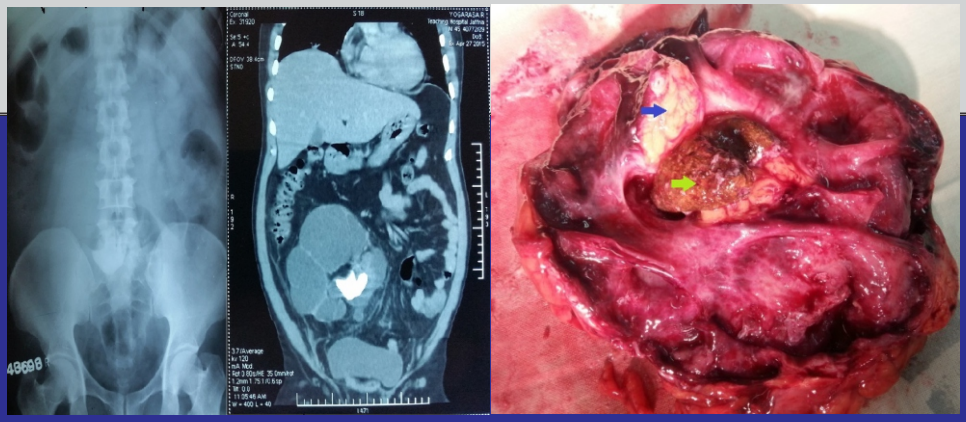
THE SRI LANKA JOURNAL OF SURGERY

October 2018 Volume 36, No.3 ISSN 1391-491X



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- Mucinous adenocarcinoma of renal pelvis in a crossed ectopic kidney
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- Pseudopapillary tumour of pancreas in a 13 year old girl

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The Sri Lanka Journal of Surgery

Journal of
The College of Surgeons
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Augustl 2018 Volume 36, No.3 - Quarterly. ISSN 1391-491X e - journal ISSN 2279 2201

Mission: "To reach the highest standard of scientific surgical practice by dissemination of high quality scientific information and to foster and promote the growth of scientific surgery in Sri Lanka and in the region"

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Printed by
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A centre dedicated for men's health and wellbeing for the first time in Sri Lanka - End your suffering with an effective treatment for Erectile Dysfunction

Lanka Hospitals PLC, a premier health care provider in Sri Lanka, announces its latest addition to the Centres of Excellence- the Male Wellness Centre (MWC) – in a bid to offer services to improve health and wellbeing of men. It's also significant that a fully-fledged wellness centre dedicated solely for men has been established for the first time in Sri Lanka.

The MWC caters to a host of services including Personnel fitness scheduling and programming, Sport health and injury management, Dietary & Nutritional advices, Pre-marital counseling and health screening, Management of premature ejaculation, Management of Erectile dysfunction, Cosmetic surgeries (Bariatric / Ocular / Dental). In addition to the General health screening, patients can obtain screening for Liver, Kidney, Respiratory, Cardiac, Diabetic, Endocrine-Hormonal, Cancer and Sexually Transmitted Diseases in addition to Substances and Alcohol abuses. Furthermore, apart from leading physicians MWC offers the service of competent consultant specialists such as Cardiologist, Endocrinologist, Diabetologist, Venerologist, Urologist, Nephrologist, Oncologist, Surgeon, Vascular Surgeon, Psychiatrist as well as Counsellor.

Erectile Dysfunction (Impotence) is a common health issue suffered by men, defined by the difficulty in achieving and maintaining a penile erection during sexual intercourse. In the Sri Lankan context, the issue is hardly brought into light especially by those who suffer and often show reluctance to seeking proper medical attention. Often, incorrect and misleading advice not only aggravates the issue, but also lead them to face unwanted complications. A special Shock Wave Therapy unit was established within the Male Wellness Centre by the Lanka Hospitals to specifically treat impotence.

The Centre conducts in-depth studies and comprehensive medical analysis to precisely identify the causes for impotence such as Vascular, Psychogenic, Neurological, Hormonal, Structural and others. Being a newer and less invasive way to treat this common sexual challenge shock wave therapy has proven to be effective even when oral medication has failed. Also known as penile extracorporeal low-intensity shockwave therapy, this method involves the use of low intensity acoustic pulse waves that lead to release of factors which promote growth of new blood vessels in the penis Therapy compromises of a handheld device being angled towards the shaft of the penis. One of the main advantages of this treatment method is that it has no clinically relevant side effects. Each treatment session can last approximately 20 minutes.

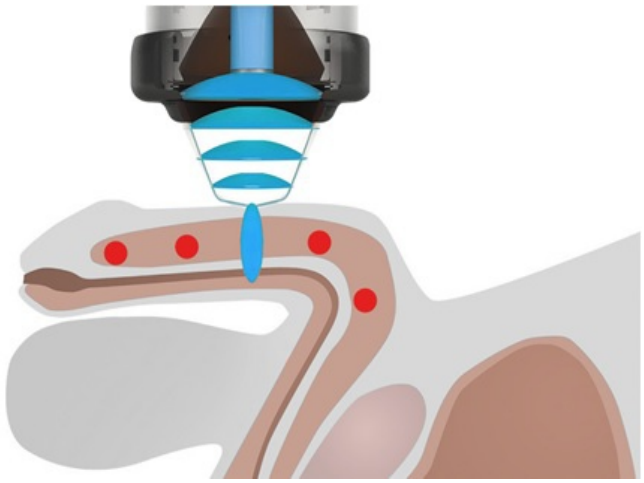

Figure 1. Shock wave therapy

Shock wave treatment is a completely painless way to treat what can be a life altering condition and a regular course of treatment usually comprises of six sessions. The frequency of these session can be tailor made as below and would be decided by the consultant:

1) Every day for 6 days
2) Every second day over an 11 day period
3) Twice a week for 3 weeks

The outcomes include gaining of more frequent erections, more rigid erections, ability to maintain an erection and perform entire act of sexual intercourse and freedom to reduce or omit medication. Therefore the use of a treatment which researchers claim is “really a breakthrough” could be good news for men who have erectile dysfunction.

As a hospital staying abreast with latest medical technology, Lanka Hospitals established Male Wellness Centre in a bid to provide world class health care services to Sri Lankan as well as International patients. Moreover, when catering to health issues and conditions that are highly sensitive and personal, Lanka Hospitals delivers complete confidentiality to its patients with the assistance of its specially trained staff.

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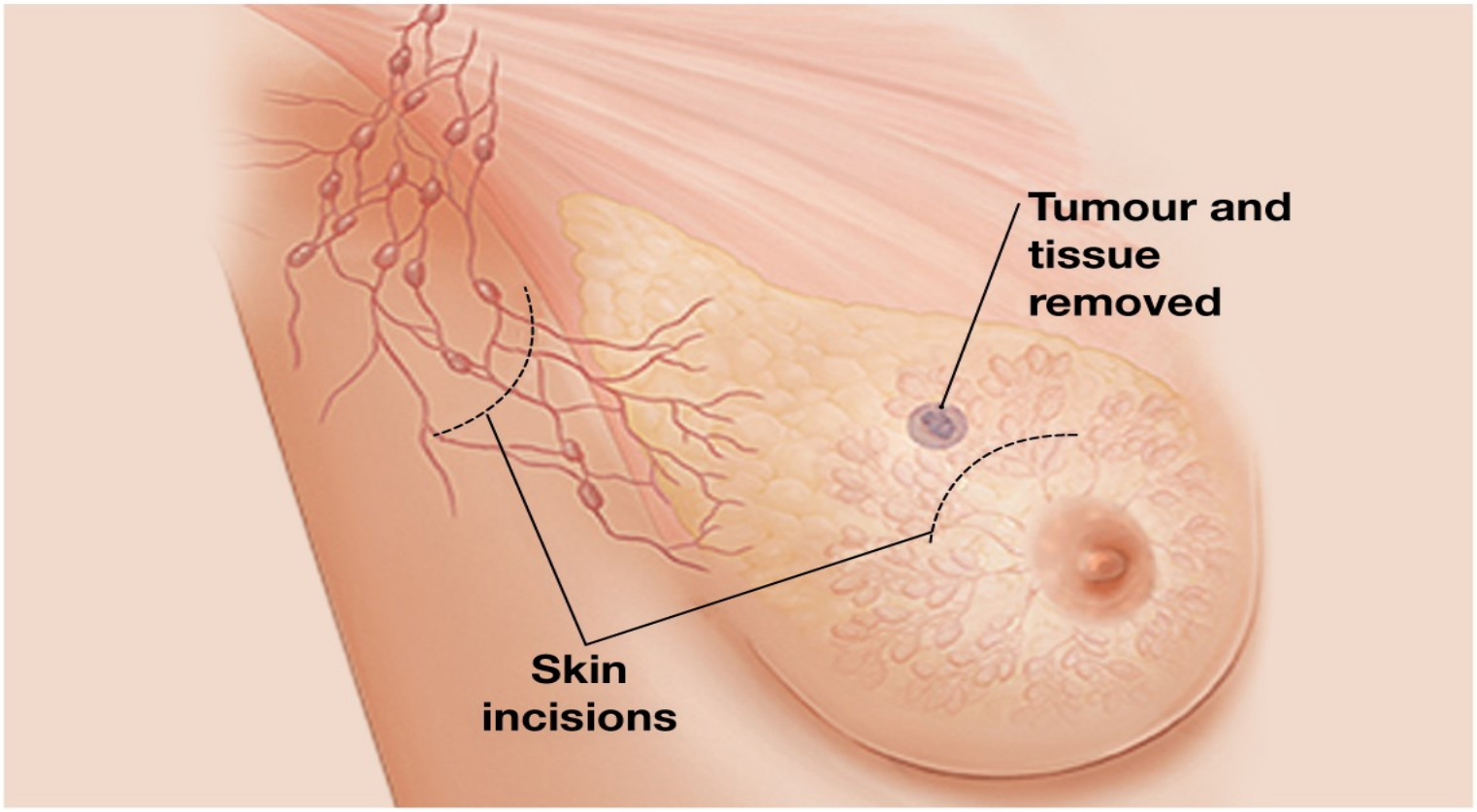

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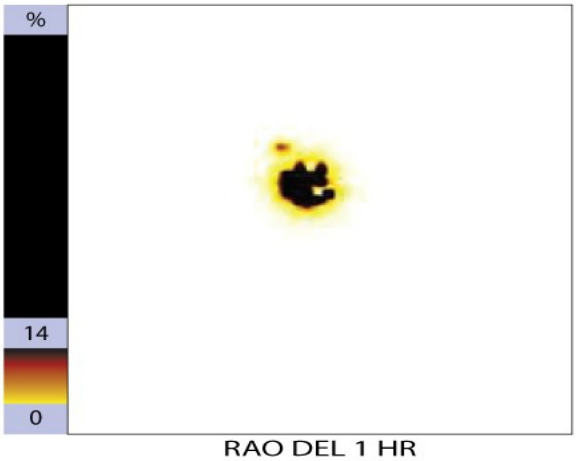

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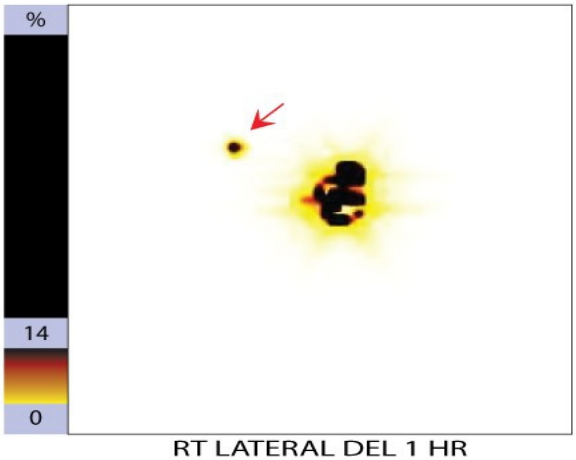

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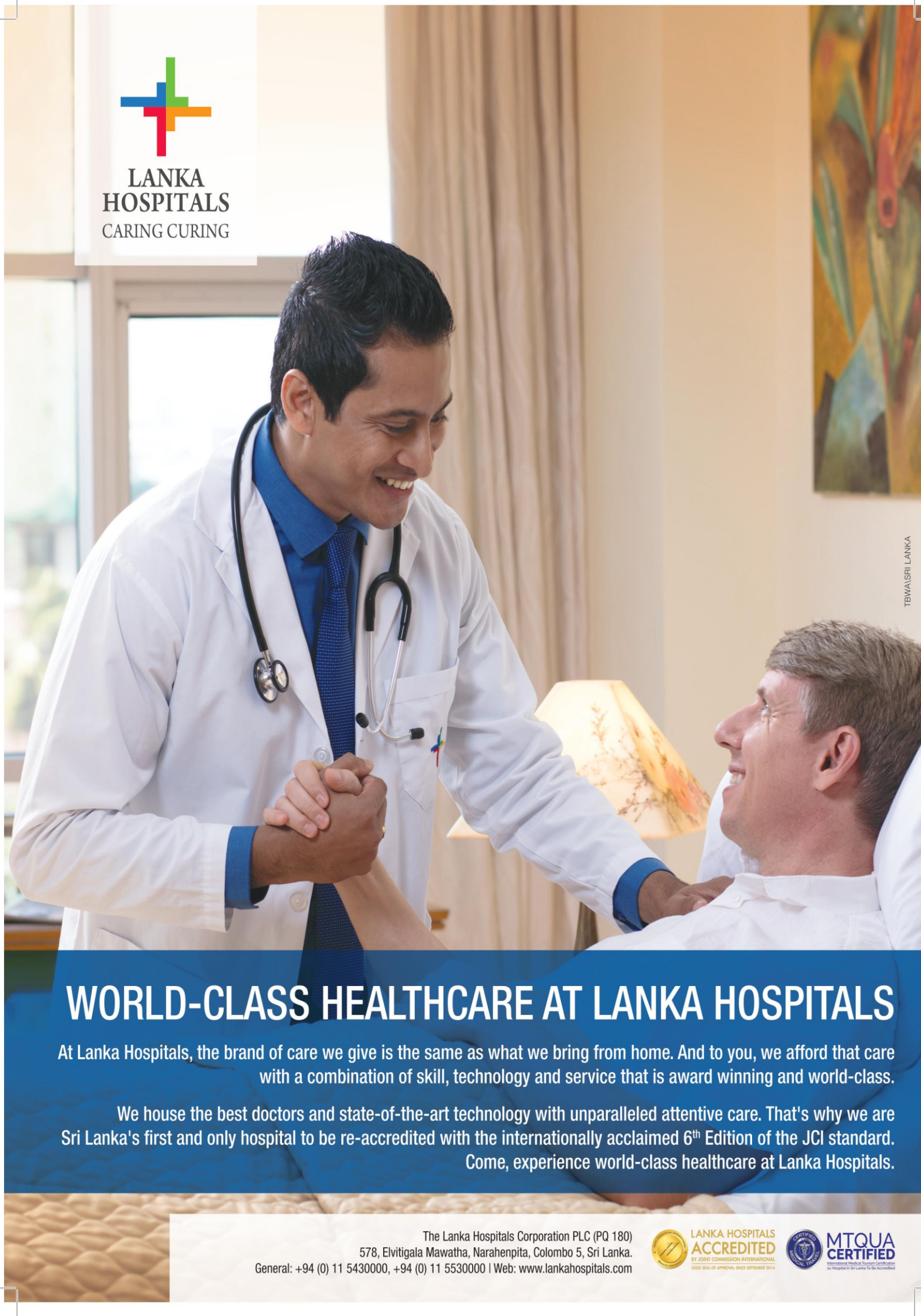
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The Sri Lanka Journal of Surgery

October 2018 Volume 36, No. 3

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CASE STUDY

Torsion of a wandering spleen

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Key words: Wandering spleen; splenectomy; splenopexy

Introduction

The spleen is a small organ usually located in the left upper abdomen. The spleen combines innate and adaptive immune system in a unique way. The function of the spleen is to remove older erythrocytes, cellular debris and blood-borne microorganisms from the circulation [1].

It is usually present in the left upper abdomen but sometimes due to lack of supporting ligaments it may be displaced from its original position. This is known as a wandering spleen. A wandering spleen is an uncommon clinical occurrence with less than 500 cases reported[2]. A wandering spleen may be either congenital or acquired. There is no hereditary predilection. Most of the cases are acquired. Acquired wandering spleen usually occurs during adulthood and it is due to trauma or other underlying conditions (e.g. connective tissue disease or pregnancy) that cause weakening of the various ligaments that hold the spleen in its normal position [3]. Wandering spleen is more common in females of age 20-40 years. Diagnosis is often elusive and depends on abdominal imaging.

The treatment of a wandering spleen is usually splenopexy. But if torsion of the pedicle is present with infarction of the spleen, splenectomy is the treatment of choice. Here we report a case of torsion in a wandering spleen.

Case Report

A 30 year old female presented to the surgical emergency department with a lump in her lower abdomen for two months and pain in her lower abdomen for four days. The lump appeared two months ago with pain tenderness in abdomen for which she was treated at her local hospital. Four days ago, she had developed acute pain in lower abdomen for which she was referred to the tertiary centre. She had no significant past history other than sterilization (tubal ligation) 1½ years ago. On examination, the lower abdomen was tense and tender with guarding. Bowel sounds were present. Vitals were stable except tachycardia 120 beats/minute. A provisional diagnosis of peritonitis was made. Initially she was managed conservatively. Routine blood investigations were within normal limits except her haemoglobin which was 6 gm%. Ultrasonography showed a vague mass in the lower abdomen with an absent spleen. Contrast Enhanced Computerized Tomography scan showed a well-defined cystic lesion of approximately 128×84×169 mm with non-enhancing material in the umbilical and the hypo-gastric region. The vascular pedicle was attached to its mid part and it had twisted around the pedicle. The lesion was abutting on the anterior abdominal wall anteriorly and dome of urinary bladder and fundus of uterus inferiorly. The lesion was displacing adjacent bowel loops. The spleen was not visualized. Hence the provisional diagnosis of infarcted wandering spleen was made and patient underwent a laparotomy. On entering the peritoneal cavity, the spleen was visualized in lower abdomen. There were adhesions of bowel, bladder and the omentum to spleen which were divided. Pedicle of the spleen was very thin and found twisted. The spleen was delivered out of the peritoneal cavity and the pedicle was ligated and divided. After exploring rest of peritoneal cavity, a single drain was put and wound was closed in layers. Post operatively patient did well and was discharged after one week.

Discussion

The spleen is usually located in the left upper abdomen at the level of 9th to the 11th intercostal spaces. It is a crescent shaped structure, it has a convex outer margin, and inner

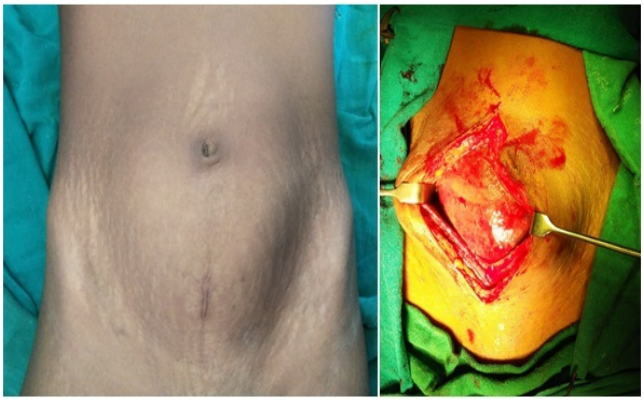

Figure 1. Spleen in lower Abdomen

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DOI: http://doi.org/10.4038/sljs.v36i3.8524

margin is indented [4].

Normally the spleen is fixed to the stomach and posterior abdominal wall by gastrosplenic and splenorenal ligaments. The phrenocolic ligament also helps in fixation of the spleen with the diaphragm and the upper abdomen. If any or all of the gastrosplenic, splenorenal and phrenocolic ligaments are poorly developed or if they are absent or loose, the spleen migrates from its normal position to the left lower quadrant or other regions of abdomen; and gravity leads to the migration of spleen along with its vascular pedicle to the lower abdomen [5].

In a wandering spleen, the spleen is only attached by its vascular pedicle Instead of ligaments. If this pedicle is twisted due to the movement of the spleen, the blood supply to the spleen may be interrupted and it may undergo infarction. As there is very little or nothing to hold the spleen in place, the spleen moves or "wanders" in the lower abdomen or pelvis due to gravity, and it may be mistaken for an unidentified abdominal mass. "Acquired" wandering spleen usually occurs during adulthood and it is due to trauma or other underlying conditions (e.g., connective tissue disease or pregnancy) that cause weakening of various ligaments that hold the spleen in its normal position [3].

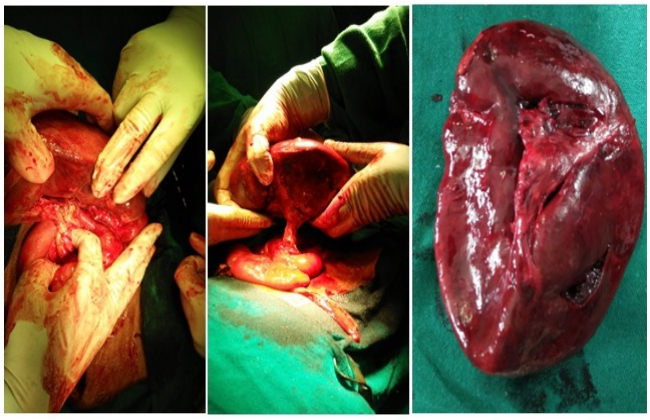

Figure 2. Torsion in Pedicle which was untwisted, ligated and splenectomy done

A wandering spleen is very rare occurrence, and is seen in women between the age of 20 to 30 years and young children. Clinically, diagnosis is very difficult.

The wandering spleen may be found in any part of the abdomen or pelvis depending on length of its vascular pedicle. Abnormally fixed spleen may be twisted on its vascular pedicle, causing ischemia to the spleen which may progress to infarction if not treated in time. Clinical presentation of a wandering spleen is varied. The clinical presentation may be acute or chronic; it may be present as an asymptomatic mass, a mass with pain or an acute abdomen.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Most of the cases of wandering spleen are acquired, and are more common in young females.
- A wandering spleen may occur in young adult due to trauma or other underlying conditions (e.g., connective tissue disease or pregnancy) that cause weakening of the ligaments that hold the spleen in its normal position.
- Treatment of a wandering spleen is usually splenopexy. But, if torsion of pedicle is present with infarction of spleen, a splenectomy is the treatment of choice.

CASE STUDY

Congenital lobar emphysema: a rare cause for respiratory distress in newborns and infants

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Keywords: Congenital; lobar emphysema; respiratory; distress; newborn

Introduction

Congenital lobar emphysema is a rare disorder commonly presents with neonatal or infantile respiratory distress. The rarity and the associated cardiac abnormalities have made it a diagnostic challenge in paediatric practice. High degree of clinical suspicion aided with contrast enhanced computerized tomography of the chest (CECT) is the key to diagnosis. Prompt diagnosis with timely thoracic surgical intervention is essential to save affected lives.

Case 1

A four months old baby with ventricular septal defect (VSD) presented to a paediatric cardiac unit due to worsening respiratory symptoms. Two dimensional echocardio-graphy demonstrated a large muscular VSD with a significant left to right shunt and moderate pulmonary hypertension. Early surgical intervention was planned and poly tetrafluroethane patch repair of VSD performed with uneventful intra-operative and immediate post-operative period.

Progressive respiratory distress with diminished breath sounds on right chest was noted on 7th post operative day, however there wasn't any clinical and biochemical features of lower respiratory tract infection. Chest X- ray revealed diminished lung markings at the centre of the right hemi thorax with collapsed upper and lower lobes. Clinical suspicion of pneumothorax was made and re-evaluated by the thoracic team where the CECT thorax demonstrated the lobar emphysema of right middle lobe.

After fibre optic bronchoscopy (F.O.B) the baby underwent axillary thoracotomy under single lumen endotracheal tube intubation. Hyper expansion of right middle lobe with collapse of upper and lower lobes noted per-operatively and middle lobe resection done securing the pulmonary vessels to middle lobe and middle lobar bronchus. Baby was successfully extubated on second post-operative day.

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http://orcid.org/0000-0003-3526-1684
DOI: http://doi.org/10.4038/sljs.v36i3.8525

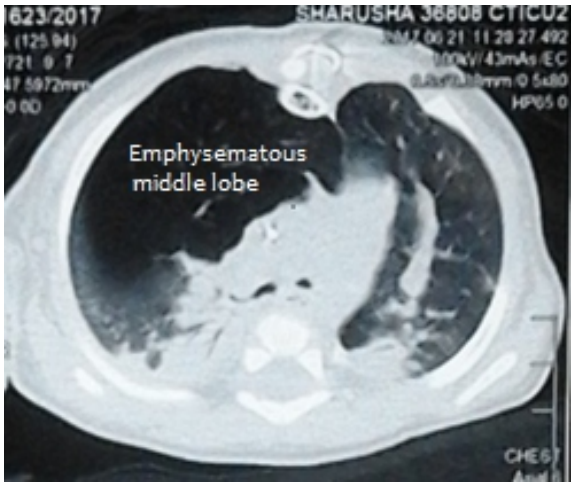

Figure 1. CECT chest demonstrating Middle lobe emphysema

Case 2

A six months old baby was investigated for cough, shortness of breath and fever for 1 week. A hyper lucent right lung was noted on Chest X-ray. CECT thorax revealed a hyperlucent anteromedial aspect of the right hemi thorax with mediastinal shift suggestive of either congenital lobar emphysema or congenital bullae involving right middle lobe. As the patient was progressive in symptoms thoracic team planned for the right thoracotomy and exploration.

Child was intubated with a single lumen endotracheal tube and underwent right axillary thoracotomy and exploration. The hyper expanded middle lobe compressing the upper and lower lobes was witnessed and middle lobectomy was performed after securing vascular pedicle and middle lobar bronchus. Rest of the surgical procedure was uneventful. Patient was extubated on the following day.

Case 3

A two months old baby girl transferred from a local hospital due to progressive shortness of breath, tachypnea, chest recessions and fever while managing as right upper lobar bronchopneumonia. Baby needed ventilator support for respiratory failure and the chest X ray revealed hyper lucent left lung fields with mediastinal shift to right side and inflammatory markings on right lung field. CECT revealed emphysematous left upper lobe with ipsilateral lower lobe collapse and right lower lobar consolidation and congenital

lobar emphysema of left upper lobe. Thoracotomy and exploration was planned early as child could not come out of the ventilator. Externally compressed left upper lobe bronchus was noted on pre- operative F.O.B.

Left axillary thoracotomy was done with single lumen endotracheal intubation. On exploration, emphysematous left upper lobe with collapsed left lower lobe was found and left upper lobectomy done after securing pulmonary vessels and left upper lobar bronchus. Child had a protracted recovery due to coexisted bronchopneumonia of the right lung.

Discussion

Two major pathological forms been discussed in the CLE as hypo alveolar and poly alveolar depending on the number of over distended alveoli present in histopathological specimens compared with normal lung architecture. Abnormalities of the

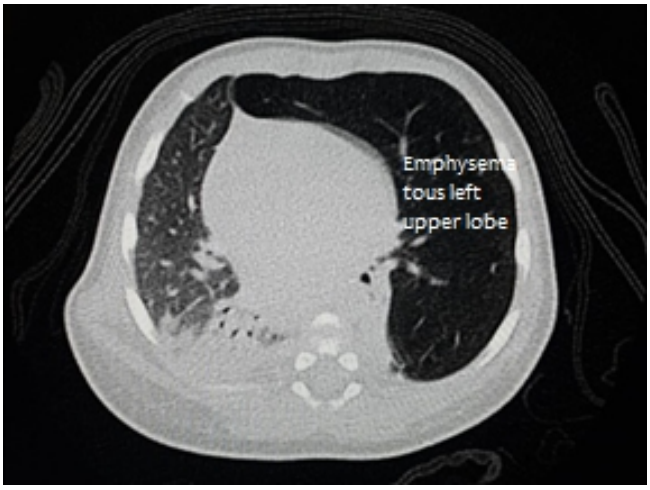

Figure 2. CECT chest demonstrating Left upper lobe emphysema.

bronchial cartilage development and resultant ball valve effect with air trapping , vascular abnormalities resulting bronchial stenosis and bronchogenic cysts and congenital cytomegalovirus infection have been postulated as possible etiopathogenesis of the condition [4]. In up to 15% of the cases there have been demonstrable congenital cardiac defects including VSD or patent ductus arteriosus [1, 3].

Upper lobes have more predilections for involvement in CLE. Left upper lobe (40 – 45%) is the most commonly affected preceded by right middle lobe (30%) and right upper lobe (20%). Hence right hemi thorax is the most commonly affected side [4].

Soon after the birth, the affected lobe is seen as homogeneously opaque region in chest X ray due to fetal lung fluid and distended lymphatic channels. Hyperlucency and diminished vascular lung markings with shifting of mediastinum and diaphragmatic depression are seen in established cases with clinically significant respiratory compromise. CECT of the chest is the investigation of choice for further anatomical details of the thorax in great detail.

Depending on the clinical presentation and radiological findings bronchial atresia with air trapping, congenital cystic adenomatous malformation (CCAM), congenital pulmonary airway malformation (CPAM), pulmonary arterial hypoplasia, pulmonary hypoplasia, Sawyer-James syndrome due to post infective obliterate bronchiolitis should be considered as the differential diagnoses.

Surgical resection of the affected lobe is the definitive management in CLE. The age of the child, clinical presentation, degree of respiratory compromise and associated heart disease are key factors to consider in timing of surgical intervention. In patients with cardiorespiratory compromise with mediastinal shift and compression of unaffected lobes necessitate lobectomy of affected segments. Histopathology of above mentioned cases revealed over distended alveoli confirming the diagnosis.

The diagnosis and management of CLE poses a great challenge upon clinicians in paediatric practice. High degree of clinical suspicion with timely thoracic surgical intervention can reduce associated morbidity and mortality.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Congenital lobar emphysema is a rare disorder which commonly presents as respiratory distress in newborns and infants.
- Timely thoracic surgical intervention can reduce morbidity and mortality associated with this condition.

CASE STUDY

Successful treatment of xanthogranulomatous pyelonephritis in an elderly lady with percutaneous drainage

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Keywords
Xanthogranulomatous pyelonephritis; renal cell carcinoma; renal calculi

Introduction

Xanthogranulomatous Pyelonephritis (XGP) is a severe form of pyelonephritis characterized by destructive changes to renal parenchyma due to chronic inflammation by bacterial infection. In general a significant number of patients undergo a nephrectomy as these lesions mimic renal tumors. If the diagnosis of XGP is suspected, a partial nephrectomy may be attempted. When the infection is extensive, a nephrectomy together with debridement of surrounding tissue may be needed. Less commonly, the condition can be treated with intravenous antibiotic alone or percutaneous drainage. Here, we report an elderly lady who presented with painful right lumbar mass. Magnetic Resonance Urography revealed a large renal collection with mixed density. The lady recovered following percutaneous drainage.

Case presentation

An 80 years old Chinese lady presented with right lumbar pain and mass for 2 weeks. It was associated with lethargy and appetite loss. She had a history of right renal stone which had been treated with open surgery 15 years ago. She had chronic kidney disease with baseline creatinine of 210umol/L. She had consumed over the counter analgesic whenever she had right lumbar pain. She had no diabetes mellitus. Dextrostix was 5.9mmol/L. Her blood pressure was 150/92mmHg and pulse rate was 116 beats per min. A right lumbar mass was felt. It was non-tender and measuring 20x15cm in size.

Her urine biochemistry revealed cloudy urine with negative nitrite and leucocytes of 2+. Urine culture and sensitivity had a negative yield. White cell counts were 13.2x10^9/L, haemoglobin 84g/L, platelet 77x10^9/L, urea 24.5mmol/L and creatinine 480umol/L. Her blood gas revealed compensated metabolic acidosis with pH 7.377, pCO2 25.8, base deficit of -9.4.

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Kidney ureter bladder radiography revealed right lumbar homogenous density with a large radiopaque stone. Ultrasound (USG) revealed grossly enlarged right kidney with heterogeneous echogenicity and multiple renal calculi. Non contrasted computed tomography of kidney, ureter and bladder (CT KUB) revealed diffusely enlarged right kidney measuring 14.3cm x10.7cm x10.5cm. The kidney contained cystic and complex solid components. There were multiple renal calculi largest 4.9cm in its diameter (Figure 1).

In view of suspicious features on CT KUB and USG, the diagnosis of renal tumour was not able to be excluded.

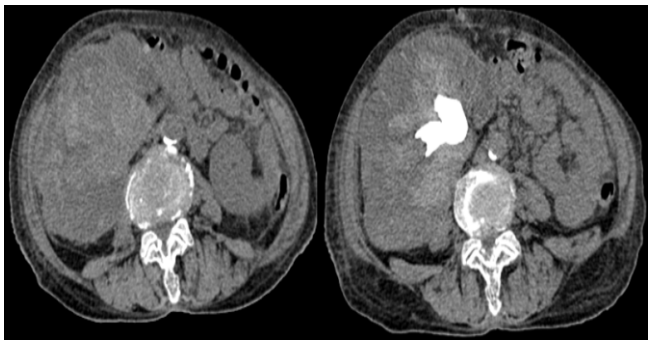

Figure 1. Plain CT axial view showing hypodensity area within the right renal mass

Therefore, magnetic resonance (MR) kidney, ureter & bladder was done due to impaired renal function. A large right renal collection with mixed density was identified. The centre and anterior portion of the lesion showed hypointense signal on T1 weighted imaging suggesting a fluid collection (Figure 2).

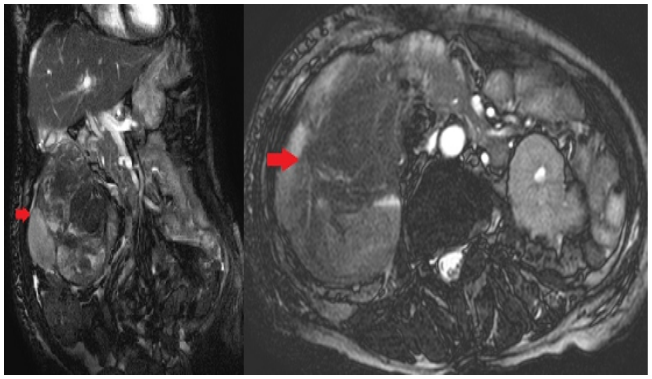

Figure 2. MRI showing right renal mass lesion (red arrow) in coronal and axial view

Based on the collective findings of all the imaging results, a diagnosis of xanthogranulomatous pyelonephritis was made. A percutaneous drain was inserted into the right renal collection.. One litre of haemopurulent fluid was drained on aspiration. Subsequently, it drained about hundred ml per day for 3 days. Her pain reduced and appetite was regained following the procedure.

Discussion

Xanthogranulomatous pyelonephritis is an uncommon and severe form of pyelonephritis with destructive changes involving renal parenchyma[1]. The entity commonly involve individual of the age of 45 to 55 years with female predominance. It present in 2 forms, either diffuse or local type. Clinical presentation and radiological findings may mimic a neoplastic aetiology [1, 2]. In the current case, we faced a similar challenge to differentiate the cause of the renal mass from a renal cell carcinoma.

Computed tomography (CT) scan is regarded as the main modality to diagnose the condition [1, 3].The CT findings may include hydronephrosis (90.9%), renal calculus (72.7%), pyonephrosis (45.5%), intraparenchymatous collection (45.5%), cortical renal atrophy (45.5%), nonfunctioning kidney (36.4%), abscess (36.4%), or perinephric fat accumulation (18.2%) [1].Hydronephrosis, renal calculus, and intraparenchymatous collections were present in the CT scan of the case. However, the non-contrasted nature of the scan limited the ability to differentiate from a malignant etiology.

Magnetic resonance imaging (MRI) gives extra information in addition to non- contrasted CT. As XGP presents like a pseudo-cystic mass, it generates low-intensity signal on T1-weighted images and high-intensity signal on T2-weighted images. This was observed in this case. Histologically in XGP, the renal parenchyma is replaced by lipid-laden foamy macrophages in combination with an inflammatory granuloma and a lymphoplasmacytic infiltrate. MRI is sensitive to identify the lipid laden foamy macrophages as high-intensity signal on spin-echo, T1-weighted images. However, angiomyolipoma (AML), retroperitoneal liposarcoma, and renal cell carcinoma (RCC) occasionally contain fatty tissue. The limitation of MRI is often described in differentiating RCC from two common benign entities which are minimal-fat AML and oncocytoma. AML demonstrated decreased T2 signal intensity, increased T1 signal intensity, high signal loss on opposed-phase images, and high ratio in signal intensity between early and delayed post contrast image. This overlaps with features of RCC on MRI. Similarly in the case of oncytoma, central scarring with areas of necrosis and local aggressive behaviour of perirenal fat invasion or renal vein branch invasion can mimic a RCC. Therefore, this confirmation of the renal mass pathology can only be relied on biopsy [4].

In this case, the diagnosis was made after consideration of the clinical history and combination of several radiological imaging modalities. The collective findings such as intra-parenchymatous renal collection and renal calculi prompted an infectious etiology. The immediate drainage of the lesion successfully relieved the symptoms of the elderly lady in short term. In conclusion, diagnosing XGP prior to any intervention remains difficult. The combination of CT and MRI may aid in differentiating from a renal cell carcinoma. Percutaneous drainage is essential measure to relieve acute symptoms. A subsequent nephrectomy maybe required for complete cure of this disease.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Xanthogranulomatous pyelonephritis can mimic renal cell carcinoma. Combination of clinical history and few imaging modalities can be useful to differentiate from renal cell carcinoma.
- The disease (XGP) is usually treated with nephrectomy. However, selected patients may benefit from initial percutaneous drainage with the intent of avoiding nephrectomy.
- MRI is an alternative imaging modality whenever doubt exists with routine imaging with radiography and computed tomography scan.

CASE STUDY

Mucinous adenocarcinoma of renal pelvis in a crossed ectopic kidney

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Keywords: Mucinous adenocarcinoma; renal pelvis; renal tumour; urolithiasis

Introduction

Most renal tumours arise from renal parenchyma. Urothelial carcinomas constitutes less than 10 % of renal tumours. Less than 1% of renal pelvic epithelial malignancies are adenocarcinoma [1]. Mucinous adenocarcinoma (MA) of renal pelvis is extremely rare and constitutes less than 0.3% of renal pelvic tumours [2]. We report a case of mucinous adenocarcinoma of crossed ectopic renal pelvis occurring in association with pyonephrosis secondary to obstruction by a large staghorn calculus.

Case report

A 46-year-old Sri Lankan Tamil male patient presented with right lower abdominal pain and fever for one week. He had haematuria on and off for the past 5 years for which he did not get treatment. He was passing milky urine for the last three months. He had a tender intra-abdominal mass palpable in the right iliac fossa.

His urine full report and blood analysis confirmed a severe urinary tract infection. The x ray KUB demonstrated a large staghorn calculus in the midline at the level of L5 vertebra (figure 1). Renal ultrasound (USS-KUB) and CT KUB revealed left kidney being situated below the right kidney. There was a cystic mass in left kidney with dilated pelvicalyceal system with no measurable renal cortex and a large staghorn calculus within renal pelvis. Right kidney and ureter were normal. The radiological diagnosis was crossed ectopia of nonfunctioning left kidney with large staghorn calculus and hydronephrosis (figure 1). He responded well to intravenous antibiotic treatment.

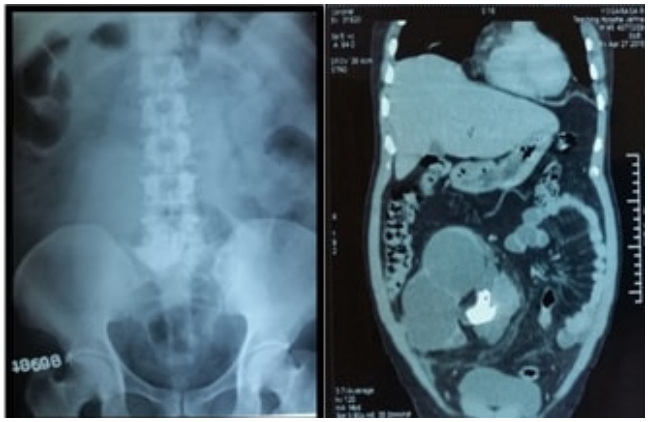

Figure 1. X ray KUB and CT KUB revealing large staghorn Calculus at the level of L5 vertebra level and crossed ectopia of left kidney with hydronephrosis and calculus respectively

A lower midline laparotomy & nephrectomy for non-functioning crossed ectopic left kidney was performed after a month. The crossed ectopic left kidney was found as a large cystic mass located in the right iliac fossa extending up to just left of the midline at the level of L5 vertebra. It was supplied by two separate vascular pedicles which were originating from the left common iliac artery. The right kidney and ureter were identified and preserved. Severe adhesions and inflammatory response were noted between the left kidney and perirenal tissue.

The enlarged kidney measured 11 x 10 x 08 cm with a large staghorn calculus measuring 5 x4 x 3 cm. The staghorn calculus was tightly impacted in the renal pelvis. A whitish

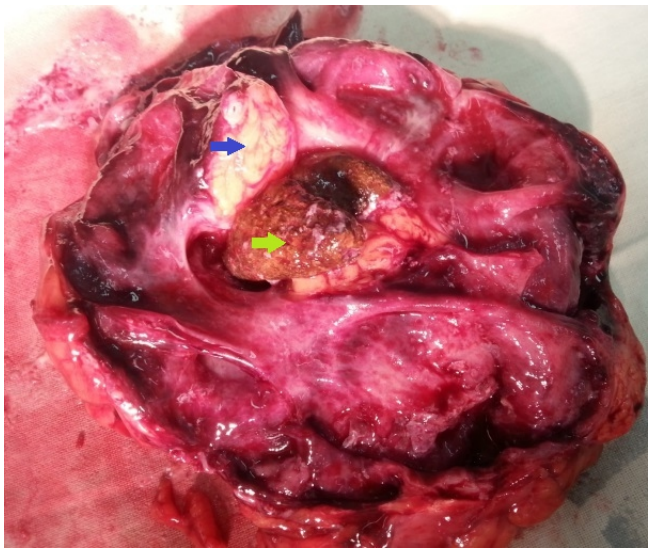

Figure 2. This image shows the tumour at pelvic ureteric junction (blue arrow) and staghorn calculus impacted within dilated pelvis (by green arrow).

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lobulated area was noted in the renal pelvis (figure 2).

Histopathology report of this specimen revealed excessive dilatation of pelvicalyceal system with mucinous metaplasia and dysplasia. Thinned out renal parenchyma had sclerosed glomeruli and tubules in fibrotic stroma. A whitish lobulated area near the pelviureteric junction, measuring 40x 30 x 30 mm, was a poorly differentiated adenocarcinoma composed of lobules and irregular cell islands in a haemorrhagic necrotic background. The tumour infiltrated into perinephric fat.

He had an uneventful post-operative recovery. Following oncology referral, he was given limited radiotherapy to left ectopic renal bed after insertion of indwelling ureteric (JJ) stent to the right ureter.

Discussion

Hasebe et al first described renal pelvic mucinous adenocarcinoma in 1960 (3). Since then few cases with mucinous adenocarcinoma have been reported mainly from Asian countries. MA occurring in ectopic kidney and horseshoe kidney are extremely rare [1].

The pathogenesis of MA is multi factorial. Glandular metaplasia of urothelium can occur due to chronic irritation, existing teratoma and epithelial sequestration during renal parenchymal development [2]. In our patient, the presence of staghorn calculus and pyonephrosis signifies the possibility of chronic irritation and ectopic nature of kidney points to the likelihood of epithelial sequestration. Apart from renal pelvis, the MA can be located in renal calyxes, ureter and in urinary bladder [4].

Most of the patients with MA have no symptoms. Few can present with passing blood or mucous in urine, loin pain and ballotable intra-abdominal masses [4]. Our patient presented with haematuria, pyuria, fever and abdominal pain mimicking urinary tract infection. He also had an intra-abdominal mass in right lower abdomen suggesting an ectopic nature of kidney.

Imaging plays a key role in the diagnosis and staging of renal tumours. Thus the management is usually decided upon the findings of CT – IVU. This may not be applicable in the case of mucinous adenocarcinoma of renal pelvis [5]. Furthermore the mucinous cystadenomas can have varying morphologies that are radiologically categorized in different Bosniak classifications [4]. Decision making on the need for surgical intervention is quite difficult in this situation. In our patient the CT – KUB revealed an obstructed crossed ectopic left kidney with hydronephrosis and thinned out renal cortex suggesting a non functioning renal system. Thus an IVU was not performed in this patient.

Our patient had clinical and radiological diagnosis of crossed ectopic left kidney with hydronephrosis due to obstruction of renal pelvis by a staghorn calculus. Since it was a non-functioning kidney with pyonephrosis, it was decided to proceed with nephrectomy. MA of renal pelvis was only identified by histopathological examination of the nephrectomy specimen.

The exact diagnosis of this type of tumour is made after the pathologic examination of specimen. Characteristic pathologic features of MA are the stratified columnar epithelium with vacuolated cytoplasm and hyperchromatic pleomorphic nuclei. Signet ring cells in mucin substance are also seen. Infiltration into renal parenchyma, breach in the capsule and extension into the perinephric fat are features of local infiltration [2]. Clinical suspicion of MA is of utmost importance when dealing with cysts filled with large pools of mucin and gelatinous solid areas [1]. This type of tumour has been reported in congenitally abnormal kidneys [6]. Our patient had locally advanced MA based on histopathological report.

The treatment of MA is nephro ureterectomy with removal of cuff of urinary bladder [1]. The ideal treatment may not be possible in most instances as MA is seldom diagnosed pre operatively. Careful dissection of cysts is of utmost importance to prevent tumour seedling. In this patient the left lower ureter was not excised and he would need a second exploration for the lower ureter. This case was discussed at the multidisciplinary team (MDT) meeting and it was decided to delay the second surgery as he had locally advanced tumour with infiltration of renal bed. It was also decided to give limited radiotherapy to renal bed of ectopic left kidney. Very good response to chemotherapy with oxaliplatin, fluorouracil, leucovorin (FOLFOX4) has been reported in similar tumours occurring in urinary bladder [7]. Because of the high local recurrence of this tumour, it is mandatory that the patient will have to be followed up regularly at frequent intervals. Regular screening with USS-KUB and flexible cystoscopy has been planned for this patient.

Conclusion

MA of renal pelvis is rare. MA occurring in crossed ectopic kidney is extremely rare. In our patient staghorn calculus, hydronephrosis, infection, inflammation and chronic irritation could have predisposed to glandular metaplasia in renal pelvis leading to MA.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- MA is rare tumour of kidney and pre-operative diagnosis is difficult.
- Possibility of MA should be kept in mind in patients with enlarged cystic ectopic non-functioning kidney.

CASE REPORT

Catamenial pneumothorax sans thoracic endometrial deposits

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Key words: Recurrent spontaneous pneumothorax; catamenial pneumothorax; VATS pleurectomy

Introduction

Although spontaneous pneumothorax is a more common entity among males a higher number of recurrences are observed in females. Catamenial or endometrial related pneumothorax is attributed as the main cause for this high recurrence rate [1]. Recognition of this entity in a female presenting with two or more episodes of spontaneous pneumothorax is important, as treatment can prevent another episode. Here we present a case report of a 43 year old woman, who was treated for recurrent catamenial pneumothoraces at National Hospital for Respiratory Diseases (NHRD), Welisara.

Case Report

A 43 year old female was referred to NHRD following two episodes of spontaneous right sided pneumothoraces. The episodes occurred over two consecutive months, symptoms starting one day before her menstrual periods began. The first episode was treated with a tube thoracostomy at the local hospital. A contrast CT of the chest performed one day after tube thoracocostomy was unremarkable apart from a small residual pneumothorax on the right side.

She had a history of treatment for primary subfertility and a diagnostic laparoscopy done 6 years before was apparently normal. One year back she underwent excision of a lump at her left groin and the histology was that of an endometrioma.

As the clinical picture was suggestive of endometrial related recurrent spontaneous pneumothorax she was scheduled for a video assisted thoracoscopic (VATS) procedure under general anaesthesia with single lung ventilation. A three port entry was utilized. The right dome of the diaphragm was normal. The right lung was unremarkable except for a small area suspicious for scarring noted at the apex of the upper lobe. A wedge resection of that segment was done using endoGIA staplers and a parietal pleurectomy was performed. Apart from a chest infection which was treated with intravenous antibiotics her post-operative recovery was uneventful. The histology of the resected lung specimen was negative for endometrial tissue. The patient was referred back to her gynaecology team to continue their management of possible endometriosis and associated subfertility.

Discussion

Catamenial pneumothorax is defined as a spontaneous recurrent pneumothorax in a women of reproductive age, with a temporal relationship with menstruation [2]. Although the exact timing in relation to the menses is variable across the literature [2], a time frame of 24 hours before and up to 72 hours after the onset of menses is reasonable [1]. This accounts for 3-6% of spontaneous pneumothoraces in females, and the mean age of onset is between 32-35 years [3]. The majority are seen in the right side (87.5-100%) but rarely left sided and bilateral cases have been reported [2].

The diagnosis is entirely clinical, depending mostly on the history. The patient will present with symptoms due to pneumothorax such as chest pain, shortness of breath and cough. These symptoms are recurrent and will have a temporal relationship with menstrual periods. Primary or secondary subfertility, a diagnosis of pelvic endometriosis, a history of uterine procedures and rarely catamenial haemothorax/haemoptysis/haemopneumothorax can be associations [2]. Our patient had a history of primary subfertility. Although a diagnosis of pelvic endometriosis was not confirmed, ectopic endometrial tissue was found in the excised groin lump of hers, which fits in to the diagnosis of endometriosis.

Radiologically there are no pathognomonic features to differentiate this condition, but chest X ray, CT and rarely MRI can be helpful [2]. Elevated CA 125 levels are associated with endometriosis, and a high CA 125 level with a spontaneous pneumothorax can support a diagnosis of endometriosis associated pneumothorax [2].

The pathogenesis of this entity is still not clearly understood [4]. During menses the mucous plug occluding the opening of the uterine cervix is absent, and as a result air enters the

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DOI: http://doi.org/10.4038/sljs.v36i3.8528

peritoneal cavity through the female genital tract. This air can enter the thorax through congenital or acquired defects in the diaphragm causing a pneumothorax. Another theory postulates that prostaglandin F2 levels which increase with menstruation causes strong contractions of blood vessels and bronchioles leading to rupture of some alveoli [1, 3]. The above theories are independent of endometriosis.

Possible mechanisms for endometrial deposits in the thoracic cavity are by retrograde menstruation, via lymphatic or blood borne emboli of endometrial tissue, and metaplastic change of mesothelial cells lining the pleura [4]. During menstruation endometrial tissue deposits on the lung surface are sloughed off causing air leaks.

Characteristic pathological lesions associated with this condition are defects in the diaphragm, nodules and spots visualized in the diaphragm, visceral and parietal pleura [2]. However rarely some cases may have none of these abnormalities [2], as in our patient. In the study by Marshall et al, 2 out of 8 (25%) with catamenial pneumothorax did not have any abnormality identified on initial thoracoscopy but one of them one was subsequently found to have lung parenchymal endometrial deposits when she presented with a recurrence [4]. According to Alifanao et al, out of eight patients with catamenial pneumothorax all 8 (100%) had defects in the diaphragm and 7/8 (87.5%) had histologically proven endometrial deposits in the diaphragm [5]. This strong association between diaphragmatic defects and endometrial deposits was seen in the study by Marshall et al as well and they postulated that the defects were in fact caused by the deposits [4].

Surgery is the mainstay of treatment of this condition, as it is associated with less recurrences compared to medical treatment alone with hormonal manipulation [1, 2]. Diaphragmatic defects should be looked for and either resected or repaired [4]. Identified bullae, endometrial deposits on the lung surface should be excised/ wedge resected and a pleurodesis or a plurectomy performed [2]. A minimally invasive VATS approach is preferred [1, 2]. However a more invasive mini thoracotomy/thoracotomy may be necessary for reoperations or to repair a large diaphragmatic defect [2]. Surgical treatment for catamenial pneumothorax is associated with zero mortality and a negligible morbidity. However even with surgical treatment recurrence rates can be as high as 40% at 4 years [4].

Endocrine treatment should be offered for coexisting endometriosis. For those who are unfit for general anaesthesia and surgery endocrine therapy can be used as the sole treatment modality to prevent recurrences [3].

Conclusions

The diagnosis of catamenial pneumothorax as a cause for recurrent spontaneous pneumothorax in females is increasing, mainly due to heightened awareness of the condition. Once diagnosed surgery is the mainstay of treatment, preferably with a minimally invasive VATS approach. Endocrine treatment should be offered for coexisting endometriosis.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Catamenial pneumothorax should be considered as one cause for recurrent pneumothorax in young females.
- Endometriosis and congenital or acquired defects in the diaphragm are the most common associated factors with this condition.
- Surgery is the mainstay of treatment with a VATS approach and medical management plays a secondary role.

Pseudopapillary tumour of pancreas in a 13 year old girl

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Teaching Hospital Batticaloa

Keywords: Solid pseudopapillary tumour; Frantz's tumour

Introduction

Solid pseudopapillary tumour of the pancreas is rare. It is a low grade malignant tumour with minimal metastatic potential among the children [1] [2]. The tumour has a good prognosis following complete surgical excision. Here we report a case of a solid pseudopapillary tumour presented as an acute abdomen.

Case report

A 13 year old previously healthy girl admitted to Teaching Hospital Batticaloa with a history of abdominal pain for 1 week duration, worsened during last 24 hours. It was associated with fever, nausea and vomiting for one day. She had one episode of hematemesis. Urinary and bowel habits were normal. There was no recent weight loss or jaundice. On admission she had a tachycardia (115bpm) but blood pressure was normal. Abdominal examination revealed an epigastric tender mass. There was guarding but no rigidity. She was clinically pale. Urgent full blood count showed neutrophil leucocytosis (18,740 / mm3) with low Hb (9.6g/dl). Renal, liver profile, urine analysis and coagulation screening were normal. Serum amylase was within normal limits.

ESR was found elevated to 55mm/1st hr as well as the CRP to 46. Urgent ultrasound abdomen showed a 14.3cm *9.6cm*10.8cm highly vascular heterogenic lesion at epigastric region. She was kept under close monitoring. IV antibiotics were started. Consequent upper gastrointestinal endoscopic study revealed erosive antral gastritis with reflux disease. Computed tomography of the abdomen was not done due to machine failure the parents declined transfer to another institution for this purpose. Exploratary laparotomy revealed highly vascular tumour arising from retro peritoneal area and involving middle body of the pancreas. 12cm*10cm in dimension. Peritoneal survey was negative for any metastatic deposit. The tumour removed with the body of pancreas.

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The head, neck and tail of the pancreas were preserved considering the child was too young for insulin dependency. Common bile duct, splenic artery portal vein were identified and preserved. The remaining parts of the pancreas were anastomosed to preserve the continuity. Histology revealed capsulated solid pseudopapillary tumour of pancreas with no evidence of capsular or perineural invasion. Areas of necrosis and haemorrhages seen microscopically. Resection margins were negative. Post-operative period and follow up was uneventful. The patient was directed for oncology follow up. But no additional therapy was instituted.

Discussion

Solid pseudopapillary tumour is referred to as Frantz's tumour after ther person who described it in 1959.

It commonly affects the young and has a female preponderance [1]. It is a low grade malignant neoplasm. Abdominal pain is a common initial presentation but some present with asymptomatic abdominal mass [2].This patient presented with abdominal pain and the abdominal mass elicited by the clinical examination. Elevated amylase levels were observed in some cases, but this patient's serum amylase was within normal limit. In contrast to the adults, children with solid pseudopapillary tumour have a low distant metastatic potential which corresponds with our case as well [3] [4].

The tumour commonly involves pancreatic tail followed by the head, and the involvement of the body is rare. But in this case the tumour was localized to the body, sparing head and tail [4]. Computed Tomography (CT) of abdomen was done preoperatively in most of the literature[3][4]. Surgical resection without preoperative CT was challenging for us. Therefore doing preoperative imaging will be a benefit for surgeon to analyse.

But our case due to the above mentioned reasons, deviated from the routine norm. Even-though in the early years pancreatoduodenectomy was performed, newer studies recommend enucleation of the tumour or partial pancreatectomy [4]. Also literature recommends complete surgical resection of the tumour alone is the best treatment [4]. In this case, we managed the patient alone with complete surgical resection of tumour as with the literature and the post-operative follow up was uneventful.

Conclusions

Solid Pseudopapillary tumour of pancreas is a low grade malignant disease of with minimal metastatic potential. Most of the time surgical enucleation alone will be sufficient.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points

- Solid Pseudopapillary tumour commonly present with abdominal pain but asymptomatic cases also reported.
- Common sites of origin are Pancreatic tail and head in the descending order.
- It has a low metastatic potential in children.
- Preoperative imaging of the tumour would be a fringe benefit for surgery.
- Most of the time surgical enucleation alone will be sufficient.

Symptomatic large infrarenal abdominal aortic aneurysm with concomitant pelvic tumour

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Key words: Abdominal aortic aneurysm; pelvic tumour

Introduction

The management of abdominal aortic aneurysm (AAA) with the presence of intra-abdominal tumour is controversial and the management plan remains a challenge to the clinician. Most of the literatures published has been focusing on the option of management and the risk involved with either a staged approach or a simultaneous approach to the management of concomitant AAA and intraabdominal malignancies.

We report a case of symptomatic huge abdominal aortic aneurysm with incidental finding of a huge pelvic tumour.

Case Presentation

An 86-year-old lady, presented with a complaint of left-sided abdominal pain for one-week duration. The pain was pricking, not relieved with simple analgesics and occasionally radiating to the back. Abdominal examination revealed a pulsatile mass occupying her left side of abdomen with slight tenderness on palpation. Her distal pulses were palpable with good volume.

CT angiography of abdomen revealed a huge fusiform aneurysm of the infrarenal abdominal aorta, measuring 7.5 x 8 cm with no evidence of contrast leakage (figure 1). There was an incidental finding of a huge multiloculated cystic mass at both adnexa measuring 12.9 x 13 x 15 cm, possibly of ovarian origin (figure 2).

Her tumour markers were taken following the findings of the CTA to rule out potential malignant pelvic tumour. Ca 125, bHCG and alpha-fetoprotein all within normal value. Further staging assessment of the mass were carried out such as chest X-ray, sigmoidoscopy revealed no significant abnormality. A referral was made to the gynaecologist for a combined surgery. In view of the huge pelvic tumour, a planned perioperative ureteric stenting was performed 4 days earlier by the urologist.

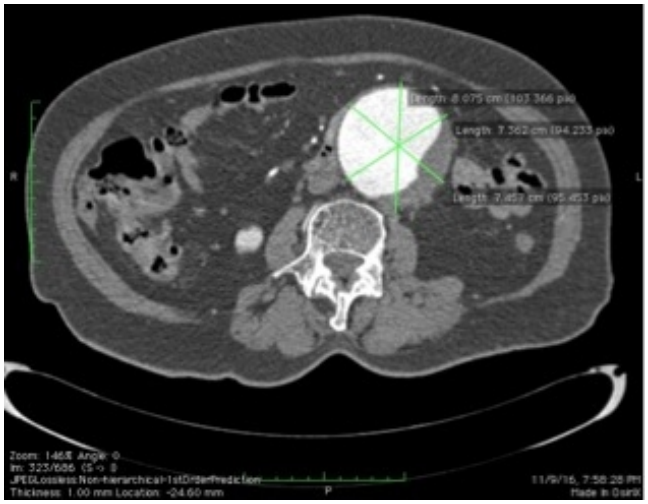

Figure 1. 8cm infrarenal AAA

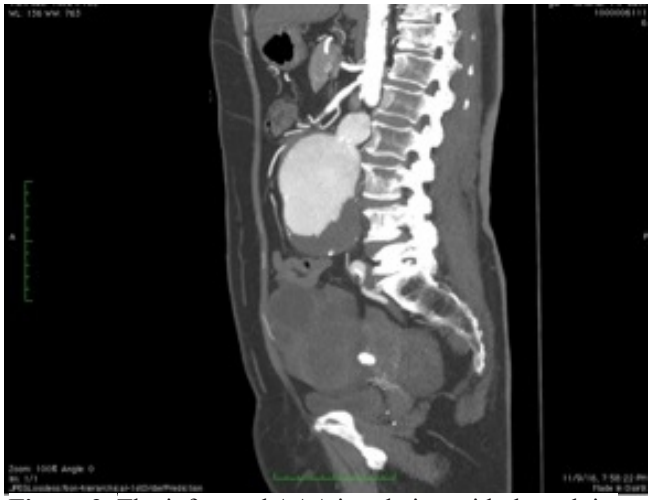

Figure 2. The infrarenal AAA in relation with the pelvic tumour

Bilateral salphingo-ophrectomy was carried out by the gynaecologist. After a vigorous wash of the abdomen cavity, the 8 cm aneurysm was repaired with a bifurcated in-lay graft. The infrarenal clamping time was about 45 minutes. The peritoneal fluids were sent for cytology assessment as well as culture and sensitivity test. The postoperative period was uneventful and she was discharged after 8 days with antiplatelet. Her first post-operative clinic assessment one month after that was uneventful. Her renal profile was unchanged from the pre-operative readings. Histopathological report of the ovarian tumour revealed a benign serous cystadenofibroma of both of the ovaries.

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Discussion

The occurrence of AAA and visceral malignancy increases with advancing age. At the time of open AAA repair, the chance to encounter intra-abdominal malignant disease has been found in up to 4% of patients [1]. Surgical treatment remains the best option of management on these two potentially life-threatening conditions, but the best appropriate timing of intervention remains controversial. Graft infection basically remain the main concern when the aneurysmal repair performed simultaneously with other non-vascular intraabdominal procedures [2]. The key point for simultaneous approach open surgery is choosing the best timing for the intervention, balancing with risk of aneurysmal rupture and complication from the intraabdominal malignancy such as obstruction and bleeding. Repeat anaesthesia in a staged procedure carries a risk especially in patients at advanced age groups [4].

The occurrences of AAA with concomitant intraabdominal malignancy are seldom encountered. Majority of the reported cases are associated with colorectal malignancy, bladder tumour and haematological malignancy including tumour of the aorta itself. However, most patients are generally asymptomatic, and one lesion is often discovered incidentally during imaging assessment of the other lesion [2]. The decision for surgery either through a one-staged or two-stage procedure will depend on the behaviour of each disease. A ruptured symptomatic AAA require an urgent emergency surgery. However, elective repair of an AAA with co-existent large abdominal tumour often carries dilemmas in decision making and eventual management. For an instance, the resection of the colorectal tumour may render the aortic graft to become infected if a single stage procedure were carried out. Overall, the behaviour and invasiveness of the tumour also needs to be evaluated in detail prior making the final decision regarding the sequence of surgical intervention [4].

The discovery of pelvic tumour in this case was only encountered during the initial imaging assessment of her pulsatile abdominal mass. She had minimal symptoms until the time of presenting to hospital. Thorough assessment of the pelvic tumour has revealed a benign lesion biochemically and radiologically except for the size.

A two-stage procedure if it erre to be performed, would carry additional risks to the patient. For instance, proceeding with the AAA reconstruc-tion first could pose significant challenges during surgical exposure and vascular control due to the extreme size of the pelvic tumour. Furthermore, the extreme size of the tumour may actually carry the risk of intra-operative iatrogenic rupture thus predisposing to tumour spillage and contamination. The tumour if deemed to be malignant, may cause recurrent deposit near the graft, and expose it to the risk of thrombosis. If the pelvic tumour were to be excised first, the second-stage procedure for reconstruction of the AAA would also be deemed difficult due to the scaring of the abdominal cavity and the retroperitoneum [3].

Conclusion

In conclusion, managing an AAA in an elderly patient with concomitant tumour carries its own risks in relation to the patient's physiological reserve, the size of the AAA and tumour behaviour. In such cases, endovascular intervention such as EVAR (Endovascular Aneurysm Repair) would be a good option of surgery and complication such as heart failure and graft infection can be prevented. Unfortunately due to the acute angulation of the iliac artery in this case has made the option out of the way.

A thorough perioperative discussion and plan with the involved units plays an important role for the success of such major surgery. Presence of invasive malignant tumour may render the AAA to be treated conservatively provided there's no evidence of leaking or rupture. Conversely, in the presence of a presumed benign tumour, an informed decision needs to be made regarding staged or simultaneous resection and repair. Primary determinants in the eventual decision regarding staged or simultaneous surgery depends on patient's physiological reserve to tolerate prolonged surgery, size of the AAA and tumour biology.

Finally, take home messages for a better care of patient in such circumstances will includes thorough perioperative discussion with the involved units for a good postoperative result which also depends on patient's overall physiological reserve and anatomical consideration of the lesion.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- A thorough perioperative discussion and plan with the involved units plays an important role for the success of such major surgery.
- Option for a single versus 2-staged procedure in such patient depends on patient's overall physiological reserve and anatomical consideration of the lesion.

A rare presentation of tuberculosis

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Keywords: Tuberculous synovitis; arthritis, extrapulmonary;

Introduction

Tuberculosis is characterized by chronic granulomatous inflammation, caseous necrosis and Langhans type giant cells. The disease burden of tuberculosis in developing countries is catastrophic. Among patients with tuberculosis, less than 1% was reported to have lesions on hand [1].

Case presentation

A 54-year-old female, a diagnosed patient with autoimmune haemolytic anaemia on immunosuppressive medications, presented with a swelling in the dorsal surface of the left wrist for six months. The swelling was painful and gradually enlarging. She denied fever. On examination, there was generalized swelling of the hand, redness, warmth with pitting oedema. Hand functions were markedly restricted with a reduced range of movements at the metacarpophalangeal, interphalangeal and wrist joints. Her Erythrocyte Sedimentation Rate (ESR) 64 mm at 1 hour, C-Reactive Protein 7 mg/dl, White Blood Count $7.21x10^{9}$ L-1 with 86% neutrophils, 0.4% lymphocytes and $580x10^{-9}$ L-1 platelets. She was initially suspected to have cellulitis. She did not respond to a ten-day course of intravenous Flucloxacillin.

She was then treated with a trial of Colchicine and Non-Steroidal Anti-Inflammatory Drugs suspecting gout. Her symptoms failed to improve with treatment. After five months, she developed a tender, laxly cystic lump in the dorsal aspect of the left wrist (Figure 1a). It cross fluctuated with a less prominent lump in the volar aspect of the wrist. Distal sensations were intact. X-ray of the wrist showed severe osteopenia and patchy intramedullary, endostial erosions with preservation of joint space. Magnetic Resonance Imaging (MRI) showed regional bone oedema, osteopenia and erosions, joint effusions with thickened synovium and multiple fluid pockets surrounding tendons suggestive of tuberculous arthritis (Figure 1b). MRI features

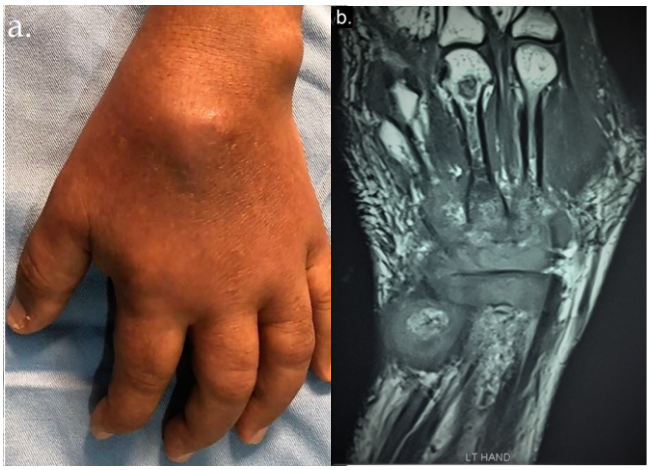

Figure 1a. Lump on the dorsal aspect of the wrist
Figure 1b. Magnetic Resonance Imaging of the wrist showing tuberculous arthritic changes

also suggested reflex sympathetic dystrophy. Upon further questioning, she denied a chronic cough, pyrexia, night sweats and weight loss. The patient did not have any past or contact history of tuberculosis. Chest X-ray had no evidence of tuberculosis. During excision biopsy of the lump, a volar wrist cold abscess was detected with diffuse synovial hypertrophy and caseous material. Histology showed multiple caseating granulomas within a heavily inflamed stroma with inflammatory cells surrounding sheets of histiocytes. Mycobacterium tuberculosis was isolated in tissue culture.

She was started on intensive antituberculosis treatment (Isoniazid, Rifampicin, Pyrazinamide, Ethambutol). Due to the liver function derangement, her drugs were withdrawn. Once liver functions were normalized, the desensitizing regime was commenced. Currently, she is on Ethambutol, Streptomycin and Levofloxacin which are well tolerated by the patient.

Discussion

Extrapulmonary tuberculosis manifestations occur as a result of the immune deficiency in individuals with existing medical conditions [2]. Due to the rare occurrence of hand infection in tuberculosis; the diagnosis is made rather later in the course of the disease [3]. Bayram, et al., reported a case of tuberculous synovitis presenting with non-specific symptoms such as

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DOI: http://doi.org/10.4038/sljs.v36i3.8531

swelling and pain at the wrist [4]. In most instances as observed in our case, the patients are treated for arthritic diseases or cellulitis at the initial encounter by the medical practitioners [4]. These patients may or may not have history and examination findings suggestive of tuberculosis. Clinicians should be aware of the unusual sites of tuberculous infections as it is a common disease in our country.

A similar picture of monoarthritis could occur due to chronic infectious tenosynovitis, tumours, and gout. Chronic regional pain syndrome (CRPS) such as reflex sympathetic dystrophy is a diagnosis of exclusion. Although the patient had pain and swelling suggestive of CRPS, she did not have abnormal sudomotor activity in the region of pain. The surgical and histopathological findings were also suggestive of a tuberculous disease. A high ESR value is seen in most cases of tuberculous synovitis [1,3]. However, the ESR level can be high in other causes of monoarthritis. X-ray findings of swelling in the soft tissues and sometimes periarticular osteoporosis were also seen in tuberculous joint involvement [1,4]. Synovitis due to tuberculosis is seen in Magnetic Resonance Imaging as thickening of the adjacent tendons, osteomyelitis, joint effusions erosions into bones and nerve entrapment [2]. Diagnosis confirmation involves histology and microbiology analysis [3].

There were studies comparing anti-tuberculin drugs versus surgery combined with anti-tuberculin medication; in which both methods were found to be equally effective [1]. In our case, we did a radical synovectomy followed by antituberculous medication. The patient is currently showing improvement with the treatment, nevertheless, she needs to be followed up. In cases, with tuberculous synovitis, the intensive treatment phase continues for two months followed by a continuation phase regimen for seven months [5].

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Hand tuberculosis infection is an uncommon presentation
- Diagnosis requires Magnetic Resonance Imaging (MRI) with confirmation by histopathology and cultures for tuberculosis
- Surgical treatment should be coupled with antituberculous treatment to avoid recurrence
- A high degree of suspicion is needed to diagnose extrapulmonary tuberculous manifestations

Adenoid cystic carcinoma of the maxilla

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Key words: Adenoid cystic carcinoma; carcinoma of maxilla; maxillectomy; perineural spread

Introduction

Adenoid cystic carcinomas (ACC) are malignant tumours of secretory glands mainly affecting major and minor salivary glands of the head and neck. It also involves lacrimal glands, ceruminous glands and occasionally female genital excretory glands. Although palate, the floor of the mouth and buccal mucosa are commonly affected, uncommon sites like lungs (tracheobronchial tree), oesophagus, also been reported [1].

ACCs are the malignancy in lacrimal glands and 2nd most common malignancy in salivary glands. They are well known for the prolonged unpredictable clinical course and the tendency of delayed metastasis. Lungs are the most commonly involved metastatic sites followed by bone, brain, kidney and liver. Tumour size, grade, stage, lymph node involvement, neural invasion and margin status are the most important prognostic factors [2]. Long-term survival can be achieved by a combination of surgery and radiotherapy. Immunohistochemistry prevents misdiagnosis and helps to determine appropriate multimodality management protocol. Here we present a rare case of adenoid cystic carcinoma affecting the palate and maxillary sinus which was managed with surgery and radiotherapy at Chittaranjan National Cancer Institute, Kolkata, and the centre of Eastern India.

Case report

A 77 years old male presented with slowly progressing hard bony swelling in right half of roof of the mouth for several years but recently increasing in size during the last 3 months. He was a regular smoker for last 55 years. but not alcoholic. He gave the history of nose blockade for the last 3 months with numbness over the right side of upper lip. Extra oral examination showed the fullness of the right maxillary region without any cervical lymphadenopathy. Intraoral examination showed an oval-shaped solitary, non-tender, firm, fixed, erythematous mass with diffuse borders in right maxilla involving hard palate and extending anteriorly from palatal rughae to posteriorly up to soft palate, medially 1 cm away from midline to laterally from lateral incisor to 1st molar tooth without any mucosal ulceration. Teeth in relation to the swelling were non-mobile / non-carious / non-tender.

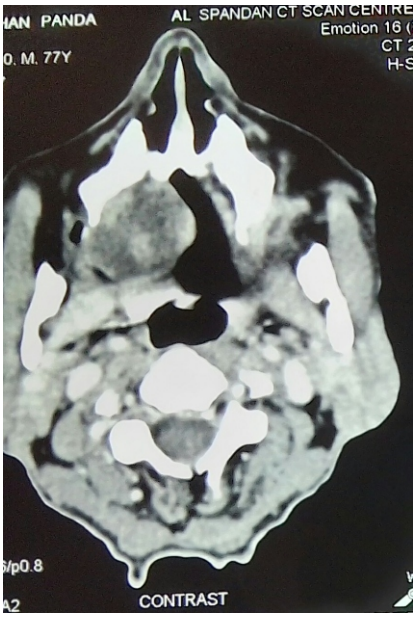

Figure 1. CT Scan showing the tumour

Panoramic radiograph showed a unilocular radiolucency obliterating the right maxillary sinus and involving the cortical plate above the lesion. Contrast-enhanced CT scan showed an enhanced 5 x 4.5 x 3 cm mass in right maxilla with the destruction of maxillary sinus and involvement of hard palate. Fat-suppressed T1 weighted MRI with gadolinium enhancement showed a high-density tumour in right palatal region extending to the buccal region of the alveolar bone in coronal section and a high-intensity area on right half of hard palate with the involvement of the major palatine nerve in axial section. No cervical lymphadenopathy, metastatic site or any other second primary was seen radiologically.

Incisional biopsy showed monomorphic cells arranged in the glandular and canalicular pattern with focal areas of cribriforming with mucin-like material inside. Fibrous stroma was seen with areas of hyalinization and islands of uniform cells arranged in the cord-like pattern with hyperchromatic

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DOI: http://doi.org/10.4038/sljs.v36i3.8532

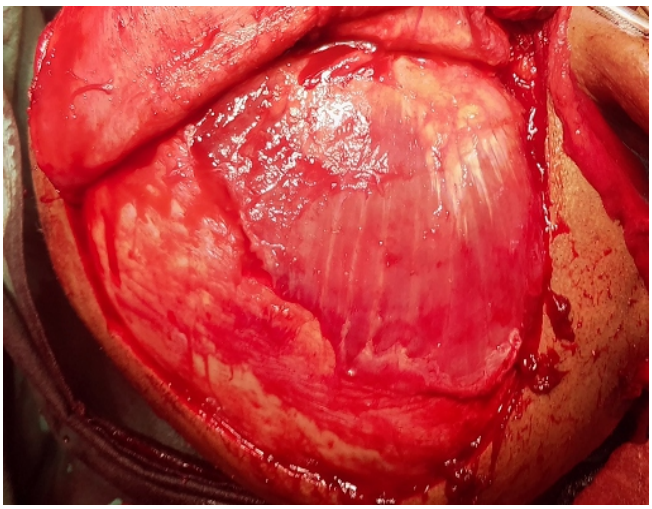

Figure 2. Intraoperative photo

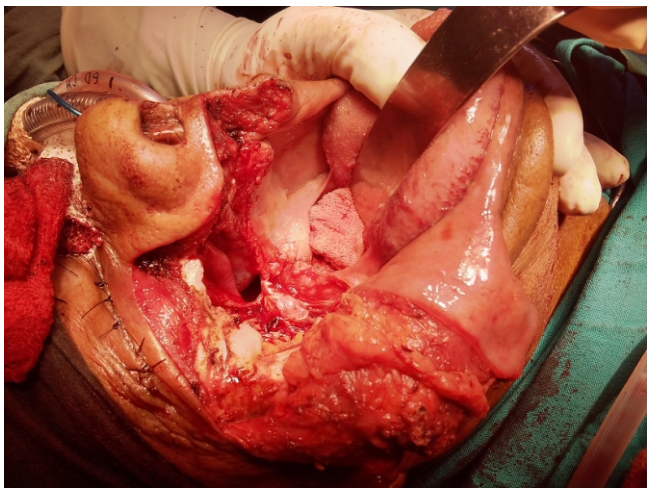

Figure 3. Harvesting of temporalis flap

nuclei enclosing round to oval pseudocystic spaces containing eosinophilic coagulum. Immunohistochemistry showed positive reaction pattern to S-110 (diffusely cytoplasmic and nuclear staining), pan-cytokeratin (AE1/AE3) (cell membrane staining) , Vimentin , p53 , C-Kit , Alpha Smooth Muscle Actin , p63 and a ki 67 labelling index of 60% (nuclear staining). CD 117 was strongly positive confirming the cribriform type of ACC. The patient was treated with right-sided subtotal maxillectomy followed by reconstruction with temporalis muscle flap. Histopathology showed adenoid cystic carcinoma with a clear margin, no lymphovascular invasion was identified but the presence of perineural invasion was noticed. The maxillary cavity was treated with adjuvant radiotherapy of 60 Gy delivered in 30 fractions. The patient is surviving for 2 years now without any locoregional or distant recurrence.

Discussion

ACCs were first described by Bilroth in 1856 as 'Cylindroma' due to its characteristic histological appearance. It is a very rare malignancy accounting for <1% of all head and neck malignancies and around 10% of all salivary gland neoplasms. ACCs show female predilection with a female: male ratio of 1.2: 1. These are aggressive, relentless, slowly growing tumours with the insidious destruction of surrounding tissues and perineural invasion. Pain is often an early sign (even before the appearance of swelling) because of neurotropism. Perineural invasion is the pathognomonic factor responsible for high rate of local recurrence. Lymph node involvement is rare and is usually caused by contiguous spread rather than lymphatic permeation or embolization [3].

Three distinct patterns of ACCs have been described: solid, cribriform and tubuloductal, cribriform being the most common, solid having worst prognosis and highest recurrence rates. 5 years recurrence rates of 100%, 89% and 59% have been reported for these three patterns respectively [4]. Differential diagnoses are polymorphous low-grade adenocarcinoma (PLGA), basal cell adenoma (BCA), mixed tumour and basaloid squamous cell carcinoma (BSCC). Immunohistochemistry demonstrates pseudocysts positive for periodic acid Schiff (PAS) and alcian blue and contain basement membrane components like type IV collagen, heparin sulfate and laminin isoforms. Epithelial cells are positive for carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA), duct lining cells are positive for CD-117 and myoepithelial cells are positive for S-100 protein, calponin, p63, smooth muscle actin and myosin. Perineural invasion is often indicated by the presence of S-100, glial fibrillary acidic protein (GFAP) and neural cell adhesion molecule whereas p53 is often involved with tumour progression and recurrence. High Ki-67 index and presence of more than 50% solid areas also indicate an aggressive clinical course. Alterations in chromosomes 6q, 9p and 17p 12-13 are most frequently observed [4].

CT and MRI are very useful to delineate a tumour, to plan extent of surgery and to look out for recurrences on postoperative follow-up, especially to determine the submucosal extent and perineural spread which can be difficult to assess clinically. The centre of the lesion usually presents with low density on CT, a characteristic feature that can help to differentiate it from squamous cell carcinomas. The changes on MRI are characterized solely by an increase in intensity with the preservation of the shape.

Surgical excision with oncologically acceptable margins and adjuvant radiation remains the mainstay of treatment with a 5 and 10 years survival rate of 77% and 57% respectively. Chemotherapy is yet to find a role in ACCs. Neck dissection should be performed in presence of clinically and/or radiologically evident cervical lymphatic metastasis. Prognosis is highly dependent on a solid histological type, perineural spread, and distant metastasis and locoregional recurrences. Clinically and radiologically undetectable micro invasion of bones often leads to locoregional recurrence even

after successful treatment completion. Bone metastases usually correspond to rapid tumour dissemination and death, whereas lung metastasis leads to a less aggressive clinical course. Hence, a close surveillance and long-term follow up is recommended to prevent local recurrence and distant metastasis [5].

Conclusion

ACCs are one of the most destructive and unpredictable malignancies with an indolent growth pattern. They often show aggressive behaviour, infiltrating into surrounding tissues and may present with regional lymphadenopathy and/or distant metastasis at the time of discovery. Early diagnosis and adequate surgical resection with adjuvant radiotherapy are the mainstays of treatment. Although our case did not have any recurrence disease for 2 years, a dose long-term follow up is needed, as 100% solid form of ACC could recur.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Adenoid cystic carcinoma (ACC) is a rare malignant tumour mainly affecting major and minor salivary glands .
- Long term survival can be achieved by surgery and radiotherapy .
- Perineural invasion is responsible for high local recurrence .
- Prognosis is dependant on solid histological type , perineural spread , distant metastasis and locoregional recurrences .

Managing isolated left internal iliac artery mycotic aneurysm

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Keywords: Isolated internal iliac artery aneurysm; salmonella infection; mycotic internal iliac artery aneurysm

Introduction

This incidence of isolated internal iliac artery aneurysm (IIIAA) is around 0.04% of all aortoiliac aneurysmal disease [1]. The conditions are extremely rare and the detection of internal iliac artery aneurysm (IIAA) usually coincidental when investigating other pathology. It has been reported that 40% of all cases present with aneurysmal rupture [2]. Most of the symptoms that have been reported associated with IIAA include abdominal pain (31.7%), urological (28.3%), neurological deficit (18.3%), groin pain (11.7%), hip or buttock discomfort and gastrointestinal symptoms (8.3% respectively) [1, 2].

We report a case of isolated mycotic IIAA and the management of the case will be discussed.

Case Presentation

A 63-year-old man with underlying diabetes mellitus and hypertension, presented with a 1-week history of left iliac fossa pain associated with high-grade fever. Investigated for possible diverticulitis, a Computerized Tomography Angiogram (CTA) was done and noted left saccular IIAA. It also showed a ruptured atherosclerotic plaque with a surrounding fluid collection and multiple air pockets (Figure-1). His blood culture revealed Salmonella species, which was sensitive to Ceftriaxone. His Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP) level were elevated on presentation with the value of 120 and 450 respectively.

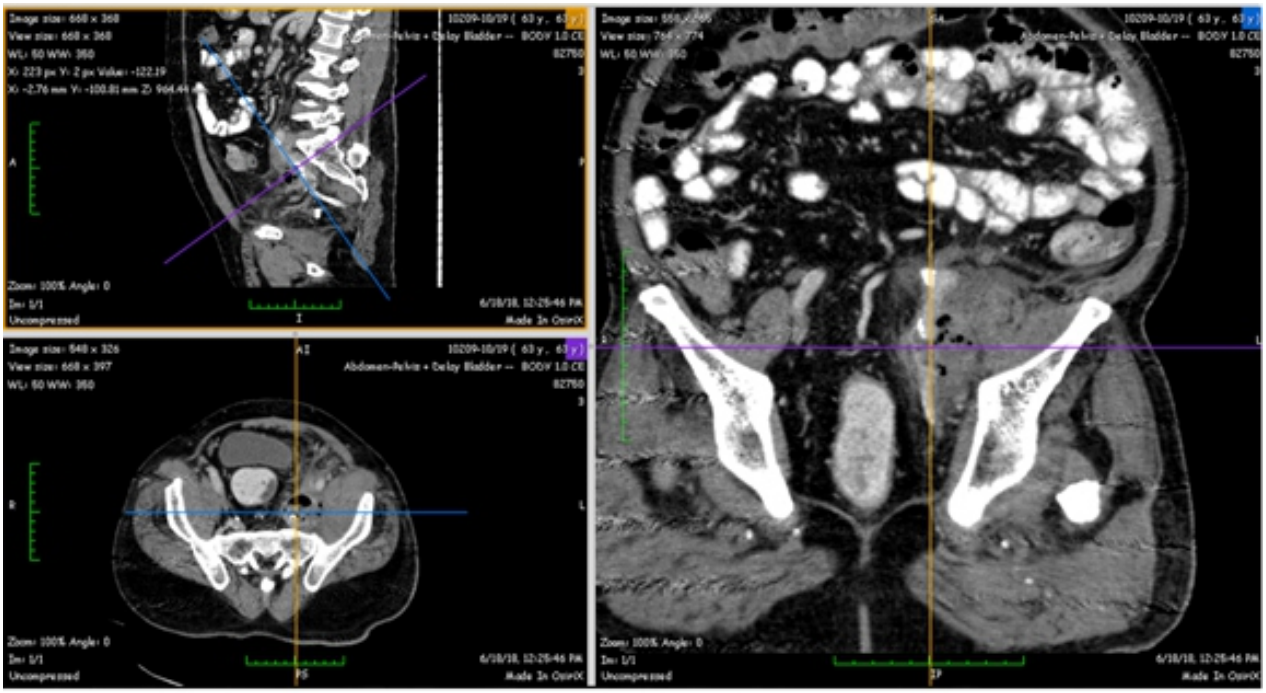

Figure 1. Noted the air pockets (thick arrow) surrounding the left internal iliac artery aneurysm with ruptured atherosclerotic plaque (thin arrow)

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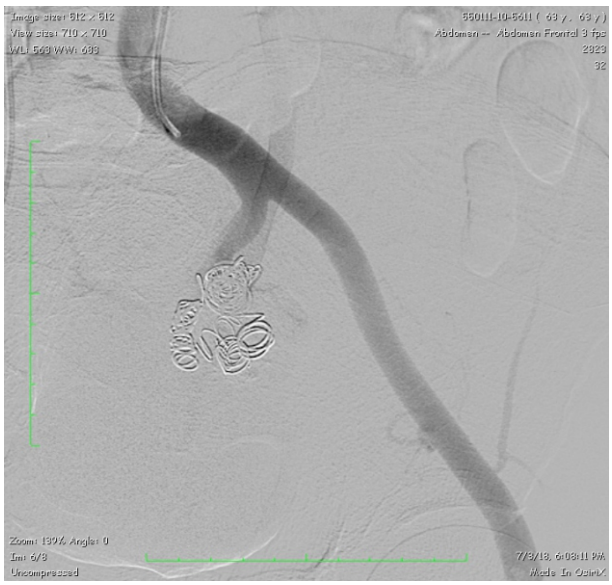

Figure 2. Successful coiling of the aneurysm and the left internal iliac artery

Based on the above, a diagnosis of isolated mycotic left IIAA was made. An intravenous ceftriaxone 2g daily dose was initiated via peripherally inserted central venous catheter (PICC) line in his right basilic vein. The patient responded well with the antibiotics and latest blood culture didn't grow any new organism after 2 weeks of antibiotic. Consultation with the interventional radiologist was made to embolize the outflow and inflow of the left IIAA and the saccular aneurysmal sac after 1 week of antibiotic (figure 2). The contralateral femoral artery was cannulated retrogradely and crossover to the left side. Amplatz vascular coils were used for the embolization.

A repeat CTA of the vessel was carried out later after 2 weeks post-embolization revealed similar collection remained with total occlusion of an aneurysm and left internal iliac artery. The left ureter was stented and open surgical debridement was carried out soon after, together with the long-term plan for IV antibiotics administration for a total of 3 months.

Discussion

Aneurysms of the iliac arteries are relatively rare. Nonetheless, their successful management demands a knowledge of their natural history. The majority of these aneurysms are atherosclerotic in type though occasional mycotic types do occur. The mycotic type of aneurysms represent only 2.5% of all aneurysms. [1, 3]. In this case, the ruptured plaque of the IIA may somehow be infected with a blood-borne organism causing a mycotic aneurysmal configuration.

The most common cause for mycotic type is Salmonella, accounting for 18% to 50% of cases [1]. Although a significant decrease in its incidence has been observed since 1965, it is still associated with 10% of all infected type of an aneurysm that has been found in the bacteriology cultures such as in this case [1]. Most infection occurs through oral route and cultures of stool or blood are frequently positive in such cases presented without gastrointestinal symptoms and is helpful for the selection of appropriate antibiotics. Inflammatory markers such as white blood count and C-reactive protein will be raised and series of these result may be used to observe progress following treatment.

Their onset is insidious and their occult location in the pelvis precludes the early diagnosis. Most of these cases have a pre-existing atherosclerotic disease at the site of an aneurysm that subsequently becomes infected, although in isolated cases Salmonella has been reported to invade even healthy intima [1]. The risk factors for the development of a mycotic aneurysm, in this case, includes local pathology of the ruptured atherosclerotic plaque of the left internal iliac artery and systemic cause from his underlying diabetes mellitus.

Salmonella infections can be divided into five categories: gastroenteritis, enteric fever, bacteraemia, localized infections, and chronic carrier state [1]. Vascular infections, osteomyelitis and meningitis are the most common non-gastrointestinal tract localized Salmonella infection. Salmonella vascular infections based on frequency, involve the aorta, coronary and peripheral arteries, prosthetic valves and vascular grafts.

The gold standard management of repair for IIAA is by open interposition graft [2], with the primary indication for repair being to prevent rupture in an aneurysm more than 4 cm in diameter [3, 4]. Other indication for repair includes abdominal pain and severe back pain. [3]. whilst the prognosis of the asymptomatic IIAA can be promising, the symptomatic patient such as retroperitoneal rupture or rupture into adjacent organ carries a limited prognosis [3, 4].

Endovascular techniques have excellent early outcomes when treating aortoiliac aneurysmal disease since the bifurcated end grafts were developed in the mid-1990s [4]. However, anatomical locations of IIAA deep in the pelvis make an endovascular approach to this entity quite challenging.

An infected IIAA may rapidly progress to rupture and prompt open surgical intervention is mandatory and life-saving although the procedure can be complicated since the IIAA is situated deep in the pelvis. Endovascular therapy such as embolization of the IIAA may alter the prognosis by controlling the acute bleeding and overall improve the survival rate in ruptured cases [4].

Conclusion

In conclusion, isolated mycotic aneurysm of the internal iliac artery is an extremely rare but serious condition of which vascular surgeons need to be aware. Their clinical manifestations are nonspecific. Being left untreated, it may

rupture from continued expansion and carries high mortality rate. The high index of suspicion and availability of radiologic modalities such as CT scan and angiography makes early diagnosis possible and hence early intervention can be initiated to improve survival rate in IIAA.

Finally, a successful management of this rare entity depends on collaboration from the infectious disease physician, interventional radiologist and the vascular surgeon. Observation of the clinical parameters of the patient is mandatory to ensure successful planned intervention of this rare yet serious disease. Early detection and diagnosis is the key point for early treatment initiation

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Successful management of a rare isolated IIA mycotic aneurysm depends on collaboration from the infectious disease physician, interventional radiologist and the vascular surgeon.
- Careful observation of clinical parameters of the patient is mandatory to ensure successful planned intervention of this rare yet serious disease.
- Early detection by means of high clinical suspicious index of the disease is mandatory for early treatment initiation.

Eviscerated bowel through the anus: managed by primary repair of rectosigmoid perforation in a young male

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Keywords: Eviscerated bowel; spontaneous rectosigmoid perforation; rectal prolapse; primary repair; surgical emergency

Introduction

Bowel evisceration through the anus is an uncommon condition to see and represents a true surgical emergency. Appropriate and timely surgical intervention can yield good postoperative results. Management begins at the point of first medical contact and should be individualized depending on the hemodynamic status of the patient and intraoperative findings.

Case Presentation

A 40-year-old male presented to the surgical emergency with his bowel loops protruding through the anus for the past 8 hours. On arrival, the patient had tachycardia but was normotensive. He gave a history of lower abdominal pain with protrusion of a mass through the anus during straining at defecation. Past history revealed grade 2 rectal prolapse for the past 2 years for which he has not sought medical advice. Abdominal examination revealed lower abdominal tenderness and per rectal examination showed approximately 4 feet of small bowel protruding through the anus. Bowel loops had a dusky discolouration but no signs of overt gangrene.

The patient was started on intravenous fluids and oxygen by face mask. The prolapsed bowel loops were wrapped in a sterile towel moistened with warm saline and patient was rushed to the operation theatre. A lower midline laparotomy was performed and approximately 4 feet of small bowel was seen herniating through a 4 cm tear in the rectosigmoid area (figure 1). The bowel was gently reduced into the peritoneal cavity. After adequate oxygenation and wrapping of the bowel loops with warm and moist surgical towels, examination revealed a viable small intestine. Since the margins of the rectosigmoid perforation were healthy, the patient was hemodynamically stable and there was no visible peritoneal contamination, the perforation was repaired primarily (figure 2). A 28 French drain was put in the pelvis and abdomen was closed in layers.

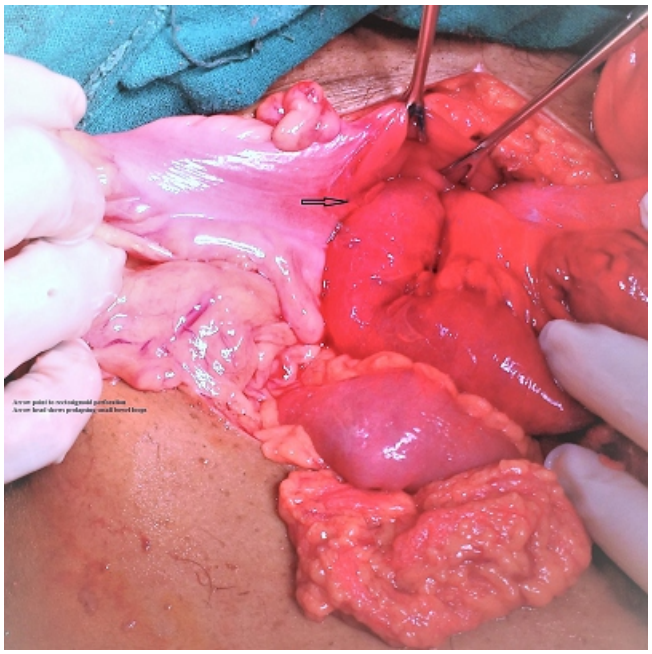

Figure 1. Small bowel herniating through the rectosigmoid perforation

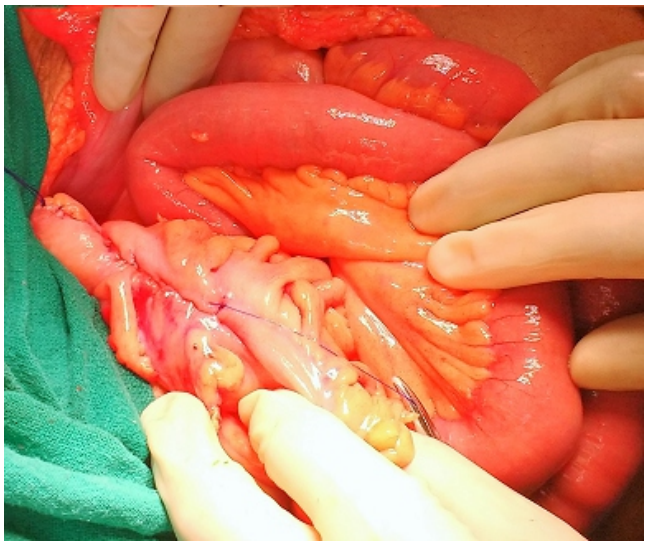

Figure 2. After primary repair of the rectosigmoid perforation

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Postoperative Course

Postoperatively the patient was extubated and was shifted to the surgical ward. He had an uneventful recovery. He was started on liquids orally on a postoperative day (POD) 4 and was started on a soft diet on POD 5. The drain was removed on POD 6. The patient was subsequently discharged on POD 8 with the postoperative advice on stool softeners and pelvic floor muscle training exercises.

At 6 months of follow up, the patient is doing well and is planned of definitive rectal prolapse surgery for grade 2 rectal prolapse.

Discussion

Ever since its first description by Brodie1 in 1827, bowel loops protruding through the anus continues to be a rare but important surgical emergency. Most of these cases give an underlying history of chronic rectal prolapse and increased intra-abdominal pressure is often the precipitating event.2-3 early operative intervention is essential to prevent bowel ischemia and gangrene.4 Patients may present late if coming from remote areas with inadequate medical facilities and also due to delay in transit. It is important to wrap the eviscerated bowel loops in a sterile towel moistened with warm normal saline at the point of first medical contact.

It is a surgical emergency and does not warrant any imaging. Endoscopy can be performed in the follow-up period, to evaluate for any predisposing condition for the perforation. After an adequate initial resuscitation patient should be immediately taken to the operation theatre. The decision to do a colostomy or a primary repair is based on the degree of peritoneal contamination, hemodynamic and performance status of the patient and margins of the perforation. A protective diverting ileostomy can be avoided if the conditions are favourable for a healthy anastomosis, but it remains a matter of surgeon's preference and experience.

Conclusion

Bowel loops through the anus, though rare represents a true surgical emergency. We report a case of a 40-year-old gentleman who presented to the surgical emergency with a grotesque picture of bowel loops through the anus. Exploratory laparotomy revealed a perforation in the rectosigmoid region with small bowel prolapsing through it. Bowel loops were reposited back and tear was repaired primarily. We highlight the importance of timely and appropriate surgical intervention which saved a major morbidity and yielded uneventful recovery.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- Spontaneous rectal perforation with eviscerated small bowel loops represents a true surgical emergency.
- Though most of the case occurs in elderly individuals with a history of chronic rectal prolapse, young individuals with a short history of rectal prolapse can also be affected.
- It is important to wrap the eviscerated bowel loops in a sterile towel moistened with warm normal saline at the point of first medical contact.
- Early operative intervention is essential to prevent bowel ischemia and gangrene.
- The decision to do a colostomy or a primary repair is based on the degree of peritoneal contamination, hemodynamic and performance status of the patient and margins of the perforation.

A floating first rib causing vascular thoracic outlet syndrome

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Keywords: Thoracic outlet syndrome; subclavian stenosis; arterial insufficiency

Introduction

Anomalous first rib is a rare cause of thoracic outlet syndrome (TOS) [1]. The incidence is 0.34% with equal gender distribution [1]. We present a case of a floating first rib causing dynamic obstruction of subclavian artery and vein.

Case Report

A 19-year-old male presented with insidious onset left arm and hand pain for three months. Pain aggravated during hyper-abduction and excessive manoeuvring of the limb. He denied of any past trauma or paraesthesia. Examination revealed fullness, thrill and a bruit over the left supra-clavicular region. Pitting oedema was present with no embolic phenomena or neurological manifestations. Radial and brachial pulses disappeared with Adson's test. X-rays of his chest and cervical spine were initially reported to be normal. Triplex Doppler scan of the left upper limb vessels was normal. There was segmental narrowing in the left subclavian artery at the costoclavicular space during abduction manoeuvre in Computed Tomography angiogram (Figure 1).

The luminal diameter of the subclavian artery in adducted and abducted positions was 8.5 mm and 1.5 mm respectively with a reduction of 46%. The subclavian vein was compressed with no visible flow during the abducted position. Rest of the subclavian vessels had no filling defects. The first rib on the left was short and ended halfway close to the mid-clavicle. It failed to articulate with the manubrium sterni. The left middle scalene muscle appeared larger in anteroposterior diameter than the right side. Retrospective examination revealed a short rib on the chest X-ray (Figure 2). The patient was explored using a supraclavicular approach. Intra-operatively compression of subclavian vessels by tight bands of scalenous anterior and medius was observed. The first rib was resected lateral to the sympathetic trunk with scalenectomy. Postoperative period was unremarkable.

Discussion

TOS occurs due to a cervical rib, anomalous first rib and ligamentous or fibrous bands of scalene muscles [2]. An anomalous first rib arises from the transverse process of the first thoracic vertebra. It is attached to the superior surface of the second rib with osseous or fibroligamentous connections [1]. If not, the anterior end of the first rib may remain free,

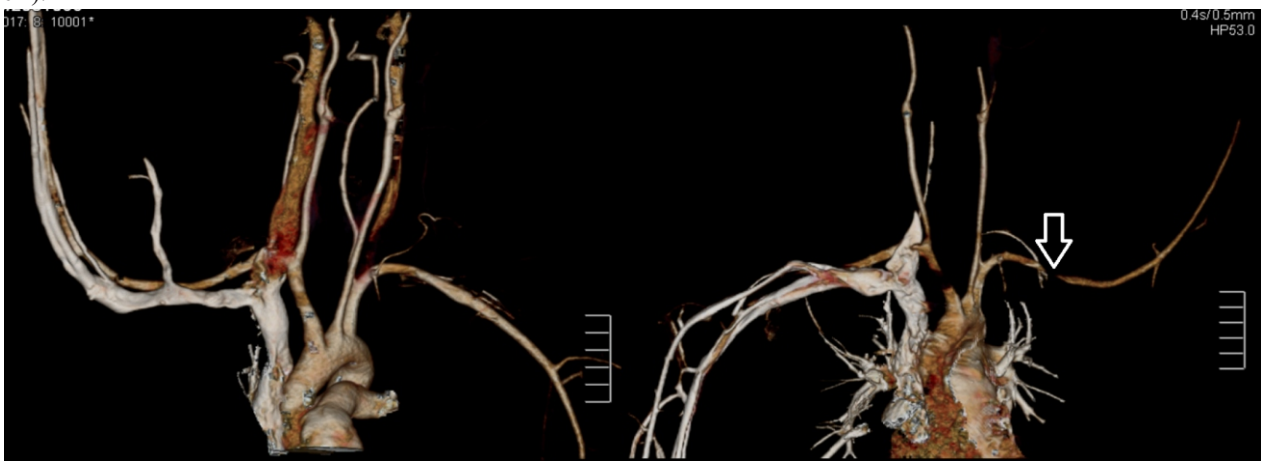

Figure 1. Computed Tomography angiogram showing segmental narrowing in the left subclavian artery at the costoclavicular space during abduction manoeuvre (marked with an arrow)

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DOI: http://doi.org/10.4038/sljs.v36i3.8535

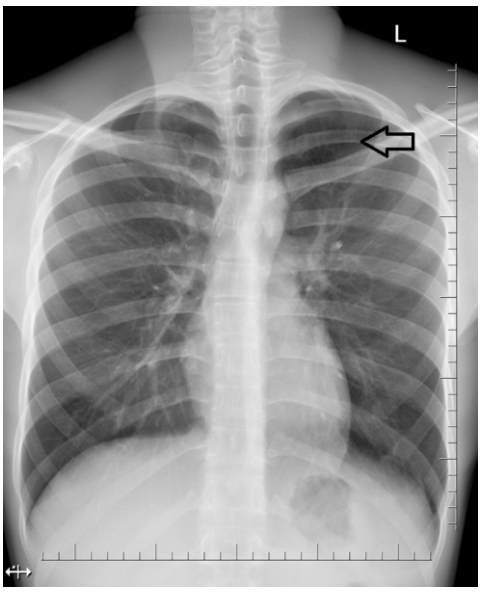

Figure 2. Chest X-ray showing a short rib on the left side. The anterior end is marked with an arrow.

termed a "floating first rib". Subclavian artery and trunks of brachial plexus run between anterior and middle scalene muscles. Subclavian vein arches over the first rib anterior to scalenous anterior. During forced inspiration or lateral flexion of the neck to the opposite side, traction of the scalenous muscles causes a floating rib to move upwards and compress these neurovascular structures. Similarly, when the ipsilateral upper limb is hyper-abducted, the available space within the posterior triangle is reduced.

Pete described the TOS for the first time in 1956[3]. In 80% of the times, TOS is caused by trauma [1]. TOS can be neurogenic, arterial, and venous or mixed, the commonest being neurogenic [2]. Arterial TOS can be due to dynamic obstruction, fixed stenosis or post-stenotic aneurysm of the subclavian artery [1]. Dynamic obstruction presents as pain, paraesthesia and Raynaud's phenomenon during hyper-abduction. The examination may reveal a thrill and a bruit in the supraclavicular region over the stenotic subclavian artery. Adson described the disappearance of the radial pulse on inspiration and turning of the neck towards the ipsilateral upper limb in arterial TOS in 1927 [4].

Arterial TOS can also be asymptomatic until thrombosis or embolism occurs. The latter might present with the cardinal features of acute arterial insufficiency. Venous TOS presents with transient oedema, pain, pallor markedly on hyper-abduction and prolonged working. If deep venous thrombosis occurs, rapidly progressing and persistent oedema accompanied by the severe throbbing type of pain will be noted [5]. Our patient had both components of arterial and venous insufficiency.

Unlike cervical ribs, anomalous first ribs are difficult to diagnoses in chest radiographs, because they arise from the transverse processes of first thoracic vertebrae [1]. It was missed initially in our case. Careful comparisons of both hemithoraces may reveal a thin, superiorly placed rib which does not articulate with the manubrium [1]. Doppler ultrasound and angiogram with provocative manoeuvres are sensitive in diagnosing both arterial and venous TOS [6]. The normal triplex study in our case may be due to the avoidance of provocative manoeuvres during imaging.

Vascular TOS can be managed by surgical excision of the anomalous rib. Murphy conducted the first successful surgical excision of an anomalous first rib [7]. DeBakey incorporated resection of anterior scalene muscle with improved surgical outcomes [8]. Supraclavicular and trans-axillary approaches are described for excision of anomalous first ribs [1]. We used the supraclavicular approach in our patient as it provided a better exposure of the subclavian artery and to excise the anomalous rib.

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- The anomalous first rib is a rare cause of vascular thoracic outlet syndrome
- Presence of thrills, bruits and positive Adson's test are indicative of arterial thoracic outlet syndrome
- Imaging studies such as Doppler ultrasound and angiograms need to be done with provocative manoeuvres
- Excision of the anomalous first rib with scalenotomy is the treatment of choice

Inflammatory fibroid polyp (Vanek's tumour) of the ileum causing ileocolic intussusception

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Keywords: Inflammatory fibroid polyp; adult intussusception; ileocolic intussusception; Vanek's tumour, PDGFR α mutation

Introduction

Inflammatory fibroid polyp (IFP) is a rare reactive, benign gastrointestinal tract lesion, affecting, mainly elderly population. Its clinical, endoscopic and radiological pictures may mimic that of a malignancy and can give rise to a diagnostic dilemma. Patients with IFP may present with intussusception, intestinal obstruction or haematochezia. Treatment options include surgery and in limited cases endoscopic resection.

Case Report

A 55-year-old Sri Lankan Tamil female presented to the surgical casualty ward with a history of acute onset abdominal pain, constipation and abdominal distention for 2 days. Her past medical and surgical histories were unremarkable. She did not have a significant family history of gastrointestinal tract malignancies. On examination, she was pale, dehydrated, and her abdomen was tender and distended with no palpable masses. Per rectal examination was unremarkable. Initial lab works were unremarkable except a mild anaemia (11.2 g/dL). Contrast-enhanced CT of the abdomen revealed an ileocolic intussusception but did not show the leading cause. Her colonoscopy revealed a pedunculated growth at the hepatic flexure region, and biopsies were taken. Biopsies showed only inflammatory changes without any specific diagnosis. Subsequently, she underwent an exploratory laparotomy and found to have a growth at the ileal region causing an ileocolic intussusception. The lesion with a segment of bowel was resected en bloc followed by an end to end primary anastomosis. Gross pathological examination showed a 40mm size, grey, firm, pedunculated, lesion arising from the submucosal layer with a homogeneous pale white cut surface. There were no haemorrhagic or necrotic areas. Rest of the ileum and colon were macroscopically unremarkable. Microscopic examinations revealed a well-circumscribed lesion with numerous dilated thin walled vessels and spindle cells in an oedematous stroma without any malignant features. She recovered from the surgery without complications and was discharged in three days.

Discussion

Inflammatory fibroid poly (IFP) is a rare gastrointestinal tract reactive lesion. The first case was reported by Vanek in 1949, but the name was proposed by Helwig and Ranier in 1953. IFP has been traditionally described as a non-neoplastic lesion, but recent molecular studies show activating mutations of platelet-derived growth factor receptor α (PDGFR α) in 70% of the cases, suggesting a possible neoplastic pathology [1]. IFP affects all the age groups without a sex preference. Majority of cases have been reported in the elderly population with a peak incidence in fifth to seventh decades [2].

Approximately 90% of cases of intussusception in adults are secondary to a pathologic condition that serves as a lead point for the intussusception, such as carcinomas, Meckel's diverticulum or rarely IFP. Patients with IFP may also present with intestinal obstruction and haematochezia. IFP patients presenting with lower urinary tract symptoms are also reported. IFPs can arise from any part of the gastrointestinal tract, but gastric antrum is the commonest site, accounting for 70% of the cases followed by small intestine in 20%. It rarely affects the oesophagus and colorectum [3]. There are cases of IFP affecting gallbladder reported as well [1]. Endoscopically approximately 70% of the IFPs appear semi-pedunculated and the rest are sessile, arising from the submucosa covered by mucosa. Occasionally the polyp can invade the mucosa or extend into the muscularis propria. Most of the IFPs measure less than 5cm in diameter, although IFPs up to 20cm are reported [4].

Contrast-enhanced CT is sensitive in diagnosing intussusception, showing an outer intussuscipiens and inner intussusceptum, giving a sausage-shaped mass appearance in its longitudinal axis and as a target lesion in cross section. Even though CT is sensitive in diagnosing the intussusception, it fails to identify the cause as IFP in most of the

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instances [4].

The reported incidence of malignancy in ileocolic and colo-colic intussusception ranges between 43 and 100%. Since the preoperative investigations fail to identify the cause in a majority of the cases, surgical intervention is needed as diagnosis and treatment [4]. There is an argument whether to proceed with an en bloc resection or a more limited resection after reduction. Reduction before resection may allow a more limited resection; but manipulation itself increases the risk of intraluminal seeding or tumour dissemination of a malignant lesion [5]. In cases of small intestinal intussusception, malignancy found to be the cause in up to 47%, and most lesions are metastatic [4]. Therefore, it is recommended to reduce the intussusception before a limited resection [5[. However, in ileocolic and colo-colic intussusception the possibility of a malignancy ranges from 43-100% with the majority of the lesions being primary rather than metastatic. Therefore, it is recommended to resect the lesion en bloc without reduction [4,5]. It can be approached by open method via a midline laparotomy as in this case report. Laparoscopy has been shown to be an equally effective alternative with manual reduction done after extra corporealization [2].

Even though endoscopic resection is a justifiable treatment approach due to the benign nature of IFP, use of therapeutic endoscopy for IFP is rare due to the size of the lesion, failure of preoperative confirmation of the diagnosis, and the urgency of symptoms [3].

Microscopically the IFP comprises of numerous small vessels, fibroblasts in an oedematous stroma, infiltrated by inflammatory cells mainly eosinophils and plasma cells [3]. The fibroblasts are arranged in a concentric “onion-skin” pattern centered on blood vessels giving rise to frequently seen whorled appearance. In immunohistochemical stains, IFPs express positivity to CD34, S100, Fascin and CD35 and negativity to CD117, Bcl-2 and c-kit. Positivity to CD35 and Fascin has been presented as evidence of dendritic cell origin. Negativity to CD117, Bcl-2 and c-kit differentiate IFPs from Gastrointestinal Stromal Tumours (GIST) [3].

All authors disclose no conflict of interest. The study was conducted in accordance with the ethical standards of the relevant institutional or national ethics committee and the Helsinki Declaration of 1975, as revised in 2000.

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Learning Points:

- IFP is one of the rare causes of adult intussusception, which may mimic malignancy.
- Preoperative diagnosis is difficult, and hence warrants surgical intervention for both diagnosis and treatment.
- Colonic intussusception, in general, should not be manually reduced to reduce the potential risk of malignant venous dissemination.
- Endoscopy plays a limited role in terms of definitive management.

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