large pancreatic calculi in and around the head of the pancreas, recurrent obstructive jaundice and pancreatic diabetes mellitus.[3] We report a case of a 69 year old male with painless tropical pancreatitis, who presented with obstructive jaundice secondary to large pancreatic calcifications.

Case Presentation

Mr. R a 69-year-old previously healthy patient presented with obstructive jaundice for 3 weeks duration. He denied having abdominal pain, nausea or vomiting. He was a non – smoker and did not consume alcohol. On examination he was thin and wasted with deep icterus. There was no hepatosplenomegaly or lymphadenopathy.

The liver functions confirmed the obstructive jaundice. The ultrasound scan of the abdomen revealed atrophic and calcified pancreas with dilated main pancreatic duct (MPD) with a calculus in situ. Distal common bile duct (CBD) was also dilated due to a calculus. There were no gall bladder calculi. The X – ray abdomen revealed large calcification around the head of the pancreas. Contrast enhanced CT of abdomen revealed dense pancreatic calcifications in the pancreatic duct as well. There were no biliary strictures and no evidence of any masses or porta – hepatic or para – aortic lymph nodes.The fasting blood glucose was 7.1 mmol/l. Hepatitis serology and ANA was negative.

The patient underwent Endoscopic Retrograde Cholangio Pancreatography (ERCP). Spinchter -otomy was done and pancreatic duct was stented. The symptoms gradually improved following stenting, however patient continues to have recurrent episodes of obstructive jaundice and 6 monthly ERCP with re – stenting is planned. He receives Gliclazide for his diabetic control.



Discussion

Chronic pancreatitis is a progressive inflammatory and fibrotic disease of the pancreas. The aetiology for chronic pancreatitis may be due to alcohol, hereditary, idiopathic, auto – immune or tropical, with alcohol being the commonest in the west and Tropical pancreatitis being the commonest in India.[5]

The is no definitive information regarding the prevalence of Tropical Chronic Pancreatitis in Sri Lanka.[1]The aetio-pathogenesis of TCP is multi factorial, including; malnutrition, toxic effects of cyanide derived from frequent cassava consumption, genetic factorsand increased oxidant stress from vitamin C and A deficiencies.

The classic clinical picture reported in patients with TCP

in India is of younger age group, who present with abdominal pain, diabetes and malnutrition.[5] This is in contrast to our patient who was an elderly male with no abdominal pain at presentation, this is conforming with the study done in 2005 in patients with TCP in Sri Lanka, where most of the patients were reported to be much older than their Indian counterparts.[1]Fibrocalculous pancreatic diabetes (FCPD),[3] liver cirrhosis, acute cholangitis and pseudocyst formation are recognized complications.

Repeated ERCP with stenting and broad - spectrum antibiotics were necessary to manage the recurrent admissions with obstructive jaundice and cholangitis due to large pancreatic head calculi.

Conclusion

Tropical Chronic Pancreatitis, though classically described in the young, can occur in the middle aged or elderly, without a history of abdominal pain, as shown in this case. Therefore a high index of suspicion is necessary in early diagnosis and treatment. Recurrent stone formation, recurrent obstructive jaundice and cholangitis and micro and macrovascular complications of FCPD may complicate the management.

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STERILE PSOAS ABSCESS IN A PATIENT WITH GRANULOMATOSIS WITH POLYANGITIS (WEGENER'S GRANULOMATOSIS) – A CASE REPORT

Jayasinghe VP, Arulmoly K

Keywords : Vasculitis,Granulomatosis with Polyangitis, Psoas abscess, Immunosuppression, ANCA

Introduction

Granulomatosis with Polyangitis (GPA) formerly known as Wegener's Granulomatosis is a rare multisystem small and medium vessel vasculitis of known origin. The hallmark of the disease is formation of non – casiating granulomas and small and medium size vessel vasculitis with positive Anti –neutrophil cytoplasmic antibody (ANCA)[1]. Psoas abscess is a collection of pus in the iliopsoas muscle compartment [2]. It may arise from hematogenus spread from distant site (Primary Psoas abscess) or from contagious spread from adjacent structures (Secondary Psoas abscess).Other conditions such as plasmacytomas, lymphomas and foreign bodypseudo-tumors may mimic psoas abscess, however GPA presenting with sterile psoas abscess is a very rare presentation. We have found no account of similar presentations in local literature and only one presentation was found in international literature[3].

Case Report

Mrs. K a 50-year-old female with diabetes, chronic kidney disease and Granulomatosis with Polyangitis (GPA) since 2009, presented to us with fever with chills and rigors, weight loss, loss of appetite, back pain, dysuria and difficulty in walking for 3 months duration.

She was first diagnosed with GPA in 2009 when she presented with acute kidney injury, renal biopsy at that time revealed Pauci – immune, C - ANCA positive vasculitis. She had undergone induction with 6 pulses of Cyclophosphamide and 3 pulses of Methylprednisolone and rescue hemodialysis. She was on Azathioprine 50mg maintenance, however her disease control was poor and the Birmingham Vasculitis Activity Score (BVAS) [4] was 17. She was febrile and pale. There was Right renal angle tenderness and Right sided Psoas sign was positive.

She had neutrophil leukocytosis (WCC 10.63*109, N 80%), high inflammatory markers (ESR 122 mm/1st h, CRP – 122 mg/dl), deranged renal functions (Creatinine 2.5 mg/dl) and her Urine full report revealed field full of pus cells. Repeated blood and urine cultures were sterile. The ultrasound abdomen revealed only chronic renal parenchymal disease and she underwent Contrast Enhanced CT of abdomen and pelvis, which revealed a right-sided psoas abscess. Ultrasound guided aspiration of the abscess was done 3 times, however it failed to completely resolve the abscess; hence she underwent surgical incision and drainage under spinal anesthesia. The abscess fluid was purulent and full of neutrophils, however repeated aerobic, anaerobic and fungal cultures failed to yield an organism. The abscess fluid was sent to National Hospital for respiratory diseases for Tuberculosis PCR and culture, which turned out to be negative. Fungal studies of the abscess fluid were negative as well. Unfortunately the abscess wall was not sent for histology and hence no granuloma formation could be detected. Incision and drainage of the abscess alleviated her symptoms and it was decided that the best course of management would be to commence her on Rituximab and Methylprednisolone for better control of GPA.

Discussion

Granulomatosis with Polyangitis (GPA) is a rare multisystem autoimmune disease of unknown etiology. The hallmark features are necrotizing granulomatous inflammation and Pauci – immune vasculitis in small and medium sized blood vessels. Non-specific systemic symptoms of GPA include fatigue, fever and weight loss. Pulmonary manifestations, musculoskeletal manifestations, cutaneous manifestations, ENT manifestations, renal manifestations and urogenital manifestations are common with GPA. Sterile abscess formation also has been reported in literature, namely parotid abscess[5], prostate abscess[6], breast abscess and lung abscess. There are no case reports of psoas abscess following GPA in local literature, however there is one report of GPA mimicking psoas abscess[3]. The patient had very poor disease control despite being on maintenance of Azathioprine and the absence of an

organism in multiple cultures led to the suggestion that the abscess may have occurred as a result of poor control of GPA. Ideally the abscess wall should have been sent for histology, however only pus from the abscess was sent for cytology, which failed to reveal anything.

Conclusion

This case report highlights that physicians must be aware of the rare manifestations of Wegener's Granulomatosis, which includes findings that mimic pyogenic abscesses.

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RARE PRESENTATION OF MICROSCOPIC POLYANGIITIS WITH NASAL SEPTAL PERFORATION - CASE REPORT

Kuruppu Arachchi AN,Thebuwana KL, Jayasinghe VP, Sundaresan KT, Umakanth M, Mayurathan P, Rushdie MNM

Keywords : microscopic polyangiitis, nasal septal perforation, ANCA associated vasculitis

Abstract

Microscopic polyangiitis(MPA) is a ANCA associated small vessel vasculitis characterized by the presence of Anti Neutrophil Cytoplasmic Antibodies and few or no immune deposits in the involved vessels.Kidneys are the most commonly involved organ in 90%[1]. This is a case of 70 year old female diagnosed case of microscopic polyangiitis presents with disease flare up with renal involvement and nasal septal perforation.

Introduction

Small vessels vasculitis involves arterioles, capillaries and venules. Granulomatosis with polyangiitis, MPA and Churg-Strauss syndrome comprise ANCA associated small vessel vasculitis and characterized by the paucity of immune deposits. MPA initially considered as microscopic form of Polyarteritisnodosa, in 1994 international consensus conference in Chapel Hill redefined this as a separate entity[2].

MPA is a rare condition which can occur at any age, but typically occur in the fifth and sixth decade. Males are more affected than females[3]. It's a necrotizing vasculitis with few or no immune deposits predominantly affecting small vessels without granuloma formation[4].Since it is a vasculitis it can affect any organ, but most commonly involved are kidneys, lungs,peripheral and central nervous system and skin. Constitutional symptoms such as fever, myalgia, lossof weight are relatively common. But upper respiratory tract involvement is around 1%[2].

Diagnosis of ANCA associated vasculitis is made on basis of clinical findings, by biopsy of relevant organ involved(kidney, nasal mucosa or nerve)and the presence of ANCA[5]. In patients with MPA, 60%-80% positive for MPO-ANCA (p-ANCA) and around 40% is positive for PR3-ANCA (c-ANCA).[1]

Case report

70 years old female house wife previously diagnosed patient with acute coronary syndrome and microscopic polyangiitis in 2013, following mono neuritis multiplex as evidenced by bilateral lower limb weakness and unexplained sensory impairment this time presented to us with generalized weakness ,tiredness and exertional dyspnea and subjective weight loss over 1 month duration. In 2013 she was diagnosed as MPA with the evidence of positive p-ANCA, elevated ESR and severe sensory motor axonal type polyneuropathy in nerve conduction study. She was treated with oral cyclophosphamide and prednisolone as an induction therapy, but she defaulted clinic follow up thereafter. There were no renal or respiratory tract involvement at that time and her Bermingham vasculitis activity score is not assessed.

During this admission she was afebrile, pale and ill looking. She had flat nasal bridge without rhinorrhea. ENT examination revealed nasal septal perforation. She didn't have any vasculitic or hypo pigmented skin rashes or stigmata of connective tissue disorders or peripheral nerve thickening. Her cardiovascular, respiratory, abdominal and nervous system examination was otherwise normal.

Her full blood count revealed neutrophil leukocytosis with hemoglobin level of 7.2g/dl. Blood picture revealed moderate anemia due to chronic disease. Her ESR was 140mm/1sthr and CRP was 122 mg/dl. Her renal functions were elevated; Scr 4.7mg/dl andblood urea 167 mg/dl. Her liver functions were normal. Urine full report revealed albumin 1+,without dysmorphic RBCs. Her urine PCR was 34mg/mmol. Ultra sound abdomen revealed normal

sized kidneys with increased echogenicity and poor cortico medullary demarcation. Her renal biopsy revealed chronic glomerulonephritis with moderate tubular atrophy and fibrosis. Her chest x ray was normal. Her nasal septal biopsy showed chronic inflammatory changes without granulomas. Her renal and mesenteric CT angiogram was normal. Her p ANCAwas positive. ANA negative with normal C3 levels. Her STD screening was normal including Hep B and HIV. Dermatology referral done and excluded the possibility of leprosy.

She was diagnosed to have minor flare up of microscopic polyangiitis with established CKD, and started treatment with prednisolone and azathioprine[6]. Her Bermingham vasculitis activity score was 17.5 .

Discussion

This patient was considered to have a minor flare up as evidenced by elevated inflammatory markers, positive ANCA and in the absence of the major organ involvement. Raised serum creatinine was considered due to already established chronic kidney disease rather than active vasculitis. Nasal septal perforation is a common presentation among wegener's granulomatosis with granuloma formation in nasal biopsy. Upper respiratory involvement is a rare presentation in MPA, although there is one case report of nasal involvement confirmed by biopsy from nasal cavity and para nasal sinuses[7]. Minor flare ups can be treated with re introduction of maintenance therapy[6] and this patient was treated accordingly.

Conclusion

Microscopic polyangiitis is a small vessel vasculitis.If not treated promptly it can lead to multi organ failure. So early treatment and close follow up is essential in disease management. Bermingham vasculitis activity score can be used to detect disease flare ups together with inflammatory markers.

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MELIOIDOSIS WITH MULTIPE SKIN AND SPLENIC ABSCESSES WITH SEPSIS - A CASE REPORT

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Keywords : Melioidosis, Splenic Abscess, Sepsis, Burkholderia pseudomalle

Abstract

Melioidosis is an infection with Gram Negative bacterium Burkholderia pseudomallei. It is now emergent in Sri Lanka as an important pathogen causing various clinical manifestations. This case report is of a 67-year-old patient with multiple skin and splenic abscesses with positive blood culture and pus culture for Melioidosis.

Introduction

Melioidosis is caused by the saprophytic soil bacterium Burkholderia pseudomallei, important cause of sepsis in East Asia and Northern Australia. It is characterized by formation of abscesses especially in the lungs, liver, spleen, skeletal muscle and prostate mostly among immuno compromised adults such as Diabetes mellitus, chronic renal disease and alcoholism.[1] Infection is acquired through percutaneous inoculation (penetrating injury or open wound), inhalation or ingestion (through contaminated food and water) and Melioidosis has also been transmitted to infants through breast milk from mothers with mastitis.[2] Correct microbiological diagnosis and prompt antimicrobial therapy is important in order to reduce high mortality and morbidity.

Case Report

67-year-old female housewife presented with abdominal pain for 1-monthduration, which was progressively worsening and most prominent in left upper quadrant. She had evening pyrexia for 2 weeks with associated chills and rigors, also complained of loss of appetite, fatigability and exertional dyspnea. During her in - ward stay she developed right-sided knee joint and right hand superficial abscesses. She is a known diabetic with poor glycemic control, hypertension and ischemic heart disease for which she underwent Coronary Artery Bypass grafting 8 years ago. She is a non – alcoholic and non – smoker. She has an exposure to muddy water during the period of flood in 2016.

Examination revealed an ill looking febrile patient. She was pale but non – icteric and had no palpable lymphadenopathy. Her cardiovascular, respiratory and neurological examination was normal. Her abdomen was not distended, mild left hypochondrial tenderness was noted. There was no organomegaly or free fluid. She had superficial abscesses over Right side knee joint and Right hand.

Her investigations revealed high inflammatory markers(ESR-142mm/hr, CRP-234mg/dl), full blood count showed neutrophil leukocytosis and hemoglobin of 8.3g/dl. Her AST was 82 U/l and ALT was 48 U/l. Blood picture revealed Anemia of Chronic Disease. 2D Echo done to exclude infective Endocarditis which revealed, left ventricular hypertrophy with septal hypokinesia and there were no vegetations. Ultrasound scan of abdomen revealed four small hypo echoic lesions in the spleen, largest of which was 8cm in diameter, suggestive of splenic abscess. Contrast enhanced CT chest and abdomen confirmed the presence of abscesses. Blood culture was positive for Pseudomonas. Species and confirmed as Burkholderia pseudomallei by agglutination test. Pus cultures from superficial abscesses revealed Pseudomonas. Ultrasound guided splenic abscess aspiration was done and culture was positive for Burkholderia pseudomallei. Melioidosis antibody titer was >10240. Retro viral screening and Mantoux was negative.

We started this patient on intravenous Ceftazidime 2g 6hourly and Co-trimoxazole 1920mg bd for 1-month duration.[3] Superficial abscess were drained by the surgical team and wound dressings were applied and the largest splenic abscess was drained under ultrasound guidance by the Consultant Radiologist. Proper glycemic control was achieved with soluble insulin and later converted to oral hypoglycemic agents. After 1 month of completion of antibiotics, contrast enhanced CT abdomen was repeated and revealed abscess were diminishing in size. We discharged her on oral Co-trimoxazole for another 3 months.

Discussion

Distribution of Melioidosis probably extends beyond South East Asian and Australia as new cases emerges from Indian subcontinent and some parts of Sri Lanka in increasing number. [6] Since Burkholderia is a saprophytic soil bacterium it is likely to be more prevalent in agriculture based areas and some areas with frequent flooding as in our patient. [4] It acts as an opportunistic pathogen since it is more common in individuals with immune suppression such as diabetes, renal insufficiency, liver disease, chronic alcoholism or patients in long-term immunosuppressants, in our case she is a diabetic with poor glycemic control. Melioidosis can present in various ways; acute or chronic infection or localized or disseminated infection as in our patient. High index of clinical suspicion is needed for accurate diagnosis. Definitive diagnosis of Melioidosis is made on isolation of Burkholderia pseudomallei on culture. Lack of familiarity with the cultural characteristics of the pathogen may lead to delay in diagnosis or misdiagnosis. [5] Treatment of Melioidosis consist of intense period of at least 14 days, but usually more as local microbiological protocols. Intravenous Ceftazidime, Meropenem or Imipenem administered as first line agents along with oral Co-trimoxazole, followed by oral co – trimoxazole for further 3-6 months. Careful search of internal abscesses is recommended and adjunctive therapy for abscesses include drainage, washout or aspiration of septic joints. [2]

Conclusion

Melioidosis is a potentially treatable condition with high morbidity and mortality. Early diagnosis warrants much-improved outcome, so need to increase the awareness among the clinicians about this condition. Well trained microbiology team and improved culture techniques is a paramount to accurate diagnosis. Multidisciplinary approach with close liaisons with Physician, Microbiologist, Radiologist and Surgical team will improve the overall outcome.

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ANOMALOUS ORIGIN OF LEFT CORONARY ARTERY FROM PULMONARY ARTERY (ALCAPA)

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Abstract

Anomalous originof left coronary artery from pulmonary artery (ALCAPA) is a rare congenital anomaly with an incidence of 1 in 300,000 live births [1].The 85% of all cases are diagnosed within the first month of life. who are presenting with severe left sided heart failure and mitral valve insufficiency. Most of the untreated patients (90%) die within one year of life. The adult form of ALCAPA may occur in approximately 10% of cases due to adequate collateral blood supply to the left coronary circulation through right coronary artery.

Adult ALCAPA can present as effort shortness of breath and angina due to relative ischemia or coronary steal phenomena (which is collateral circulation between right and left coronary system ensues and left coronary artery flow reverses in to pulmonary artery trunk due to the low pulmonary artery pressure).

Here I present a 36 years old male patient presenting with exertional shortness of breath, effort angina and two episodes of loss of consciousness for about two hours duration in two separate occasion in the past and being finally diagnosed as adult ALCAPA following coronary angiogram.

Case Report

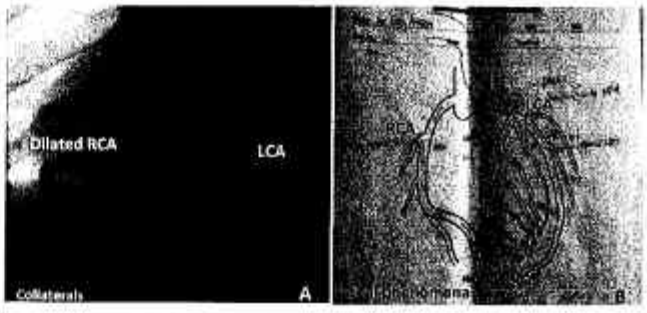
A 38-year-old, father of three children and brick binder was normotensive andnon-diabetic with no family history of ischemic heart disease but he has been smoking 10 cigarettes per day for last fifteen years. He presented with dyspnoea, effort angina since child hood and he had two episodes of loss of consciousness for about two-hours in two separate occasions in the past at the age of 17 and 21 respectively. Both episodes of loss of consciousness were followed by short period of palpitation after blunt trauma to the left chest. He was recovered spontaneously after two hours without any specific treatment in eachoccasion.

His pulse rate was 64 beats per minute with regular in rhythm and blood pressure was 120/80 mm of Hg. Cardiovascular examination revealed displaced apex beat with S3 gallop and pan systolic murmur heard at mitral area with radiated to axilla. He had normal vesicular breath sounds over the chest. His ECG showed prolonged PR interval (1st degree heart block) and partial LBBB with left axis deviation. His initial 2D transthoracic echo cardiograph showed mildly impaired Left ventricular function (EF- 50%)with globally hypokinetic Left ventricular muscles, Grade 1 diastolic dysfunction, grade 11 Mitral regurgitation with dilated Left ventricular cavity (LVEDD 67mm). Origin of the Left coronary artery (LCA) couldn't be found.

An exercise treadmill test was performed revealed ST depression change in lateral leads at stage 1V modified Bruice protocol.Apart from faintness at stage 1V, he was totally asymptomatic during this study. The patient was referred to coronary angiography, which showed an anomalous LCA arising from pulmonary artery with retrograde filling through collateral from an enlarge right coronary artery (RCA).

Blood Flow → Aorta → Large RCA → LAD/LCX → LMCA → MPA

The patient was referred to cardiothorasic surgical unit for surgical correction of ALCAPA and mitral valve repaire.



Blood Flow → Aorta → Large RCA → LAD/LCX → LMCA → MPA

Figure 1:A Dilated RCA from Aortic sinus with steal phenomenon.B, Schematic diagram of coronary angiogram

Discussion

The ALCAPA results when the left main coronary artery arises from the pulmonary trunk instead of the ascending aorta. It usually manifests as an isolated defect, but can be associated with other cardiac anomalies in 5% of cases. This abnormality only accounts for 0.25-0.5% of all congenital cardiac anomalies. Like other congenital heart defects,the condition is generally considered to be based on multifactorial inheritance and it is unrelated to utero exposure to teratogens, chromosomal abnormalities, or other risk factors.

This anomaly may result from abnormal septation of the conotruncus into the aorta and pulmonary artery, or from persistence of the pulmonary buds together with involution of the aortic buds that eventually form the coronary arteries.

The function of the left main coronary arterial territory then often requires extensive collateral formation from the right coronary artery.

There are two forms (infant and adult) based on onset of disease each of which has different manifestations and outcomes. In the first month of life, physiologic pulmonary hypertension tends to preserve antegrade blood flow within the left coronary artery, and infants usually remain asymptomatic.

The ALCAPA can present in adult life mostly with Left ventricular systolic function. But symptoms may range from dyspnea, chest pain, exercise intolerance and sudden cardiac death due to acute ischemia or malignant ventricular arrhythmia during exercise [2, 3]. The true incidence of older patient is not known with only case reports of patient older than 50 years of age [4-6].

The ALCAPA was first described by Krause [7] and Brook [8] in 1865 and 1885 respectively from post mortem examination. However, the first clinical description was published by Bland, White and Garland in 1933 [9] and the condition is also known as Bland, White and Garland syndrome.

The infants (with ALCAPA) develop severe left sided heart failure and significant mitral incompetence at the age of one month; initial symptoms are feeding difficulties, irritability, diaphoresis, tachypnea, tachycardia and cyanosis. The chest pain due to myocardial ischemia could be mistaken with infantile colic. Though it is a congenital coronary anomaly it should still be kept in mind while dealing with an adult patient with such type of presentation.

However, during the neonate period pulmonary pressure decrease and together with ductus arteriosus closure, results in an increase in systemic arterial pressure reversing the blood flow within the LCA retrograde in to pulmonary artery (PA). Therefore, degree of myocardial ischemia is dependent on the extent of the collateralization between RCA and LCA.

Coronary angiography alludes to the diagnosis, with the present of a large tortuous RCA with collateral filling the LCA system.However, a dilated RCA with retrograde Doppler flow from LCA to PA with visualization of septal flow due to collateral have been described as diagnosis echocardio -graphic criteria for ALCAPA [10].Anyway, normal echocar -diogram may be seen in 10% of adult ALCAPA patients.

Surgical correction is the treatment of choice in suitable patient as soon as possible, with the medical management as only supportive and temporary. There is acceptable corrective procedure for ALCAPA are available,

(1). Ligation of the anomalous left coronary artery at its origin.

(2). Left coronary artery was closed by device embolization [11].

(3). Reconstruction of a double coronary artery system.

First two procedures should be done only in extensive collateral supply from the right coronary artery is available. The double coronary artery system can be created in many ways.

a) Translocations of anomalous left coronary artery to the aortic sinus [12,13]

b) Aorta –left coronary artery bypass graft (saphenous vein) in combination with ligation of anomaly artery origin.

c) Left coronary artery conduits using left common carotid artery or sub clavian artery and left internal thoracic artery.

d) The Takeuchi procedure [14] (where a creation of an aorto-pulmonary window and an intra-pulmonary tunnel extending from the anomalous ostium to the window).

Post-operativelong-term survival (20years)in infants with ALCAPA was shown to be 94.8% [15].Most of the infants with corrected ALCAPA show normalization of both ventricular failure and mitral valve insufficiency.No long-term studies of large population of adult with corrected ALCAPA are available but the prognosis is generally good. Restoration of a dual coronary system will prevent further ischemia and arrhythmia.The patients with a history of ventricular tachycardia or ventricular fibrillations not associated with acute ischemic event antiarrhythmic treatment must be considered. Andcan consider ICD implantation or catheter ablation.

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ABERRANT ORIGIN OF COMMON HEPATIC ARTERY – A TECHNICAL CHALLENGE DURING PANCREATICODUODENECTOMY.

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Abstract

The gastroduodenal artery requires division at the upper border of duodenum, as it branches off the common hepatic artery during pancreatico-duodenectomy. In 2-4% of population, the common hepatic artery is originating from superior mesenteric artery (SMA), known as the replaced hepatic artery which can be accidentally ligated.

Therefore recognition of vascular anatomy on preoperative imaging is important to plan the surgery.

In the patient presented, at the time of surgery, a replaced hepatic artery was found. This highlights the importance of vigilance regarding anomalies during surgical dissection, even if the pre-operative imaging doesn't display such anomalies.

Keywords : Replaced hepatic artery, Gastro duodenal artery, Pancreaticoduodenectomy.

Keywords : Replaced hepatic artery, gastroduodenal artery, pancreaticoduodenectomy

Introduction

The gastro duodenal artery, supplying the pancreatic head and the duodenum needs division during pancreaticoduodenectomy[1]. A precise identification of this artery branching off the common hepatic artery (CHA) is essential to avoid inadvertent ligation of the latter which oxygenates the liver. However in 2-4% of population, the common hepatic artery is originating from superior mesenteric artery (SMA), known as the replaced hepatic artery [2]. The gastro duodenal artery is given off on way.

In the patient presented, at the time of surgical dissection, a replaced hepatic artery was found.

Case Report

A 68yr old male underwent a pancreaticoduodenectomy for carcinoma of distal common bile duct. During the supra-duodenal dissection the hepatic artery did not have its usual pathway of coming from the left side originating at the celiac trunk, giving off the gastro-duodenal artery and continuing up, to the hilum of the liver. The hepatic artery was found to be originating from the SMA giving the gastro-duodenal artery on way. The hepatic artery was dissected along its pathway up to liver hilum. Division of the common bile duct facilitated the dissection and ligation of gastro duodenal artery.

The resection and anastomosis were completed with the standard technique

Discussion

Various studies have enumerated that a classical common hepatic artery giving off the gastro duodenal branch and continuing as proper hepatic artery dividing in to right and left hepatic arteries is evident only in 50-60% of the population. Michel et al in 1955 classified variations seen in 40-50% [4]. Most variations involve replacing the classic origin of right and left hepatic arteries, often recognized by pre-operative imaging. However a majority of these are clinically not significant.

A critical evaluation of CHA giving off the gastro duodenal artery is important, since division of the latter is a mandatory step in pancreatico-duodenectomy. Inadvertent ligation of hepatic arteries renders the liver and bile ducts ischemic and may cause biliary fistulas[3]. Recognition of vascular anomalies on preoperative imaging is preferable to identification at surgery.

In 2-4% the common hepatic artery originates from SMA, known as the replaced hepatic artery [5]. This variant was found in the patient presented, at surgical dissection.

Conclusion

Recognition of vascular anatomy on preoperative imaging and vigilance during surgical dissection regarding anomalies is important for safe surgery.

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Case Report

HYPOGASTRIC HERNIA - AN UNUSUAL VENTRAL HERNIA

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40y old previously healthy male patient admitted to emergency department with lower abdominal swelling, pain and inability to pass urine for 2h duration.

The swelling had developed suddenly without any predisposing cause followed by development of severe colicky abdominal pain with in few minutes. There was associated nausea and vomiting and patient was not able to void after the occurrence of pain. Although patient was in distress due to pain he was haemodynamically stable apart from mild tachycardia of 90bpm.

On abdominal examination there was a triangular shape, tense tender lump in midline between the umbilicus and pubic symphysis with undistorted umbilicus [Fig 1]. There was no cough impulse at the lump and the percussion note was resonant. No bowel sounds were audible over the lump. There was no inguinal or para umbilical hernia found.

Urinary catheter inserted only drained 150ml of urine with no reduction in size of lump. Ultrasound scan of abdomen revealed a suprapubic region midline hernia containing entrapped bowel loops with no peristalsis suggestive of obstructed, strangulated hernia.

Patient underwent urgent surgery where subcutaneous hernia sac with strangulated small bowel loop was identified. The neck of the hernia was through a small infraumbilical midline defect in the linea alba. Following release of tight neck, bowel regains its normal color and peristalsis. Contents were reduced and anatomical repair of the defect in the rectus sheath was done followed by onlay mesh repair since there was no contamination of wound. Post-operative recovery was uneventful and patient was discharge 2 days after surgery.

Figure 1 – patient had infra umbilical swelling with non-distorted umbilicus.

Discussion

Ventral hernias are hernias occurring in the anterior lateral abdominal wall excluding inguinal hernias. They include midline hernias [Epigastric hernia, umbilical hernia, Para umbilical hernia and hypogastric hernia] and lateral hernia known as Spigelian hernia [1]. They occur as a result of defects in aponeurotic system of anterior abdominal wall.

This patient had a hypogastric hernia. It is a rare midline hernia occurring below the umbilicus which has high risk of strangulation [2, 3]. In contrast to umbilical and para umbilical hernia, umbilicus is not distorted in hypogastric hernia.

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GASTROSCHISIS

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Key words: Gastroschisis, Congenital anomaly

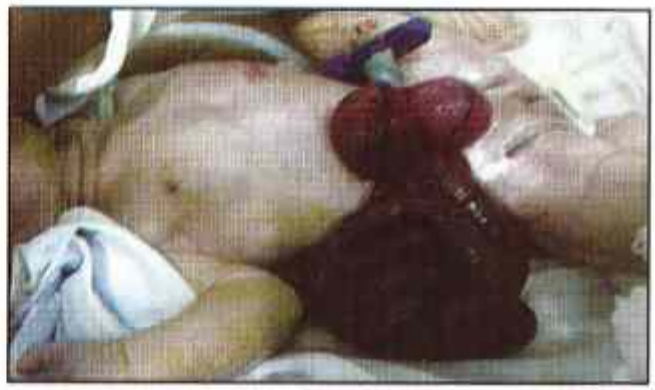
Introduction

Gastroschis is a congenital anomaly which is characterized by a small abdominal wall defect located to the right of the umbilical ring through which the gastrointestinal structure is herniated into the amniotic cavity (1, 3). The incidence of gastroschisis is between 1 in 6000 and 1 in 10 000 live births and the survival rate is approximately 90% (2, 3).The risk factors for this abnormality are young maternal age, low BMI, race, smoking, low socioeconomic status, recreational drug use, and alcohol and consumption during pregnancy(2, 3,).

We report a case of a neonate with gastroschisis who was born at Kalmunai Base Hospital of Sri Lanka and an uneventful outcome.

Case Report

A baby girl weighing 1850g was born at 34 weeks of gestation by normal vaginal delivery following a spontaneous onset of labour. The parents were not consanguineous, the mother was 41year old while the father was 46years and they already had three healthy children. This was an unplanned pregnancy and mother had failed to be followed up at antenatal clinic regularly. She had been is treated for rheumatoid arthritis with oral Methotrexate and she had continued Methotrexate during pregnancy without consultation. She did not have other pregnancy related problems such as diabetes or hypertension. She neither smoked nor took alcohol. The antenatal fetal anomaly scan was not done. The baby was born in good condition with satisfactory APGAR score and required no resuscitation. The examination revealed herniation of gut loops through the anterior abdominal wall to right of the umbilicus and the herniated gut contained jejunum, ileum, large bowel , ovaries and mesenteric vessels which were twisted around each other. Apart from this no other external defects were identified .Soon after birth she had mild grunting which resolved with oxygen via nasal cannula. Otherwise her cardiovascular, respiratory and neurological examination was unremarkable. The baby was transferred to Teaching Hospital Batticaloa which is the nearest tertiary care centre (40Km from Kalmunai) which has the paediatric surgeon and the neonatal intensive care unit which can deliver post surgical care. After stabilization the baby underwent Silo bag fixation and following a week of post operative care in the neonatal unit she had an uneventful recovery. During the local hospital clinic review the baby had an echocardiogram, ultrasound brain and abdomen which were normal and her growth and development was within the limit.



Discussions

Gastroschisis is diagnosed by routine ante natal ultrasound scan between the 18th and 20th week of gestation and the maternal alpha fetoprotein of serum is also usually elevated in this condition (3, 4). Antenatal diagnosis helps planning timing, location, and mode of delivery in advance as this rare malformation requires timely surgical correction (4). This stresses the importance of prenatal diagnosis of gastroschisis for better neonatal outcome.

Advances in both surgical and neonatal care have decreased the mortality and morbidity of abdominal wall defects over recent decades. (3).Fluid resuscitation and caring of herniated viscera and their blood supply are mainstay of initial management .Nursing in incubator will reduce the heat loss of babies born with abdominal wall defects

and a nasogastric tube should be inserted to facilitate bowel decompression (3)

Methotrexate is a teratogenic agent which can cause spontaneous abortion as well as congenital malformations therefore the use of Methotrexete during pregnancy is not advisable(5)

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A CASE OF RUPTURED HETEROTOPIC PREGNANCY

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Introduction

Heterotopic pregnancy (HP) is rare and potentially fatal early pregnancy complication. The presence of an intrauterine uterine pregnancy (IUP) often makes difficulty in diagnosis of heterotopic pregnancy.

Case

A 29-year-old primipresented with lower abdominal pain without vaginal bleedingat 7 weeks of period of amenorrhea. Initially she was reassured following an assessment by a gynaecologist. At that time, her examinations were normal and a transvaginal ultrasonography (TVS) revealed an IUP.

Three days after the initial presentations, she was admitted with severe lower abdominal pain, shoulder tip pain and faintness.She was in shock and tenderness in left iliac fossa and adenexia were noted. TVS revealed an IUP with a mixed echogenic mass in the left adnexa and significant free fluid in pelvis. An urgent laparotomy was carried out and left side salpingectomy was performed. Histology confirmed the ectopic gestation. Rest of her pregnancy was uneventful and she delivered a term baby.

Discussion

Heterotopic pregnancy is defined as the co- existence of uterine and extra uterine pregnancy which is commonly in the fallopian tube. The incidence is 0.6-2.5/10,000 pregnancies and increases with artificial reproductive technique.A high-resolution TVS with color Doppler is useful in diagnosisa heterotrophic pregnancy. Treatment options are conservative, medical or surgical and it depends on clinical features and viability of pregnancies.

Conclusion

High index of suspicion is necessary to diagnose the heterotrophic pregnancy and it should be always a differential diagnosis when a woman presenting with abdominal pain even there is an IUP.

BACLOFEN INTOXICATION

Indika Prasad H.M.R., Sivakanthan K.

Introduction

Baclofen is a derivative of gamma –aminobutaric acid used for symptomatic relief of spasticity and spasms of skeletal muscle. [1] Recommended maximum pediatric oral dose for 5yearchild is 40mg/day [2]

Non therapeutic ingestion of adult prescribed drugs is a major Peadiatric health care problem which is recently increasing in incidence. [3] Many drug poisoning in Peadiatric age group take place in child's own house. Medicinal drugs are the leading cause of non-fatal poisoning in children in middle-income countries. [4]

Case

A 5 year old preschool boy transferred from local hospital following ingestion of an unknown amount of an unknown tablet which was prescribed for his paralyzed aunt. The child had taken tablets at around 12-12.30 pm and then he had eaten a mango. He vomited 4 times before admission to local hospital. Child was drowsy on admission. Mother told that the aunt has witnessed child was playing with her drugs and taking them (she couldn't resist as she is bedridden). There was no history of head injury or snake bite.

On admission to ETU child was drowsy and unconscious. Glasgow coma scale (GCS) was 7/15. Respiratory rate 20/min and good respiratory effort was there. Oxygen saturation was 98% at room air. Auscultation findings were vesicular breath sounds with coarse crepitations and conducting sounds. Pulse rate was 70-80/min, regular and blood pressure was 70/40mmHg. Peripheries were cold and muscle tone was low with reduced reflexes. Pupils were dilated, 5mm in size and reacting to light. Capillary blood sugar was 104mg/dl.

Normal saline bolus was started, kept left lateral position with oxygen and secretions were sucked out. Elective intubation done with the help of Anesthesia team as GCS was 6-7/15. Gastric lavage with normal saline 5 ml/kg (75 ml) done and activated charcoal 1g/kg (15g) in 60 ml normal saline was given via nasogastric tube. We managed to contact one of the child's relative living near their house and he vibered the photos of offending drug .It was found to be Baclofen tablets. Atropine one dose given as there was bradycardia. Intravenous antibiotics, Cefotaxime and Metronidazole started due to suspicion of aspiration. Child transferred to medical intensive care unit (MICU) for ventilation and monitoring. At the MICU blood pressure was 110/70mmHg, pulse rate was 102/min with good oxygen saturation. He was kept warm. For sedation propofol infusion started. Intravenous Ondansetron and intravenous Ranitidine also started. Facilities to measure Baclofen plasma levels were not available.

Sedation tailed off after 10 hours and self extubated by 12 hours of ICU care. Reflexes and tone started to improve and GCS was 15/15. Patient monitored at ICU for 20 hours and transferred to ward high dependency unit (HDU). Oral feeds started and patient was well by 24 hours apart from drowsiness.

Patient discharged after 72 hours of admission.

Discussion

Symptoms of Baclofen poisoning usually appear within 2 hours as its rapidly absorbed from the gastrointestinal tract and drug is excreted via kidney [5,6] Usual elimination half life is 2-6 hours (mean 3.5).85% excreted unchanged in urine and rest metabolized by liver. [4].Assessment was done clinically. As patient came within 2 hours of ingestion and the amount of drug taken is unknown. a gastric lavage after protecting the airway was done and activated charcoal was given. [7]

Central nervous system depression is well known with baclofen toxicity. [5, 8] It also causes lethargy, somnolence, hallucinations, coma, seizures, encephalopathy, respiratory depression, flaccidity, hyporeflexia and agitation. [5, 8, 10, 11] GCS was 7/15 which was an indication for intubation and ventilation to protect airway. Autonomic dysfunction is common but inconsistent. Patient had bradycardia, therefore atropine was given which has effect on improving ventilation, heart rate and blood pressure and core

temperature. [9] Hypotension was also noted and given fluid bolus. Patients can get meiosis or mydriasis; our patient had mydriasis. [11]Hypothermia is a known effect of baclofen poisoning. [8, 10, 11] Patient warmer and bed sheets were used to keep the patient warm. Convulsions and cardiac arrhythmia are known complications. [8, 10, 11] ECG monitoring of our patient was normal and fortunately there were no convulsions.

To reduce the risk of aspiration with vomiting, intravenous ondansetron was prescribed. There is a place for physostigmine in mild to moderate baclofen poisoning. [11] .

Clinical effects of baclofen poisoning last for 5-8 hours. Outcome was good with supportive care. [8, 11]

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DENGUE HAEMORRHAGIC FEVER IN A THIRTY SIX DAYS OLD INFANT

V. Thirukumar, WMP Weerakoon

Key words : Dengue Haemorrhagic Fever

Introduction

Dengue infection has become a major public health problem in Sri Lanka in past two years. Incidence of dengue infectionin Sri Lanka was 12976 up to July 2017 [1]. Dengue fever is an acute febrile illness caused by four serotypes of Dengue virus (DENV1, DENV2, DENV3, DENV4) and infection with a dengue virus may be clinically asymptomatic or may present as a undifferentiated febrile illness, dengue fever (DF), dengue haemorrhagic fever (DHF) or Unusual Dengue[2]. Reported cases of dengue fever (DF) or dengue haemorrhagic fever (DHF) are rare in neonates and early infant. We report a case of DHF in a thirty six days old infantwhich is the youngest age presented to our unit.

Case Report

A thirty six day old baby girl was admitted to paediatric unit with a three day history of fever and reduced feeding for one day. But there was a bulging fontanelle and 3cm palpable liver. Initially patient was treated with a presumptive diagnosis of septicemia associated with meningitis but septic screening was normal. Since Mother also had fever on admission and they presented from a dengue endemic area both mother and child weretested for Dengue NS1 Antigen where it becamepositive.

[1]Consultant Paediatrician,[2]Registrar in Paediatrics,TH Batticaloa, Batticaloa.

Ultrasound scans of the abdomen and chest done after admission revealed evidence of leakage. Baby was managed as DHF.In critical phase baby was managed with intravenous fluid and breast feedingaccording to hematocrit and urine output. Throughout the critical phase of the illness, he was haemodynamically stable and had no bleeding manifestations. After ten days of illness baby was discharged and Dengue IgM and IgG antibody done on 10th day of illness, ware positive.

Discussion

As the incidence of dengue infection in Adults has increased, it has also increased in breast feeding mother as well as infant. Vertical transmission of DENV has been reported[3,5] and the mechanisms of infection transmission remain unclear in the peripartum period.Clinical manifestations of DHF are more significantly associated with death in infants compared with older children. Neonatal DHF is rare.Only a few cases of DHF are reported worldwide. Complex pathogenesis of DHF during primary dengue in infants and neonates, due to the scarcity of clinical experience and unavailability of studies andguidelines, affects the probable management.

Table 1: Result of investigations

	3rd Day	4th Day	5th Day	6th Day	8th day	10th Day
White cells (mm-3)	7.17	6.62	7.89	8.29	8.78	8.88
Platelets (10^3/mm3)	160	57	59	78	88	102
Haemoglobin (g/dl)	11.7	12.2	12.1	13	12.3	12.1
HCT	34.3	37	36	35	34	34
CRP (mg/l)	<5					
NS 1 Antigen	Posi -tive					
ALT		106				
AST		45				

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VOLVULUS OF THE WANDERING SPLEEN

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Abstract

Wandering spleen is a consequence of the congenital abscence of the dorsal mesogastrium. This Floating spleen is having the risk of torsion and leading to infarction. A case of wandering spleen with situs inversus and complicated by infarction due to volvulus is reported.

Key words : *Mesogastrium, Situs inversus viscerum, Splenectomy, Splenopexy.*

Introduction

Wandering spleen is not a common condition characterized by the absence of ligamentous attachment to the left hypochondrium[1,2]. Here, we report a rare case of wandering spleen in association with situs inversus which is complicated by volvulus.

Case report

A ten year old girl was admitted with right sided upper abdominal pain of two days' duration. She also complained of vomiting in the incipient stages of abdominal pain and it settled on admission. Initial examination revealed she was afebrile, tachycardic and tachypnoeic. She had rigid abdomen with right hypochondrial tenderness.

Blood analysis was normal except increased white cell count and neutrophil leukocytosis. Ultrasound scan of the abdomen showed situs inversus and altered echogenicity of the spleen.

Laparotomy was performed and found to have situs inversus of the stomach, and intestines. The liver and the gallbladder were in the normal position. The spleen was found to be completely free floating without any ligamentous attachments and it was infarcted due to the volvulus (Figure 1). Pus was found in the pelvis. Splenectomy was done and the recovery was uneventful.

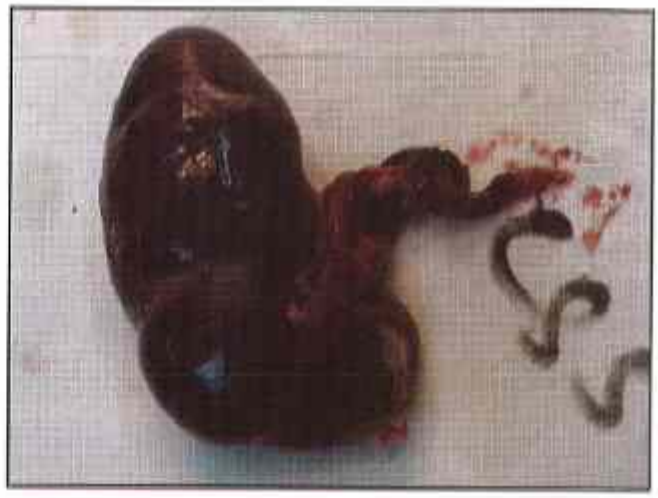

Figure 1. The infarcted spleen (Absence of the dorsal mesentry can be demonstrable in this specimen)

Discussion

The wandering spleen is only attached by an elongated vascular pedicle, leaving it to wander in any part of the abdomen or pelvis[2]. The situs inversus is the shortened form of the Latin"situs inversus viscerum", is a term used to describe the inverted position of chest and abdominal organs[3]. Both are congenital malformations of peritoneal reflections.

Most presentations are in younger than 01year; the male to female ratio is 6:1[2]. In intrauterine life, the failed fusion between the dorsal mesogastrium and the posterior peritoneum, leading to absence of formation of supporting ligaments o spleen [2,4-5]. The malattached spleen has the increased risk of torsion and it can lead to splenic ischemia and infarction[2]. Fifteen percent of patients are asymptomatic, 55% are present with abdominal pain and 90% are presented with palpable mass in the abdomen[2,6]. Sixty four percent of paediatric patients complicated with torsion[2]. The splenic torsion is usually clockwise[2].

The abdominal sonography is the preliminary study to find out the absence or presence of the spleen in the left

hypochondrium[1,2]. Inaddition, a duplex study can be conducted to assess the splenic blood flow[2]. Computed tomography (CT) determines the location of the spleen and the malposition of surrounding viscera such as situs inversus, gastric malposition[3]. Inaddition, infarction of the spleen and involvement of the pancreatic tail and surrounding fat also can be appreciated in CT[2].

Splenopexy is the surgical procedure to prevent torsion when a viable wandering spleen is found at surgery[2].

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Key points

The diagnosis and the viability of the wandering spleen can be made on the basis of the sonography and duplex study of the abdomen. But, volvulus deserved emergency laparotomy to perform splenopexy or splenectomy according to the viability of the spleen.

NON - FAMILIAL CHERUBISM

R.P. Gallage

Abstract

Cherubism is a rare self-limiting benign disease that usually manifest as a bilateral bone enlargement of maxilla & mandible in childhood. Although this is generally inherited as autosomal dominant trait, there are very few solitary sporadic cases of cherubism in the medical literature. Clinical presentation, radiological features & histological appearance of a solitary sporadic case of such entity are discussed here.

Keywords: Cherubism, Maxilla, Mandible, Radiolucency

Introduction

Cherubism is characterized by painless bilateral expansion of mandible and/or maxilla that results in a fullness of cheeks, protuberant intraoral alveolar mass, and displaced or missing teeth.Cherubism is inherited with a penetrance of almost 100% in males and nearly 50-75% in females1. Cherubism was first described by William Jones in 1933 as a familial multilocular cystic disease of the jaws accompanied by swelling of the cheeks. There are only around 300 cases have been published in the literature. The characteristic facial appearance with chubby round face & heaven looking eyes, which resemblance of the cherubs portrayed in Renaissance art, has given its name. Usually the disease presents before the age of 5 years & regress after puberty. Radiologically, this is characterized by multilocular radiolucencies with a soap bubble appearance in mandible, maxilla & occasionally in the ribs. Histopathological evaluation of the lesions shows proliferating fibrous connective tissue containing numerous multinucleated giant cells, which are osteoclasts.

While advancing age, the initially fibrous tissue is replaced by bony structures. Therefore, treatment is usually conservative, allowing natural regression to occur. If surgical re-contouring of expanded bone is necessary, it is best to defer it until after puberty.

Case Presentation

A 9 years old female child of non-consanguineous parents presented to Oral & Maxillofacial Clinic of North Colombo Teaching Hospital in July 2015 with a complaint of bilateral lower facial painless swelling for 4 years. Initially there was mild swelling and that has been gradually increased to present condition over last 4 years duration. Past medical history as well as past surgical history was negligible. There was no history of significant facial deformities in her parents or siblings. She was generally healthy on physical examination with normal intelligence.

On extra-oral examination, there was mild facial asymmetry with mild swelling of bilateral cheeks, more prominent on the right side, which was seen to involve angle of the mandible region. [Figure 1a-1d]. Skin over the swollen area was normal in color, texture as well as freely mobile & intact. Bilateral submandibular lymph nodes were slightly enlarged, mobile and non-tender. Intraorally cortical expansions were noticed in relation to right side posterior buccal regions of mandible as well as in lesser amounts on maxilla also.

A differential diagnosis of Cherubism, Fibrous Dysplasia, Central Giant Cell Granuloma and Hyperparathyroidism was proposed on clinical ground. Routine hematological investigations and biochemical investigations of serum calcium, AST, ALT levels did not reveal any related abnormality

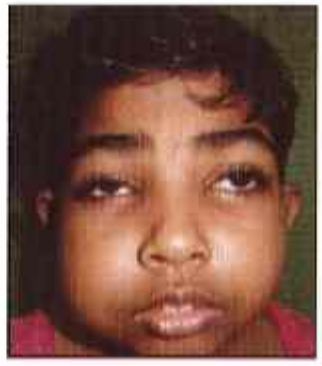

Figure 1a: Frontal View

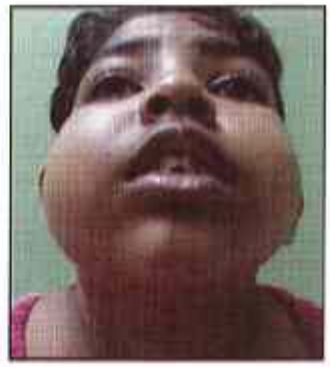

Figure 1b: Worm Eye View

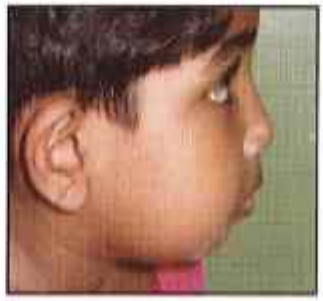

Figure 1c: Right Lateral View

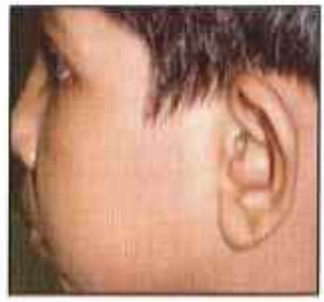

Figure 1d: Left Lateral View

Figure 2: Panoramic View Radiograph

A panoramic view radiograph was taken & found to have variably expansile, osteolytic lesions involving bilateral angle as well as ramus regions of the mandible sparing condyles. Similar type of lesions was involving the bilateral maxilla while some upper & lower permanent teeth were impacted [Figure 2].

Due to negative family history of similar diseases, an incisional biopsy was done intraorally under general anesthesia and a sample was taken from the right posterior mandible.

Histopatholgical studies revealed a lesion with collection of numerous multinucleated giant cells in a fibrocellular stroma.

Therefore, a definitive diagnosis of Cherubism was concluded on clinical, radiological & histopathological features. Conservative management was initiated with follow-up, considering age of the patient, severity of the disease and self-limiting behavior of the disease.

Discussion

Cherubism is a dominantly inherited syndrome characterized by excessive bone resorption in the jaws and accumulation of inflammatory/fibrous tissue in the lower face.3 The disorder was mapped to chromosome 4p, and able to identify amino acid missense mutations in the SH3-binding protein SH3BP2 in several cherubism families and tumor necrosis factor a (TNF-a)-dependent auto-inflammation as a major cause of the disorder.3 There are some cases without a family history in the literature; those cases without family history, just like in our case, may be due to incomplete penetrance or new mutations.

Symptoms and signs depend on the severity of the condition, and range from no clinically or radiologically detectable features to grotesquely deforming mandibular and maxillary overgrowth with respiratory embarrassment and impaired vision and hearing.4Our patient had bilateral lower facial swellings, intraoral posterior mandibular & maxillary buccal expansion with some impacted permanent teeth without the involvement of respiration, vision or hearing.

Histological investigations are essential to conclude the cherubism from other differential diagnosis as in our patient.

Since the lesion undergoes spontaneous regression, the surgical intervention is usually delayed until after puberty. However, in patients with functional or cosmetic problems or emotional disturbances, surgical intervention can be considered. A few recent studies have shown effective usage of calcitonin & steroids.5 We managed our patient conservatively expecting spontaneous regression after puberty.

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