ALEXIS Pulse

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THE EDITORIAL TEAM

FROM EDITORIAL DESK

Dear Doctors,

Greetings from Alexis Multispecialty Hospital, Nagpur!!!

We sincerely hope that you are doing well and everything is fine while you receive this communication from us. With great pleasure and humble pride we would like to share that Alexis Multispeciality Hospital is the first and only hospital in Central India and one of the 39th Hospital in all over India to receive the prestigious Joint Commission International (JCI) Accreditation, USA, which is one of the most respected Quality Certification in the world. This accreditation has placed Nagpur on the Global Map of Quality Healthcare Delivery and we feel pride in putting this feather in Nagpur's crown. In the short span of less than three years in Nagpur we have added healthcare quality to more than 76,000 families and we stand committed in continuing our legacy of being partner in good health for every healthcare seeker in future as well.

We truly feel privileged to present the second edition of our clinical journal 'Alexis Pulse' which encompasses the most critical patients being successfully managed by our team of Clinical Experts at Alexis Multispeciality Hospital, Nagpur.

We hope that these case studies will add a clinical value to your day-to-day practices and can help you to guide your patients effectively.

Please share your views or suggestions about 'Alexis Pulse' on info@alexishospital.com and we will try to incorporate the same in our next edition.

With Warm Regards,

Dr. Tushar Gawad
Senior Hospital Administrator

SUCCESSFUL SURGICAL MANAGEMENT OF HUGE CERVICOMEDIASTINAL GOITRE

A CASE REPORT BY:

DR. SURAJ AGRAWAL, DR. PUNEET JANDIAL, DR. NIMISHA MRINAL, DR. MANISH PUTTEWAR, DR. JAYANT KELWADE

ABSTRACT

Retrosternal goitre (RG) is defined as a goitre with a portion of its mass ≥ 50% located in the mediastinum (1). Surgical removal is the treatment of choice and, in most cases; the goitre can be removed via a cervical approach. Less than 2% patients of retrosternal goitre require sternotomy for their removal. We present case of retrosternal goitre, reaching beyond the tracheal bifurcation, which was successfully managed by surgical removal through combined neck and sternotomy approach. Preoperative identification of patients with retrosternal goitre, who will require sternotomy during surgery, is vital for ensuring good outcomes.

INTRODUCTION

Retrosternal goitres can be classified as either primary or secondary. Primary intra-thoracic goitres arise from aberrant thyroid tissue which is ectopically located in the mediastinum, receive their blood supply from mediastinal vessels and are not connected to the cervical thyroid. They are rare, representing less than 1% of all retrosternal goitres (2).

Secondary retrosternal goitres develop from the thyroid located in its normal cervical site. Downward migration of the thyroid into the mediastinum is facilitated by negative intra-thoracic pressure, gravity, traction forces during swallowing and the presence of anatomical barriers preventing the enlargement in other directions (thyroid cartilage, vertebral bodies, strap muscles, especially in patients with a short, large neck). These secondary retrosternal goitres receive their blood supply, almost always through branches of the inferior thyroid artery. This case report describes a case of secondary retrosternal goitre who presented with respiratory difficulty due to severe compression of trachea.

CASE REPORT

57-year-old man presented to Alexis Hospital with complains of neck swelling since 6 years. Swelling was gradually increasing in size over the years. Patient also experienced gradually increasing difficulty in breathing while walking since last 6 months. Patient was previously seen at multiple hospitals in Nagpur and was diagnosed as a case of Colloid Goitre of thyroid with large retrosternal extension. Patient had hyperthyroidism and was taking anti-thyroid medications for one year. Patient was refused surgery at many hospitals due to high-risk nature of the case.

At Alexis Hospital, patient was evaluated by a multi-disciplinary team of Oncosurgeon, Cardiac Surgeon, ENT Surgeon, Anaesthetist and Endocrinologist. Patient's CT scan was done at Alexis Hospital and it showed large diffuse goitre arising from thyroid gland, more on the left side than right. Goitre was extending into the mediastinum behind the trachea till the tracheal bifurcation, going 2-3 cm below the arch of aorta. There was severe compression of trachea, resulting in narrowed airway and respiratory difficulty. Trachea was severely pushed to the right side by the goitre. Goitre was abutting the arch of aorta and all its major branches were pushed away by the mass.

Patient and his family were counselled in detail about the risks involved in the surgery. After obtaining the informed consent and after thorough preoperative workup, patient was posted for surgery.

On 1st August 2019, patient underwent Total Thyroidectomy through Sternotomy approach, by joint effort of Oncosurgeon and CVTS Surgeon. During surgery, Median Sternotomy was done by CVTS surgeon to provide access to the tumour. Thyroid goitre was then carefully dissected out by the Oncosurgeon, carefully preserving all the important structures. Left recurrent laryngeal nerve was coursing through the goitre and had to be sacrificed. Right recurrent laryngeal nerve was identified and carefully preserved. Both parathyroid glands were preserved on right side. Superior parathyroid gland was preserved on left side however inferior parathyroid could not be identified on left side. Blood loss during surgery was 350ml. Patient was extubated in OT itself and shifted to ICU for post-operative monitoring. Bronchoscopy was performed in OT before removal of endotracheal tube to rule out tracheomalacia.

Post-operatively, patient had no respiratory difficulty and he maintained good oxygen saturation on room air.

He was noted to have hoarse voice on post-operative day 1 and direct laryngoscopy conformed that left vocal cord is not mobilising and is in paramedian position. Post-operatively, he had mild transient hypocalcaemia, which was easily managed with oral calcium supplements. His postoperative recovery was otherwise uneventful and he was successfully discharged on post-operative day 6.

DISCUSSION

RGs show a slow-growing enlargement, which usually remains asymptomatic for many years. The most common symptoms are related to compression of the airways and the oesophagus, and less commonly, signs of compression of vascular and nervous structures are present. Computed tomography (CT) scanning is, at present the most exhaustive examination for assessment of the extent of the goitre and compression effects on adjacent anatomical structures. There is general agreement that surgical removal is the treatment of choice for RG, even in the absence of clinical symptoms. There are numerous reasons for performing surgery in such cases:

1. Non-surgical treatment of RG with thyroid hormone or radioactive iodine ablation is very rarely successful
2. RG can become a life-threatening emergency if there is a sudden enlargement of the goitre, secondary to haemorrhage or malignant change
3. A diagnosis of malignancy, reported in 3-21% of RGs could be missed, considering the difficulties and potential dangers in performing fine-needle aspiration cytology in the mediastinal portion of a RG

Most RGs can be removed through a cervical approach, while a partial or total sternotomy should be performed only in a minority of patients, ranging between 1- 11%.

Many attempts have been made to specifically define the factors increasing the likelihood of sternotomy, but a general consensus has still not been reached.

Flati et al. in 2005, defined the sternotomy approach as 'inevitable' in the presence of an iceberg shaped RG with >70% of the mass lying in the mediastinum (3).

More recently, Cohen (4) identified four factors significantly increasing the need to perform sternotomy:

1. Presence of malignancy
2. Involvement of the posterior mediastinum
3. Extension of the goitre below the aortic arch
4. Presence of ectopic goitre.

Careful CT assessment of RG is mandatory before surgery to identify cases where sternotomy approach might be required. Preoperative identification of such cases enables early involvement of Thoracic surgeon in the case and allows room for better planning and consent process. It is well known that post-operative morbidity is more common in patients undergoing RG resection than in patients undergoing thyroidectomy for cervical thyroid disease (5). So, it is imperative that such cases are managed only in centres having adequate expertise and necessary infrastructure to achieve best outcomes.

CONCLUSION:

Removal of a RG is a challenging surgical procedure, presenting a slightly higher risk of complications, especially hypoparathyroidism and post-operative bleeding. An attempt to remove the goitre through the cervical incision should always be made, using all the techniques available, on account of less risk of surgical and aesthetic damage obtained with this approach. The most significant criteria for selecting patients requiring sternotomy are CT scan features, in particular presence of an ectopic goitre, thyroid gland volume and extension of the goitre to or below the tracheal carina. Therefore, the CT scan should always be included in the pre-operative diagnostic workup, when RG is suspected.

REFERENCES:

1. Katlic MR, Wang CA, Grillo HC. Substernal goiter. Ann Thorac Surg 1985;39:391-9.
2. Wu MH, Chen KY, Liaw KY, Huang TS, Lee PH. Primary intrathoracic goiter. J Formos Med Assoc 2006;105:160-3.
3. Flati G, De Giacomo T, Porowska B, Flati D, Gaj F, Talarico C, et al. Surgical management of substernal goitres. When is sternotomy inevitable? Clin Ter 2005;156:191-5.
4. Cohen JP. Substernal goiters and sternotomy. Laryngoscope 2009;119:683-8.
5. Abboud B, Sleilaty G, Mallak N, Zeid HA, Tabchy B. Morbidity and mortality for substernal goiter. Head Neck 2009;13 [Epub ahead of print]

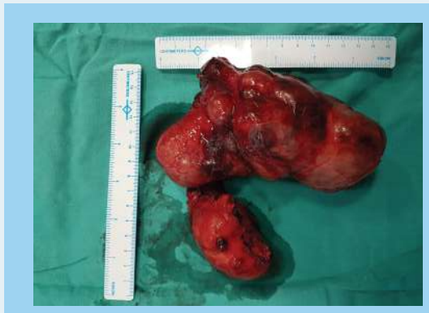


Figure: Specimen of total thyoidectomy

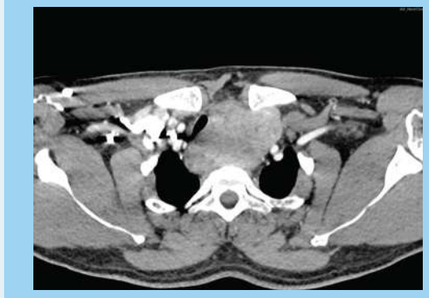


Figure: Preoperative CT scan showing Retrosternal Goitre

TWIN REVERSED ARTERIAL PERFUSION SYNDROME (TRAP) WITH CONGENITAL HYPERTROPHIC PYLORIC STE

A CASE REPORT BY:
DR. RASHMI SHINDE AND DR. SHWETA BHANDARKAR

ABSTRACT

Twin reversed arterial perfusion (TRAP) syndrome is a rare condition (0.3:10,000 births) that occurs in monochorionic twin pregnancies, resulting in coexistence of a normal 'pump' twin and an acardiac twin. The acardiac twin is dependent upon the normal twin to provide circulation by means of vascular anastomosis. Many of the bizarre defects are felt to be caused by low oxygen tension and this causes dramatic alteration in the twin fetal physiology and high prenatal mortality. Overall only 50% of pump twins survive. We present a case in a nineteen year old primigravida, 33 weeks of monochorionic diamniotic pregnancy with TRAP syndrome. In our case fetus one was having acardia and acrania and fetus two with severe intrauterine growth restriction. Following delivery twin one showed no signs of life. Twin two, the pump twin had all features of twin to twin transfusion which survived well. Later was found to have cinfantile hypertrophic pyloric stenosis for which the neonate was operated at day 26 of life with weight of 1. 34 kg. Baby tolerated the procedure well and was discharged from NICU on day 50 of life in good health.

INTRODUCTION

TRAP sequence is a rare condition, unique to identical twins. Overall TRAP affects about 1% of all monozygotic twins with 75% occurring in monochorionic diamniotic twins, remaining 25% in monoamniotic twins. TRAP is characterised by normal appearing pump twins that sustains a dysmorphic acardiac twins through aberrant arterio-arterial anastomoses within the shared single placenta. This case report narrates obstetric history and outcome of 19 years old primigravida who was unaware of her twin pregnancy and later on found to be suffering from TRAP syndrome. Above all the pump twin which survived had a rare anomaly of pyloric stenosis present at 26 day of life operated and sustained the surgery well

CASE REPORT

Mrs J, 19 years old primigravida, married life of one year, non consangious marriage, at 26 weeks of gestational age came to OPD for further management. She came with a scan report from outside suggestive of single live intrauterine fetus of 22 weeks of gestational age. But as the height of uterus was more than period of gestation, suspicion of polyhydramnios, twin gestation was made. An ultrasound was advised showing diamniotic monochorionic pregnancy with fetus A (acrania and acardia), fetus B (Live Intrauterine pregnancy of 22 weeks, 4 days). The diagnosis of monochorionic diamnionic pregnancy was made with fetus B structurally normal, and fetus A acardiac. Serial follow up scans along with a fetal medicine specialist opinion was taken. At around 31 weeks of gestation fetus B developed severe oligohydramnios and fetus A which is acardiac developed polyhydramnios. Procedure of amnioreduction was done for fetus A with amnioinfusion of fetus B with septostomy of separating membrane. Steroid was given for foetal lung maturity.

At 32 weeks of gestation, patient developed severe gestational hypertension without proteinuria. She was admitted and started on 2 antihypertensives and discharged after stabilization. It was planned to deliver by 34 weeks of gestation in view of MCDA pregnancy and severe gestational hypertension. But at 33 + 3 weeks, patient presented with premature rupture of membrane. On admission, her blood pressure was 180/100 with headache. PIH profile was deranged. A diagnosis of TRAP syndrome/premature rupture of membrane with HELLP syndrome was made. Patient was put on prophylactic magnesium sulphate. Antihypertensive were titrated. An emergency caesarean section was done. She delivered preterm babies, Fetus A – (ACARDIAC - WITH NO SIGNS OF LIFE). Fetus B -1 KG (SEVERE IUGR) Post-operation patient was shifted to high dependency unit for further management. She recovered well and was discharged on day 6. The PUMP twin, weight of 1 kilogram cried weakly after birth was stabilised and immediately shifted to NICU. As expected, the pump twin had signs of severe intrauterine growth restriction with respiratory distress due to cardiac dysfuction, metabolic complications (hypoglycaemia, hypocalcemia hypomagnesemia), thrombocytopenia and polycythemia. It managed with non-invasive ventilation, strict fluid management, partial exchange, antibitoics and phototherapy. Baby was on full feeds by day 9 of life. In NICU on 24 day of life baby developed non bilious persistent vomiting on day 26, on evaluation found to have infantile hypertrophic pyloric stenosis. For which she was operated at 1.4 kg by Paediatric surgeon. Responded well to the intervention and discharged later at 1.7kg on day 54 of life.

DISCUSSION

The incidence of TRAP syndrome is 1 in 35,000 pregnancies, 1 in 100 monozygotic monochorionic twin gestations and 1 in 30 monochorionic triplet gestations. Most cases are seen in twins with only 8% in triplets. The risk in multiple pregnancies of a higher order is even greater. Exact pathogenesis is not known. One theory explains that umbilical vascular anastomosis in early embryogenesis in fused placenta results in vascular disruption, early tissue hypoxia in one twin giving rise to atrophy of heart and other organs. Other theory postulates that severe genetic/other primary cardiac embryogenesis defect first cause the failure of cardiac development in one twin followed by anastomosis between umbilical vessels allowing survival of affected twin. In one of the case reports published, association between TRAP and VACTERL was mentioned, thus indicating that this defect occurs in early embryogenesis. Monochorionicity always highlights risk of TRAP. In our case it was monochorionic and diamniotic twins. Chromosomal analysis was not done in our case. The acardiac twin becomes dependent on the perfusion of the 'Pump' twin. The upper half of the body of an acardiac twin is extremely poorly developed and sometimes not developed at all. Head, cervical spine and upper limbs are usually absent. Oedema in the upper body consistent with cystichygroma is common. In contrast the lower half of the body although malformed is better developed. This is because of mechanism of perfusion of the acardiac twin where blood enters via abdominal aorta which is deoxygenated blood, that leaves the normal twin. So the oxygen and nutrition available is extracted allowing some development of caudal aspect. Once blood enters upper torso in retrograde fashion oxygen saturation is extremely low, halting development of heart, head and upper torso giving rise to severe deficits. The pump twin suffers from high output cardiac failure which is directly proportional to size/weight of the acardiac twin. The anomalies in acardiac twin are partial/total absence of cranial vault, holoprosencephaly, absent facial structures, anophthalamia, micro-ophthalmia, cleft lip, cleft palate, absent or rudimentary limbs, diaphragmatic defects, absent lungs & heart, oesophageal atresia, ventral wall defects, ascites, absent liver & gall bladder, oedema of the skin, single umbilical artery (75% cases), markedly different sized umbilical artery, inconsistent membrane development between the twins and occasionally umbilical artery drawn directly into superior mesentric artery. Congenital anomalies are present in about 9% of pump twins; however pump twin presents with ascites, pleural effusion, polyhydramnios and skin oedema which are features of congestive heart failure. Prenatal diagnosis by ultrasound is suspected when a twin gestation presents with discordance and bizarre malformations with retrograde blood flow in acardiac twin demonstrated by pulsed/colour flow Doppler. Anencephaly, cystic hygroma, conjoined twins, twin demise and intramniotic placental tumours are differential diagnosis to be ruled out. Recurrence of TRAP syndrome is unknown/not increased, likely to be low and the couples can therefore be counselled optimistically for future pregnancy. The prognosis in TRAP syndromes is lethal for acardiac twin. The fetal mortality of the pump twin is extremely high [50-75%] because high output cardiac failure. In our case the pump twin survived well. Infantile pyloric stenosis was managed with Ramsted's pyloromyotomy. At present the baby is doing well with good weight gain and neurological development.

CONCLUSION

The obstetrician and radiologist should be aware of this anomaly especially in twin/multiple pregnancies so that timely proper measures can be taken to survive the pump twin. 1st trimester scan is very important for the diagnosis of twin pregnancy as well as to know chronicity of twin pregnancy. Early diagnosis and good neonatal care gives better outcome.

REFERENCES:

1. R K, Bindra MS. Twin Reversed Arterial Perfusion Syndrome (TRAP or Acardiac Twin)-A Case Report. J Clin Diagn Res. 2014;8(1):166–167. doi:10.7860/JCDR/2014/7012.3965
2. Kariappa TM, Chidananda HT, Mamatha R. Acardiac twin: an unusual case report. Indian J Pathol Microbiol. 2007Oct;50(4):801–3.
3.Hanafy A, Peterson CM. Twin Reversed arterial perfusion (TRAP) sequence: case report and review of literature. Aust N Z J Obstet Gynaecol. 1997 May;37(2):187–91.

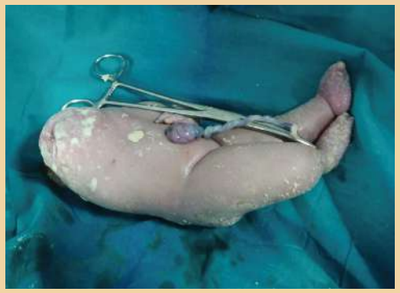


Figure : Fetus A

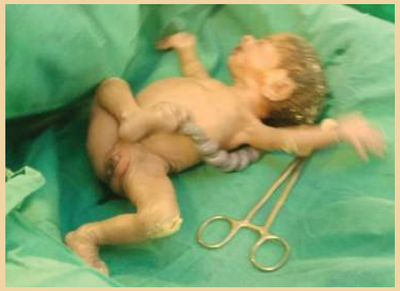


Figure: Fetus B

HARTNUP DISEASE – A RARE DERMOMETABOLIC DISORDER

A CASE REPORT BY:
DR. BHASKAR GUPTA

ABSTRACT

Dermometabolic disorders due to inborn errors of metabolism in children can manifest in a variety of ways. Disorder due to Tryptophan metabolism being rare can cause symptoms which can be confused with a variety of skin diseases. One such disorder of Tryptophan metabolism which is autosomal recessively inherited known as Hartnup disease can be very distressing to the child and family and can cause severe morbidity and death due to complications. The characteristic findings are pellagra like dermatitis following exposure to sunlight, intermittent cerebellar ataxia, psychosis and constant aminoaciduria. A proper history with clinical findings and investigations can diagnose this disorder and with proper medications the flare ups and complications can be minimised. A case of Hartnup disease in which the child presented repeatedly with severe illness and his elder sibling had died due to similar presentation was thoroughly examined and investigated and thereby diagnosed and got relieved with due medications at Alexis Hospital, Nagpur; thus highlighting the need to be alert with regards to various inborn errors of metabolism in children.

INTRODUCTION

Hartnup disease is an aminoaciduria that is usually clinically silent but can manifest with severe and intermittent episodes of pellagra like rash, cerebellar ataxia and psychosis. The reported incidence is 1 in 30,000 persons and is autosomal recessively inherited due to mutations in SLC6A19 gene coding an enzyme that transports neutral amino acids across the apical membrane of epithelial cells in the gut and kidneys. Due to this large amounts of neutral amino acids including tryptophan are present in urine establishing the diagnosis. A 3 year old child, presented with severe pellagra like skin lesions since the age of 9 months and was repeatedly hospitalised due to its complications and had failure to thrive. He was examined and investigated. With a simple medication of Nicotinamide tablets daily his symptoms were alleviated and he started with good growth and development. This case highlights the importance of always keeping in mind the rare inborn errors of metabolism in children which if diagnosed properly can reduce the mortality and morbidity.

CASE REPORT

A 3 year old boy presented with multiple severe excoriating hyper-pigmented lesions on flexural aspect all over body, perineum and neck, hands and feet with mild itching. The first episode was at 9 months of age with irritability, poor-feeding, fever and lesions and was admitted to hospital in Chhattisgarh. He was investigated and treated with emollients, antibiotics and nutritional supplements and was provisionally diagnosed as staphylococcal scalded skin syndrome. The lesions gradually reduced but continued off and on till the time at age of 16 months he again had severe flare ups with poor feeding, stomatitis, cough and irritability and was taking oral antibiotics. He was admitted again at another hospital and treated with provisional diagnosis of Steven Johnson's syndrome due to the reaction of antibiotic that had been consumed. The skin lesions gradually reduced and his third admission at age of 2 years was with diarrhoea, fever and similar excoriating skin lesions. This time he was being treated with provisional diagnosis of Acrodermatitis enteropathica and was given steroids and zinc although the Serum and Urine zinc levels sent were normal.

He was presented to Alexis Hospital in February 2018 with extensive excoriation, abdominal pain and anorexia. He was admitted and found to have weight of 11 kgs which is below 10th centile for expected age, erythematous plaques over thighs, hands and legs, perineum, ankles with bleeding at some spots and excoriation with hyperpigmentation. He was kept on IV fluids, emollients, zinc, and anti-allergics with antibiotics. His history revealed that he was born as full term gestation with birth weight of 2.8 kgs with history of consanguinity and slightly delayed developmental milestones with emotional instability and irritability and mood swings. His examination revealed that he had sunlight exposure exacerbation and hyperpigmentation with lichenification and stomatitis. Also he was found to have mild gait instability and nystagmus. His investigations revealed high WBC count with neutrophils of 62% normal Liver function tests and ASO titre of

27, normal Calcium, phosphorous and alkaline phosphatase and low vitamin D levels. His Serum Zinc was 101 which were within normal limit and his urine for porphobilinogen was negative.

A detailed history revealed that his elder sibling presented with similar recurrent skin lesions, with poor nutrition, unstable gait, uncontrollable and irritable behaviour; died at age of 3 years.

With due care his lesions improved and he was advised follow up after 4 weeks. At follow up again similar skin lesions were noted and his urine for Amino acid was sent which showed aminoaciduria with excess excretion of neutral amino acids alanine, serine threonine, tryptophan, valine leucine isoleucine and excretion of basic and acidic amino acids like proline, hydroxyprolene, and arginine were normal.

The clinical presentation of recurrent extensive photosensitive dermatitis with extensive lichenifation and emotional instability with nystagmus with aminoaciduria suggested a diagnosis of Hartnups disease. He was put on Tablet Nicotinic Acid 250 mg (Nicoglow) per day with high protein diet, Emollients and nutritional supplements with advice to avoid sun exposure and was followed up after 6 weeks. Subsequent follow ups at 6 weeks, 3 & 6 months and 1 year later revealed no skin lesions, good weight gain and appetite with reduced irritability.

DISCUSSION

Acute dermatitis and blistering with secondary crusting and scarring with lichenification and hyperpigmentation following sun exposure are seen in pellagra which is due to Vitamin B3 deficiency and is uncommon now-a-days with triad of dermatitis, diarrhoea and dementia. This child presented with neurological features along with glossitis, stomatitis, nystagmus, emotional disturbance and aminoaciduria with familial occurrence thus suggestive of the diagnosis of Hartnup disease. The lesions are exacerbated during the period of malnutrition and intercurrent infections.

Hartnup disease is autosomal recessive in inheritance with the genetic defect in 5p15.33 locus encoding for SLC6A19 this gene was identified in the year 2004. The estimated incidence is 1 in 30,000 persons. The metabolic aberration in hartnups disease results from an error in the transport of monoamino monocarboxylic neutral amino acids that affects renal tubular reabsorption and intestinal absorption. There is deficient transport of neutral amino acids including tryptophan and thus large amounts of amino acids are excreted in urine. Large amounts of tryptophan remains in intestinal lumen where it is converted to Indolic compounds by bacteria and then absorbed. These compounds are toxic to Central nervous system thereby causing neurological manifestations. Large amounts of Indican excretion is noted which can be detected by urine chromatography. Lack of tryptophan absorption leads to niacin deficiency which results in pellagra like symptoms and photosensitivity.

Thus the characteristic features of Pellagra like dermatitis following sunlight exposure with exacerbations during infections and poor nutrition along with intermittent cerebellar ataxia, neurological and emotional features with glossitis and stomatitis and constant aminoaciduria are characteristic features of Hartnup disease. Affected children are normal at birth but may have delayed milestones.

The differential diagnosis is acrodermatitis enteropathica which is an autosomal recessive nutritional dermatoses due to defective absorption of zinc from intestine presents with reddish hairs, alopecia blepharitis, corneal dystrophy poor wound health and zinc levels are very low.

Staphylococcal scalded skin syndrome known as Ritter's disease due to phage group 2 Staphyloccal strains 71 & 55 due to exfoliative toxins A & B presents in kids as bullous impetigo to generalised peeling and Nikolsky sing positive.

Biotindase deficiency is due to defective enzyme for degradation of valine isoleucine and lucine resulting in atopic dermatitis, alopecia, myoclonic seizures and hearing loss and diagnosed by enzyme estimation in serum.

CONCLUSION

Hartnup disease is an inborn error of tryptophan excretion which is named after the Hartnup family in whom it was first noted and is the second most inherited aminoaciduria after phenylketonuria. The intermittent nature of disease in the form of pellagra like dermatitis to face, neck, hands and legs with neurological manifestations and psychological disturbances are characteristic. Early suspicion of this disease is vital in reducing the mortality and morbidity and thereby nutritional and neurological outcome. A simple tablet of Nicotinamide daily with proper diet and avoiding exposure to sunlight is essential part of treatment with excellent results.

BIBLIOGRAPHY

Broer S: Disease associated with general amino acid transporters SLC 6. Curr Mol Pharmocol 2013;6;pp74-87

Nelson Textbook of Paediatrics. 19th edition Iraz Rezwani chap 79.5 pp 429-430

Scriver C R Mahon B Levy HL The Hartnup phenotype multifactorial disease. Am J Hum Genet 1987;40 pp 401

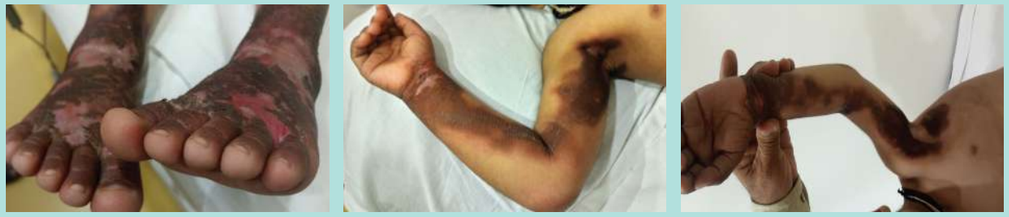


Figure: Healing lesions with only hyperpigmentation one month after starting treatment

Figure: Healing lesions with only hyperpigmentation one month after starting treatment

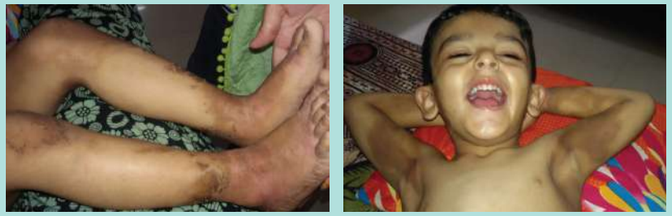


Figure: Healing Lesions after Treatment for 3 month

SUDDEN CARDIAC ARREST ATTRIBUTED TO VENTRICULAR ARRHYTHMIA IN ACUTE MYOCARDIAL INFARCTION SUCCESSFULLY TREATED

A CASE REPORT BY:

DR. SHOEB NADEEM, DR. RISHI LOHIYA, DR. SANJAY KRIPLANI, DR. RAKESH BHAISARE, DR. TAUSIQ QAZI

ABSTRACT

Sudden cardiac arrest has got a very poor prognosis and very low rates to hospital discharge. We present two cases of out of hospital sudden cardiac arrest, who were successfully resuscitated in hospital and discharged to home. Both were young males who had acute anterior wall myocardial infarction, given thrombolytic therapy after being resuscitated in emergency room, one had recanalised infarct related artery and another required angioplasty with stent, and were discharged to home without any assistance.

CASE REPORT

Two young males 32 and 36 year old respectively presented to emergency department at Alexis Hospital within 10 minutes interval, with similar history of chest pain at home about 30 minutes ago and collapsed while being transported to hospital. One of the patients received some resuscitative effort by the relatives who brought him, in the form of mouth to mouth breathing and chest compressions. They were brought to hospital in a state of cardiac arrest with no palpable pulse, and gasping breathing, with monitor showing ventricular fibrillation, were immediately defibrillated to sinus rhythm, cardiopulmonary resuscitation given according to ACLS protocol with active involvement of emergency room and code blue team, after return of spontaneous circulation ECG showed right bundle branch block with ST elevation in anterior leads. Patients were in cardiogenic shock and required high inotropic support, relatives were given option of thrombolysis or primary angioplasty, they opted for thrombolysis. Both of them were thrombolysed with Reteplase and started on heparin infusion, inotropes, IV fluids and diuretic infusion, IV lignocaine and induced hypothermia. One patient had rapid resolution in ST elevation on ECG, and early tapering of inotropes was possible, was extubated the next day. His coronary angiography done after stabilisation showed recanalised left anterior descending coronary artery. He was discharged home after 5 days. The other patient had persistent pulmonary edema with poor resolution in ST segments, had acute kidney injury, recovered gradually and extubated on third day, his coronary angiography revealed total occlusion in proximal left anterior descending coronary artery, and required angioplasty with stent to left anterior descending coronary artery. He was discharged on 8th day. Both patients had complete recovery with no neurologic disability. Follow up echocardiography showed near total recovery in left ventricular function in the first patient and significant improvement in the second patient.

DISCUSSION

Cardiovascular disease (CVD) is a leading cause of global mortality, accounting for almost 17 million deaths annually, which is 31% of all global mortality. It is estimated that about 40-50% of all cardiovascular (CV) deaths are Sudden Cardiac Deaths (SCD) and about 80% of these are caused by ventricular tachyarrhythmias. About 6 million SCDs occur annually due to ventricular tachyarrhythmias. Sudden cardiac death remains a major public health problem. The survival rate from sudden cardiac arrest is less than 1% worldwide and close to 5% in the United States (US)1. The time from symptom onset to emergency department arrival for patients with acute ST elevation myocardial infarction ranges between 110 and 140 minutes in North America, while in India, it is 180-330 minutes[2,3]. In our case the patients were brought to hospital within 30 minutes of symptom onset. This delay in presentation is attributed to several factors such as lack of symptom awareness, longer distances travelled to reach hospital and problems of transportation. Only 5.4% of patients are brought to hospital in an ambulance, with the large majority using public transport and hired vehicles4. Time-to-treatment is critical when considering the chance of survival for an SCD victim. 95% of those who experience SCD die because they do not receive life-saving defibrillation within 4 to 6 minutes, before brain and permanent death start to occur5,6 .Our patients received immediate defibrillation upon arriving to hospital. In India, most people do not know how to respond when someone collapses suddenly. One of our patients had received basic life support by the accompanying relatives. This experience emphasises the need to create public awareness of symptoms of acute coronary syndrome, also the need to seek medical attention immediately and public at large should be educated in giving basic life support. Also prompt recognition of arrhythmia in hospital and immediate defibrillation should be the norm, along with high quality cardiopulmonary resuscitation. Also the role of intensivist and induced hypothermia cannot be overemphasised.

CONCLUSION

Sudden cardiac arrest is the leading cause of death worldwide, with majority of cases attributed to ventricular arrhythmia in acute myocardial infarction. Prompt recognition of symptoms, immediate defibrillation, followed by high quality cardiopulmonary resuscitation and restoration of arterial flow is important in saving lives in such patients.

REFERENCES

1. Deo R, Albert CM. Epidemiology and genetics of sudden cardiac death. Circulation. 2012;125(4):620-37.
2. George E, Savitha D, Pais P. Pre-hospital issues in acute myocardial infarction. J Assoc Physicians India 2001;49:320–3.
3. Rajagopalan RE, Chandrasekaran S, Pai M, et al. Pre-hospital delay in acute myocardial infarction in an urban Indian hospital: a prospective study. Natl Med J India 2001
4. Malhotra S, Gupta M, Chandra KK, et al. Prehospital delay in patients hospitalized with acute myocardial infarction in the emergency unit of a north Indian tertiary care hospital. Indian Heart J 2003;55:349–53
5. American Heart Association. Heart Disease and Stroke Statistics — 2007 Update. Dallas, Texas: American Heart Association; 2007
6. Lokhandwala Y, Panicker GK, Deshpande S. Sudden cardiac death-an Indian perspective. CVD Prevention and Control 2009;4:103–108.

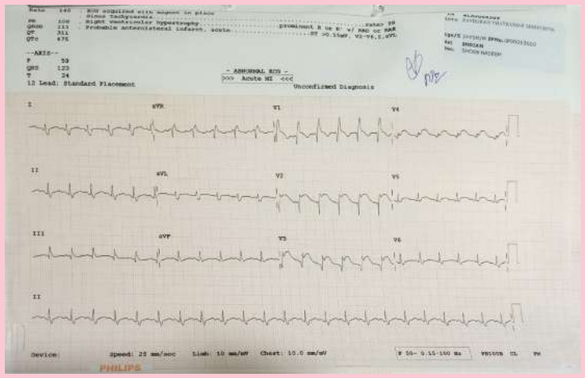


Figure : Fetus A

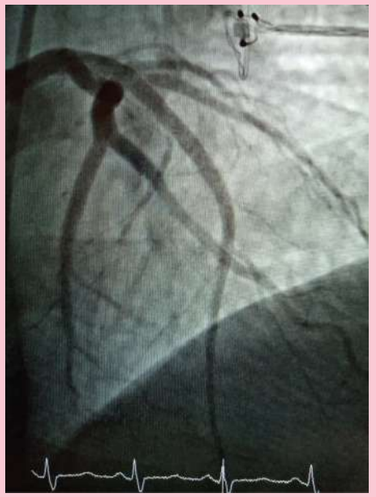


Figure : Fetus A

DENGUE ARDS IN PREGNANCY – A RARE PRESENTATION

A CASE REPORT BY:

DR. SAKSHI GAIKWAD, DR. MANOJ PETHE, DR. NITIN DAMBHARE, DR. SHILPA MOON

ABSTRACT

Dengue fever is a major public health problem in India. Dengue fever is an arboviral infection, causing a wide spectrum of illness. Dengue haemorrhagic fever (DHF) is a more severe form of dengue disease. As dengue infection has become more common, increasing number of DHF cases are associated with unusual presentation. Pulmonary manifestations are unusual in this disease except for pleural effusion. ARDS in DHF is rarely reported in literature. Of great concern is the infection in the pregnant woman. The pregnant woman undergoes great hormonal changes. The change towards greater immunological tolerance allows for a successful pregnancy. Dengue viral fever can be a multisystem disorder, with complement activation and an inflammatory cytokine storm with a spectrum of manifestation; effevescence, defevescence and recovery, related to the days of illness. The respiratory consequences with ventilation - perfusion mismatch and shunt, hypoxia may create the chaos of respiratory distress. Dengue fever encounter in pregnancy may be quite or it can be impending disaster. Dengue infection in pregnancy has risk of haemorrhage for mother as well as new-born. In addition there is risk of premature birth and fetal death and vertical transmission causing neonatal thrombocytopenia that necessitates platelets transfusion. An alert clinician is the most important vital link.

INTRODUCTION

The cases of dengue in pregnancy are on rise due to increasing incidence of dengue fever in adulthood. The sign and symptoms of dengue fever in pregnancy are similar to that in non-pregnant patient. In pregnancy infection with dengue virus is not more severe as compared to non-pregnant patient as in the case, for instance, of malaria. Infection with dengue virus does not increase the risk of foetal malformation or deformity. Dengue fever can affect the unborn baby which include - low birth weight, preterm birth, abortion and death. The endothelium is the target of the immunopathological mechanism in in Dengue and DHF. The hallmark of dengue is vascular permeability and coagulation disorder. These mechanisms can explain varied systemic involvement. Dengue haemorrhagic fever [DHF] can result in acute respiratory distress syndrome (ARDS). The usual criteria for defining ARDS are diffuse pulmonary infiltrates on chest X ray, Pao2/Fio2 of less than 200-250 mm Hg and no evidence of heart failure. Dengue haemorrhagic fever presenting with ARDS in adult has not been reported widely. This case report describes the clinical profile and management in case of Dengue ARDS in pregnancy treated at Alexis Multispeciality Hospital, Nagpur.

CASE REPORT

31 year old female Gravida 2 para 1 previous lower segment caesarean section 34 weeks of gestation, booked case at Alexis Hospital with no previous medical problem, came in OPD with history of high grade fever with chills and rigors since 4 days. Her fever profile was sent and patient was found to have dengue NS1 positive. All other investigations were normal. For the management of dengue complicating pregnancy, patient was admitted in ward. On admission, she was fibrile, pulse rate (100 per minute), temperature - (99degrre F), spo2 - (95 per cent), respiratory rate (20 PER minute). Blood pressure (124/78 mm Hg). Patient was treated with supportive management and was stable. After approximately 3-4 hrs of admission, patient developed sudden onset breathlessness and was complaining of air hunger. On examination, patient was febrile, temperature - 101degree F, pulse -114/min, tachypnic(32/min), SPO2-78% on room air, respiratory system showed decreased air entry bilaterally, bilateral basal crepts were present so patient was shifted to ICU. In ICU, posteroanterior radiograph was done with lead cover over abdomen showed large confluent and patchy air space opacities in perihilar regions with basal effusion. Arterial blood gas (ABG) analysis suggested of hypoxia. So a diagnosis of Dengue ARDS was done. Patient was not able to maintain saturation on O2 mask, so was put on non-invasive ventilation. For fetal well-being 2 doses of injection betnesol were given for fetal lung maturity. Patients other investigations - 2D ECHO - normal, H1N1PCR – normal and procalcitonin - normal. On day 3 of admission, patient developed fetal tachycardia with FHR of 200/min with normal maternal pulse rate showing uterine contractions. In view of persistent fetal tachycardia, decision of emergency caesarean section was taken. Under general anaesthesia, patient delivered a preterm male baby, weight 2.3kg and was shifted to NICU for further management. After procedure, patient could be extubated easily and was further shifted to ICU for further management and was kept on non-invasive ventilation. Patient recovered well in ICU and was shifted to room on Day 2 of LSCS.

DISCUSSION

Dengue has been associated with ARDS as a rare complication. Yet many of the Dengue patients do present with early sign of respiratory distress, tachypnea and dyspnoea. Early arterial blood gas analysis (ABG) may indicate an (A-a) DO2 gradient, the existence shunt fraction with possible pulmonary hypertension. The patient presented with tachypnea and with a hypoxemia, not responding to oxygen therapy alone but required application of positive pressure breathing. An early posteroanterior radiograph revealed large confluent and patcy air space opacities in peripheral regions with basal effusion suggestive of ARDS. Beside ECHO excluded a cardiogenic cause of pulmonary edema, indicating a non-cardiogenic cause of pulmonary edema. Her NS1 was positive for dengue on the 4th day of fever. Acute Respiratory Distress Syndrome (ARDS) was first reported 50 years ago, in 1967 by Ashburgh and Petty. There is a continuing urgency for earlier detection of ARDS ameliorates recovery and return to a normal life.

CONCLUSION

Dengue ARDS is a major cause of maternal morbidity and mortality if not managed accurately. Multidisciplinary team approach by obstetrician, physician and intensivist play a crucial role in the management. Dengue ARDS requires early diagnosis and treatment. High index of clinical suspicion is required to diagnose dengue ARDS as it is very difficult to distinguish from other cases of ARDS in pregnancy like pre-eclampsia, eclampsia, pulmonary embolism, haemorrhage and pneumonitis and H1N1 infection.

REFERENCE

Sen MK, Ojha UC et al .Dengue Haemorrhagic fever (DHF) presenting with ARDS .Indian J Chest Dis Allied Sci, 1999;41:115-9

Phupong V Dengue fever in pregnancy; a case report, BMC Pregnancy Childbirth 2001;1(1):7

Raymond O Powrie,Michael.F.Greene and William Cammon.De Swiet,S 2004

WHO regional office for South East Asia compressive Guidelines for prevention and control of Dengue and DHF WHO, New Delhi 2011

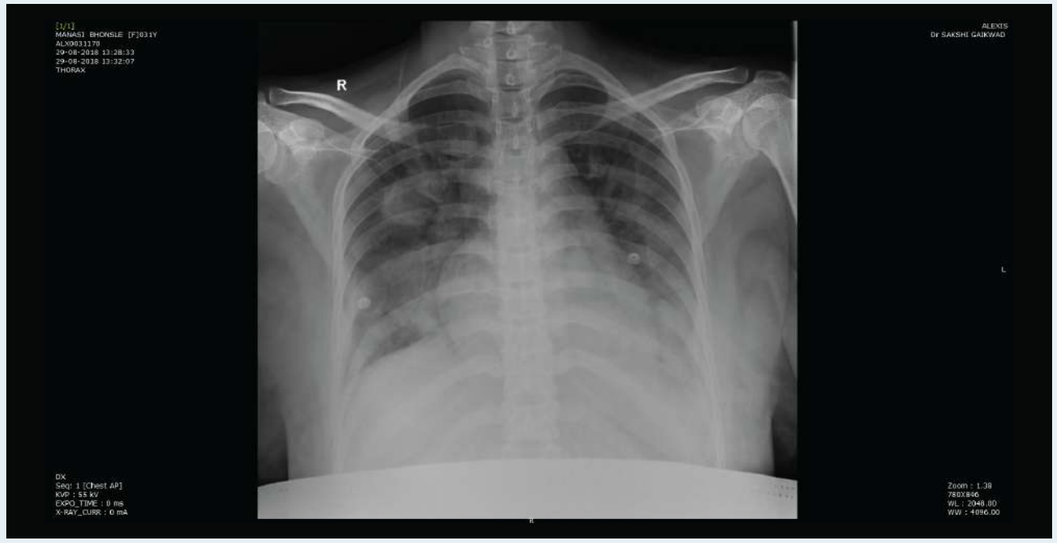
Setlic RF, Morgan J Mc Allister K et al.Pulmonary Haemorrhage fever, Southern Medical Journal 2004;97(7):688-9

Mujumdar R, Jana Ch.Ghosh s, Biswas U,clinical spectrum Of Dengue fever in a tertiary care centre with particular reference to atypical presentation in 2012,outbreak in kolkatta. J.Indian Med Ass 2012;110:904-6

Janz D, Ware LB. An approach to the patient with the Acute Respiratory Distress Syndrome. Clin Chest Med 2014,35(4);685-696

Francis CYL, Lung Ultrasound –a primary survey of the acutely dyspneic patient. J of Intensive Care 2016,457

Markhand Tunna R.T. The Efficacy of bedside chest ultrasound; from accuracy to outcome. Eur Respire Rew 2016,25;230-246.



14 YEARS POST FEMORAL OSTEOTOMY FOR FRACTURE NECK FEMUR - TREATED SUCCESSFULLY WITH A MODULAR TOTAL HIP REPLACEMENT

A CASE REPORT BY:
DR. ALANKAR RAMTEKE, DR. ANIRUDDHA SONEGAONKAR, DR. SHIVRAJ SURYAWANSHI

ABSTRACT

Fracture of neck of femur is very common with established guidelines for its management [1]. In younger and active patients hip joint preservation by means of various bio-mechanical treatment modalities like osteosynthesis, fixation and realigning osteotomy is indicated [2, 3]. In older patients, total hip replacement (THR) is preferred because of predictable outcomes, excellent success rates and early return to quality of life [2, 3]. However with any proximal femoral osteotomy the anatomy of proximal femur is distorted and THR becomes difficult as standard implants and regular surgical techniques cannot be used [4]. In the present patient proximal femoral osteotomy, bone loss, limb shortening and disuse osteoporosis posed significant challenges in surgical management.

INTRODUCTION

Fracture neck of femur is prone to complications like non-union and osteonecrosis of femoral head - sometimes needing multiple surgeries finally leading to inevitable THR if attempts at fracture union fail [2, 3]. Various types of osteotomies indicated for treatment of non-union of fracture neck femur invariably distort the proximal femoral anatomy making a future hip replacement difficult and occasionally impossible[5]. In present patient proximal femoral osteotomy, bone loss, limb shortening and disuse osteoporosis ruled out use of standard THR designs [4].

CASE REPORT

A 64 year old wheel chair bound lady presented at Alexis with complaints of back ache, 'flail' left lower limb and inability to walk since 14 years with a history of hip surgery. Patient had a history of a fall at her home in the year 2005, for which she underwent a surgery of which no medical records were available. After surgery, patient was treated with hip spica and advised bed rest for 1 year. After two years, she gradually started ambulating with a walker but with difficulty. At presentation - patient was housebound for the last 14 years and was dependent on family for activities of daily living.

On examination patient had a scar of lateral approach to the hip. Attempted range of motion was painful but not restricted. Patient was unable to perform a straight leg raising test with no active control over her 'flail' left lower limb with no distal neurovascular deficit. She had 4 cm shortening with a fixed pelvic obliquity and fixed lumbar scoliosis with convexity to left side. On radiographic examination - pelvis with both hips anteroposterior view radiograph revealed some osteotomy done at level of lesser trochanter with greater trochanter (GT) in a valgus position with no calcar and proximal femoral bone loss. Femoral head was necrotic with severe disuse osteoporosis, thin femoral cortices and osteoporotic acetabulum (Figure 1A, 1B). On lateral radiograph - GT had united in malposition (Figure 1C, 1D).

We decided to offer a THR to this patient with following challenges - avascular necrosis of femoral head, deformed proximal femur, disuse osteoporosis, limb shortening of 4 cms, potential damage to sciatic nerve while achieving leg length equality and negotiating or incorporating previous scar of surgery, poor soft tissue and hip capsule due to previous surgery leading to potentially high chances of postoperative hip dislocation.

Avascular necrotic femoral head (Figure 1A, 1B) meant that any osteotomy or fixation was not feasible and THR was indicated. Proximal femoral bone loss and deformity did not allow use of any standard cemented or uncemented stem as there was deficient proximal femoral metaphyseal bone to support the cement mantle or metaphyseal loading of uncemented femoral stem. As the femoral canal was not in a straight line in coronal and sagittal planes it was not feasible to put any standard or long diaphyseal fitting femoral stem as it would easily perforate the osteoporotic femoral cortices (Figure 1A, 1C). Also the osteoporotic and wide femoral canal ruled out use of a diaphyseal fitting stem. A standard femoral implant has a fixed anteversion which cannot be adjusted to accommodate the deformed proximal femur (Figure 2A, 2B). With a shortening of 4 cms - restoring leg length is not easy as 14 years old scar and fibrosis will not allow leg length restoration. Too much lengthening can cause sciatic nerve palsy and foot drop.

PLANNING

We planned to use an S-ROM modular cementless THR system (DePuy Orthopaedics Inc., Warsaw, Ind) for this patient. It consists of 2 modular parts (Figure 3) which are assembled on the table to suit the patient's femoral anatomy [6]. There is a proximal sleeve that consists of a conical portion and a spout that can be oriented and placed in best available bone [6]. The fully porous coated sleeve is designed to convert shear forces to compressive forces at implant-bone interface (Figure 4). Then there is a titanium stem, polished in its distal portion, with distal splines 1.25-mm in height to provide purchase and rotational stability in the diaphyseal bone. The splines surround a coronal slot, which reduces the stem stiffness. The stem is then placed through the sleeve to engage the taper proximally via a cold weld, with the splines penetrating distally into the endosteal cortical bone for rotational stability [6].

INTRAOPERATIVE FINDINGS AND CHALLENGES

Standard posterior approach was used for exposure. The osteotomy was extra-articular, at the level of lesser trochanter, well united and posterior capsule and short rotators were atrophic but intact (Figure 5). Bone was very osteoporotic, proximal femur was deformed with deficient calcar. Acetabular fixation was relatively easy and a Pinnacle cup (DePuy Orthopaedics Inc., Warsaw, Ind) was fixed in 45 degree inclination and 20 degree anteversion. We projected an imaginary line from the distal femoral shaft to proximal femur to use as our entry point for intramedullary canal reaming. As proximal femur and GT were deformed, the anatomical landmarks like pyriform fossa were not reliable references. We carefully avoided any damage to the GT as it was the only bone available and fixed the proximal sleeve in the GT as shown (picture 6 & 7). The GT was extremely osteoporotic and we carefully avoided use of excessive force. We used bone graft from femoral head to augment fixation of the proximal sleeve in weak and osteoporotic GT bone. Thereafter the distal stem was carefully inserted avoiding perforation of osteoporotic and deformed proximal femoral canal due to the coronal split at the same time using the splines to get sufficient rotational stability. The modularity of the implant was used to ensure femoral stem anteversion of 20 degree. Highly cross linked ALTRX polyethylene liner and 32 Ceramic head (DePuy Orthopaedics Inc., Warsaw, Ind) were used as bearing couple. Head was reduced with due care to avoid fractures during reduction. Reduction was very stable in flexion of 100 degree, 30 degree adduction and 45 degree anteversion on table. Combined anteversion angle on coplanar test was 40 degree [7]. Posterior capsule and short rotators were repaired using transosseous sutures. We avoided lengthening more than 3 cm to avoid sciatic nerve stretch.

POST-OPERATIVE COURSE AND FOLLOW UP

Patient postoperative stay in the hospital was uneventful. We were confident of the fixation and allowed full weight bearing walking from day 1 post surgery. Patient was discharged 4 day post-operative ambulatory with a walker. Satisfactory signs of osteointegration were seen radiographically at 6 weeks. Patient last followed up at 6 months post-surgery and is walking independently with satisfactory clinical and radiographic progress. We have advised her to use a tripod stick to avoid falls owing to her severe disuse osteoporosis.

CONCLUSION

We will continue to see patients of hip osteoarthritis secondary to various causes treated in past with osteotomy. Even though these patients pose unique and multiple challenges, they can be treated successfully with satisfactory outcomes as suitable implants, advanced surgical skills and technical know-how are now available.

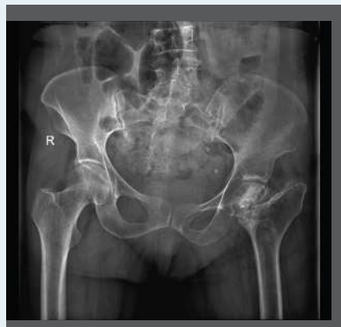


Figure 1 A - Preopaerative radiograph, PBH anteroprposterior

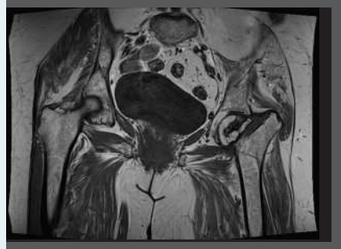


Figure 1 B - MRI showing non-union, neck resoprtion & necrotic, sclerotic femoral head

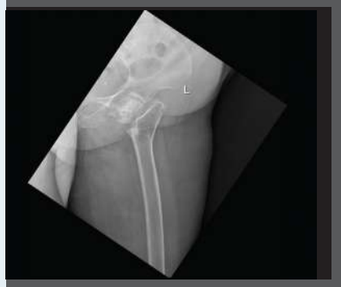


Figure 1C - Preoperative shoot through lateral view left hip and proximal femur

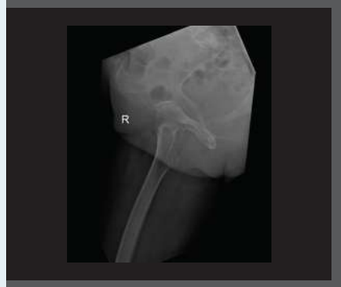


Figure 1 D - Preoperative shoot through lateral view right hip and proximal femur, for comparison

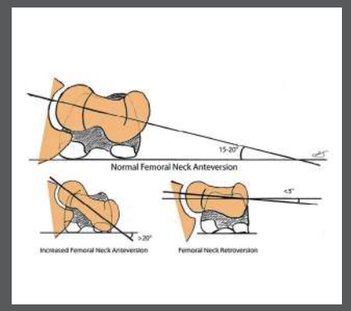


Figure 2 A - Illustration to explain femoral neck anteversion in a native hip

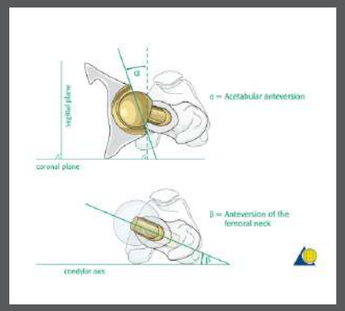


Figure 2 B - Illustration to explain combined anteversion in a THR

Figure 3 - Various parts of a modular THR system

Figure 4 - Clinical photograph of a porous coated proximal sleeve used in this surgery

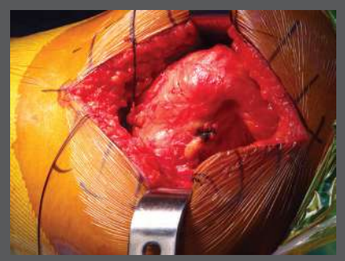


Figure 5 - Intraoperative photograph of distorted proximal femur and atrophic short external rotator muscles

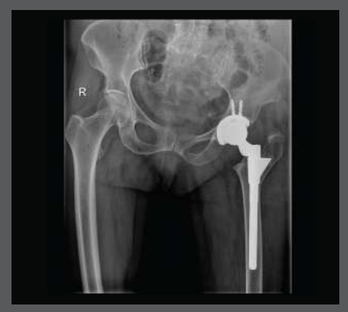


Figure 6 - Postoperative 6 weeks PBH AP radiograph showing - position of proximal sleeve in best available bone, signs of osteointregration, leg length restoration, well aligned and well fixed acetabular and femoral component

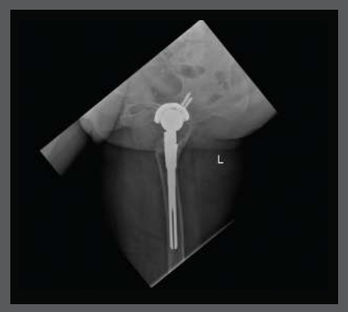


Figure 7 - Post operative 6 weeks shoot through lateral left hip radiograph showing well aligned and well fixed acetabular and femoral component.

REFERENCES

1. Rath, S., L. Yadav, A. Tewari, T. Chantler, M. Woodward, P. Kotwal, A. Jain, A. Dey, B. Garg, R. Malhotra, A. Goel, K. Farooque, V. Sharma, P. Webster, and R. Norton, Management of older adults with hip fractures in India: a mixed methods study of current practice, barriers and facilitators, with recommendations to improve care pathways. Arch Osteoporos, 2017. 12(1): p. 55.
2. Ly, T.V. and M.F. Swiontkowski, Management of femoral neck fractures in young adults. Indian J Orthop, 2008. 42(1): p. 3-12.
3. Forsh, D.A. and T.A. Ferguson, Contemporary management of femoral neck fractures: the young and the old. Curr Rev Musculoskelet Med, 2012. 5(3): p. 214-21.
4. Boos, N., R. Krushell, R. Ganz, and M.E. Muller, Total hip arthroplasty after previous proximal femoral osteotomy. J Bone Joint Surg Br, 1997. 79(2): p. 247-53.
5. DeCoster, T.A., S. Incavo, J.W. Frymoyer, and J. Howe, Hip arthroplasty after biplanar femoral osteotomy. J Arthroplasty, 1989. 4(1): p. 79-86.
6. Spitzer, A.I., The S-ROM cementless femoral stem: history and literature review. Orthopedics, 2005. 28(9 Suppl): p. s1117-24.
7. Blumenfeld, T.J., Pearls: Clinical Application of Ranawat's Sign. Clin Orthop Relat Res, 2017. 475(7): p. 1789-1790.

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ALEXIS
PARTNERS IN GOOD HEALTH

SPONTANEOUS RUPTURE OF URINARY BLADDER - RARE CASE WITH DIAGNOSTIC DILEMMA

A CASE REPORT BY:
DR. JUNED SHAIKH, DR. NISHANT DESHPANDE, DR. ABHIJIT DESHMUKH

ABSTRACT

Spontaneous rupture of the urinary bladder (SRUB) is a rare clinical condition. We report this young male who presented with SRUB which occurred after binge alcohol drinking. A Foley catheter was placed. Patient managed conservatively initially. Later during stay, CT cysto-urethrogram was done and it was confirmed intraperitoneal bladder rupture at dome. The patient underwent laparoscopic repair of bladder rupture repair. Initial presentation remains imprecise; continuous urinary leakage leads to abscesses, sepsis, and metabolic derangements. CT scan is imaging modality of choice. The condition is a surgical emergency, and necessitates urgent closure of the breach.

INTRODUCTION

Spontaneous rupture of the urinary bladder (SRUB) is a rare clinical condition, with very high morbidity and mortality rates. Idiopathic rupture is encountered in less than 1% of all cases. This condition is often difficult to diagnose clinically, even with increased timely access to computed tomography (CT). The approach requires a high index of suspicion as the patients present with non-specific abdominal pain and may not provide a clear history [1].

CASE REPORT

A 33-year-old chronic alcoholic male presented in ER with history of anuria since last 12 hours with significant abdominal distension. He was known case of right renal agenesis and seizure disorder on levetiracetam. He had history of recurrent UTI since past one year and was diagnosed as overactive bladder by urologist outside; on intermittent medication for same. He also had diffuse abdominal pain, with no alleviating or aggravating factors. Patient stated that he had consumed moderate amount of alcohol at night prior to his arrival. His vitals were stable. Cardiopulmonary examination did not show any abnormalities. Abdominal examination revealed sluggish bowel sounds, with no visible scars; abdomen was soft, diffusely tender to palpation, with no rebound tenderness or organomegaly. Rest of the physical examination was within normal limits. Laboratory findings revealed BUN of 56 mg/dL and creatinine of 3.3 mg/dL, both of which were significantly elevated from patients' baseline. The complete blood count showed WBCs of 14.7 × 103/μL with normal haemoglobin/haematocrit and platelets. LFT and viral markers were within normal limit in the emergency department, a Foley catheter was placed draining sanguineous urine. His ultrasound abdomen showed liver parenchymal disease with moderate ascites and changes of cystitis. Patient was put on conservative management with IV fluids and antibiotics. Opinion of gastroenterologist was taken and patient underwent diagnostic and therapeutic ascitic tapping; 900 ml fluid drained. Ascitic fluid examination revealed TLC of 2000, albumin of 0.4 gm/dl and LDH of 71U/litre. Urologist opinion was sought for underlying bladder problem and started with tablet alfuzosin. Patient responded well to above treatment and renal parameters became normal within 3 days. Foleys catheter was removed after 3 days. Post catheter removal patient complained of lower abdominal pain and distension with burning urination. Urgent CT cysto-urethrogram was done and it confirmed small intraperitoneal bladder rupture at dome with extravasation of urine.

The patient underwent diagnostic cystoscopy followed by laparoscopic bladder repair. Cystoscopy showed high and narrow obstructive bladder neck. Bladder was mildly trabeculated with slit shaped tear at dome with severe inflammation. Biopsy was taken from the site. Laparoscopic double layer closure of rupture was done using a continuous technique. An indwelling catheter was left to ease healing of the bladder repair. The patient had an uneventful post-operative stay. He was discharged home with a Foleys in situ and removed after 14 days. Patient was started with tablet silodosin post-operatively and was voiding well on follow up.

DISCUSSION

Pressure of more than 300cm H2O is required to rupture a normal bladder. Retention of urine alone rarely predisposes the patient to SRUB. Spontaneous bladder perforation (SBP) is an extremely rare event with almost all of the cases reported having a history of previous bladder manipulation, lower urinary tract obstruction, neurogenic bladder, pelvic radiotherapy or surgery, inflammation, and malignancy. [2] The clinical presentation is usually with features of peritonitis and most of the time, the diagnosis is made intraoperative. The condition carries high rates of morbidities and mortalities (47%) [3], attributed to the delay in diagnosis. It is often difficult to distinguish SRUB from other causes of acute abdomen especially in the absence of a history of traumatic event [4]. The spontaneous rupture of the bladder can be facilitated by excessive alcohol consumption which has a diuretic effect [5] and leads to over distension in the context of the sensitivity disturbances due to the α-sympathomimetic effect and CNS depression produced by the alcohol. In men, congestion of the prostate and prostatic urethra aggravates the obstruction of the outlet [6]. With extreme increase of the intra-vesical pressure, the most susceptible part is the intra-peritoneal bladder [7]. These patients may present with abdominal distension and oliguric acute renal failure secondary to reabsorption of urinary constituents from the peritoneum [8] and many of these cases were initially treated conservatively with urinary catheterization and antibiotics. Moreover, in patients with SBR, the diagnosis is often delayed for various reasons. First, absence of a history of trauma may prevent the physician from considering SBR. Second, because of diminished sensations and altered motor function in the abdomen and bladder, there is an absence of abdominal pain and signs which may significantly delay presentation as well as diagnosis [9]. Third, the perforation may be small and the collection often contained or loculated due to adhesions of previous surgery, radiotherapy, or advanced malignancy. Complications caused by the leakage of the urine into the peritoneal cavity and pelvis include intra-abdominal and pelvic abscesses, sepsis, and metabolic derangements. Significant peritoneal reabsorption of urea and creatinine can masquerade as acute renal failure on initial biochemical testing, as observed in our patient. Electrolyte abnormalities such as hyperkalemia, hypernatremia, uremia, or acidosis may also occur. Electrocardiogram abnormalities resulting from hyperkalemia may be seen. Several authors have noted increased mortality rates after severe symptom onset [10, 11]. Imaging modalities to evaluate a patient with suspected bladder injury have included retrograde cystography, CT retrograde cystography, and delayed CT cystography. Conventional abdominal non-contrast CT has an accuracy rate of only 60.6% for bladder injury compared to 95.9% for the retrograde cystogram [12]. However, in a patient with worsening abdominal pain/distension and vital sign abnormalities, a CT scan will likely be the imaging modality of choice.

Extra-peritoneal bladder rupture can be managed conservatively [8].

Intraperitoneal SRUB is a surgical emergency and can be rapidly fatal if diagnosis and treatment are delayed, with early and adequate treatment being essential [7, 13]. The treatment consists of laparoscopic or open suturing of bladder rupture as continuous leakage of urine does not allow spontaneous closure of the breach [7]. Identifying risk factors for SBR is important because the recurrent nature of perforation has been reported by many authors [9, 14].

Because the weakening factor may not be modifiable, it is pertinent to meticulously manage the compounding factors to prevent occurrence of SBR. In neurogenic/non-neurogenic voiding dysfunction, cystometrography provides important guidelines for evidence based management and has shown to decrease the incidence of perforation [15].

CONCLUSION

Most cases of spontaneous bladder rupture in relation to excessive alcohol consumption can have a positive outcome with early prediction and management. This diagnosis should be considered in those who present with acute abdomen, anuria and a raised serum creatinine level. Although the idiopathic SRUB is a rarely encountered, all medical professionals working in ER settings need to be aware of this presentation. Prompt surgical consultation and management would prevent a poor clinical outcome in such cases.

REFERENCES

1. Daignault MC, Saul T, Lewiss RE. Bedside ultrasound diagnosis of atraumatic bladder rupture in an alcoholintoxicated patient: a case report. Crit Ultrasound J. 2012;4(1):9.
2. Huffman JL. Atraumatic perforation of bladder. Necessary differential in evaluation of acute condition of abdomen. Urology. 1983;22:30e35.
3. Hansen HJ, Eldrup J. Spontaneous rupture of the urinary bladder - a late complication to radiotherapy. Case report. Scand J Urol Nephrol. 1989;23(4):309-310.
4. Muneer M, Abdelrahman H, El-Menyar A, Zarour A, Awad A, Al-Thani H. Spontaneous Atraumatic Urinary Bladder Rupture Secondary to Alcohol Intoxication: A Case Report and Review of Literature. Am J Case Rep. 2015;16:778-781.
5. Bastable JR, De Jode LR, Warren RP. Spontaneous rupture of the bladder. Br J Urol. 1959;31(1):78-86.
6. Kim HJ, Moon H, Sohng I, Lee HW, Lee G, Lee JG. Effects of ethanol and its metabolite acetaldehyde on responses of the rat bladder. BJU Int. 1999;83(6):686-692
7. Jurisic D, Glavan E, Cugura JF, Vidovic D. Splenic rupture following idiopathic rupture of the urinary bladder presenting as acute abdomen. J Postgrad Med. 2007;53(1):39-40.
8. McAnnich JW, Santucci RA: Genitourinary trauma. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ (eds), Campbell's Urology, 8th ed, vol 4, Philadelphia: Pennsylvannia, Saunders, 2002, pp: 3707-3744.
9. Garfinkle SE, Chiu GW, Cohen SE, Wolfman EF. Spontaneous perforation of the neurogenic urinary bladder. West J Med 1976; 124(1): 64-66.

10. Sezhian N, Rimal D, Suresh G. Isolated intraperitoneal bladder rupture following minor trauma after alcohol ingestion. South Med J. 2005;98(5):573-574.
11. Herd AM, Crofts NG, Lee LM, Girard JV, Sweetland RJ. Isolated bladder rupture after minor trauma in a patient with alcohol intoxication. J Emerg Med. 1994;12(3):409- 411.
12. Hsieh CH, Chen RJ, Fang JF, Lin BC, Hsu YP, Kao JL, Kao YC, et al. Diagnosis and management of bladder injury by trauma surgeons. Am J Surg. 2002;184(2):143- 147.
13. Arnaout F. Spontaneous atraumatic rupture of urinary bladder. Internet Journal of Urology. 2008;5(2).
14. Kato A, Yoshida KI, Tsuru N, et al. Spontaneous rupture of the urinary bladder presenting as oliguric acute renal failure. Int Med 2006 (online publication DOI:10.2169/internalmedicine. 45.1748).
15. DeFoor, W., Tackett, L., Minevich, E., Wacksman, J. and Sheldon, C. Risk factors for spontaneous bladder perforation after augmentation cystoplasty. Urology 2003; 62: 737-741

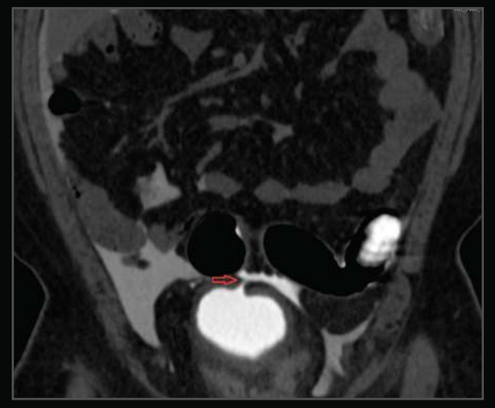

Figure 1: CT CYSTOGRAPHY

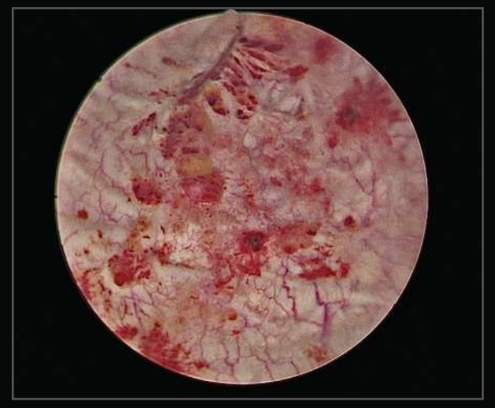

FIGURE 2: SLIT SHAPED TEAR AT DOME WITH SEVERE INFLAMMATION

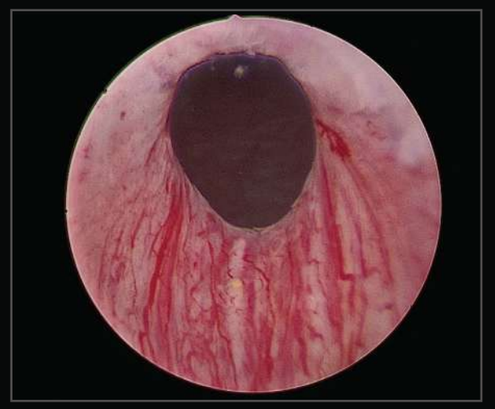

Figure 3: CYSTOSCOPY-HIGH AND NARROW OBSTRUCTING BLADDER & NECK

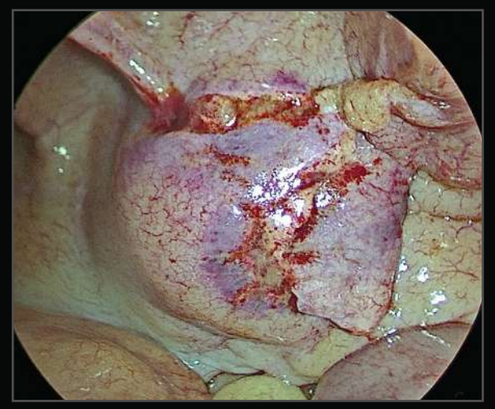

Figure 4: LAPAROSCOPIC REPAIR-SEROSAL TEAR AT DOME OF BLADDER WITH SURROUNDING INFLAMMATION

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