DISCUSSION

Congenital bladder diverticulum with out associated posterior urethral valve or a neurogenic bladder is rare. They usually occur lateral and cephalad to ureteric orifice. These types of diverticula are larger than those associated with secondary causes. The cause of these diverticula is an inherent weakness in the bladder musculature. Vesico-cutaneous fistula due to other etiological factors has been reported frequently. Common causes include extensive trauma with pelvic fractures,[1] after irradiation for pelvic malignancies,[2] postoperative causes like radical hysterectomy,[3] hip arthroplasty.[4] There are also few cases reported as sequel to large bladder calculus.[5] Anecdotal cases of vesico-cutaneous fistula from inguinoscrotal hernia,[6] antenatal bladder aspiration,[7] bladder instability,[8] factitious,[9] actinomycosis[10] have been also reported. A thorough search for the etiological factors like stones and malignancy should be made. IVU, VCU, and a cystoscopy would be useful in making the diagnosis. Other cross-sectional imaging such as CT scan and MRI is needed if the fistulous tract is complicated and malignancy cannot be ruled out with routine imaging modalities. The threat of repeated urinary tract infection and malignancy make the management of this lesion mandatory. Open surgical management with excision of the fistulous tract and interposition with myocutaneous flap is ideal for large fistulas. Extensive skin loss can be replaced by skin grafting. After a thorough search of literature we could not find any reported case of similar nature.

REFERENCES


CASE REPORT

IDIOPATHIC MEDIASTINAL FIBROSIS PRESENTING AS MEDIASTINAL COMPRESSION SYNDROME

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ABSTRACT

Mediastinal compression syndrome is a commonly seen entity. Mediastinal compression, mostly due to a space-occupying lesion, is distinct and different from mediastinitis/mediastinal fibrosis, which could also lead to superior vena cava syndrome. Idiopathic mediastinal fibrosis should also be considered as differential diagnosis of mediastinal structures with various radiological, CT and MRI and histological features if feasible. Medical therapy is disappointing while surgical cure has limitations. This interesting patient presented as mediastinal compression syndrome, which on investigation was postulated as idiopathic mediastinal fibrosis, as a diagnosis on exclusion of other causes, which is rare, hence being reported.

KEY WORDS: Fibrosis, mediastinitis, mediastinal fibrosis

INTRODUCTION

Idiopathic mediastinal fibrosis is also known as Chronic Fibrosis or Fibrosing Mediastinitis or Chronic Mediastinal Fibrosis or Cryptogenic Mediastinal Fibrosis, all terms suggestive of slowly progressive envelopment of mediastinal structures in proliferating fibrous scar tissue. Osler in 1903 published the first major review on mediastinal fibrosis,[1] since then a few case series[2][3] have been published with stray case reports.[4][5][6] Mediastinal structures are surrounded, constricted and sometimes invaded by the fibrous tissue, which may extend to affect other intrathoracic organs. The pattern of involvement of mediastinum is variable and so are the clinical features.[2] Venous structures (Superior Vena Cava and Pulmonary veins), due to thin walls and low intraluminal pressure, tend to be compressed earlier than are the arteries and tracheobronchial tree and esophagus. For this reason venous hypertension in the drainage area of superior vena cava is considered the hallmark of the disease. It is a rare cause of superior vena cava obstruction (1–2%).[4] Various hypotheses have been proposed for the fibrous lesions.[3]

Radiological,[6] CT and MRI features help in diagnosis however histological confirmation is

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difficult. Medical therapy is discouraging, surgery has limitations while stenting of vessels and dilatation of bronchi and esophagus may provide some relief to patients.

CASE HISTORY

A 30-year-old male, rickshaw-puller by occupation presented with puffiness of face and swelling over both upper limbs, more after getting up in the morning since 2 years. He also had noticed dyspnoea on exertion and dull aching diffuse chest pain since 2 years and had cough with scanty mucoid-white expectoration since 4 months. There was no history of fever, hemoptysis, edema feet, urinary complaints, dysphagia, nausea or vomiting, change in voice, palpitation. Past history was not contributory. There was no history of drug intake or exposure to radiation. Personal and family history was not contributory.

This average built man had suffused conjunctiva, face and both upper limbs. The vital parameters were normal. The jugular veins were full and nonpulsatile, prominent veins were seen over arms, chest and anterior aspect of neck with flow away from mediastinum. No edema feet. Respiratory system examination revealed bilateral ronchi and crepitations. Examination of other systems was normal.

On investigation his hemogram, urine analysis, renal and liver function tests, ECG and 2–D-ECHO were normal. Sputum for acid-fast bacilli, malignant cells, and fungus culture was negative. The X-ray chest showed mild superior mediastinal widening. Computerized tomography of chest showed multiple abnormal vessels in superior mediastinum. Abnormally dilated azygous vein and small vascular structure in the region of SVC. There was no evidence of mediastinal mass or lymphadenopathy. Venography of upper extremity revealed bilateral collaterals. Superior Vena Cava was not visualized. Mediastinoscopy, mediastinotomy or bronchoscopy may show areas of compression or distortion, which may be seen on computerized tomography of chest.

Investigations suggestive of idiopathic mediastinal fibrosis are mediastinal widening on X-ray chest, barium swallow and bronchoscopy may show areas of compression or distortion, which may be seen on computerized tomography of chest. Venogram showing collaterals and mediastinal veins are diagnostic. Mediastinoscopy, mediastinotomy may be difficult and histopathology may be confirmative and requires large sample of tissue if obtained surgically though rarely CT guided automated needle biopsy may be possible.

Our patient presented with features of superior vena cava obstruction for which no clinical or radiological cause could be demonstrated, hence a possibility of idiopathic mediastinal fibrosis was entertained as a hypothesis is discarded due to lack of histological corroboritation. The first description of idiopathic mediastinal fibrosis is ascribed to John Hunter in 1757 while others prefer the description written by Hallet of Edinburgh in 1948. The pathogenesis as reflected by the term ‘Idiopathic’ is unknown. The various hypotheses postulated for the fibrous lesions are as follows:

1. Fibrosis results from chronic infections, e.g. tuberculosis, histoplasmosis, and syphilis.
2. Repeated trauma followed by organization in a hematoma produces fibrosis, however history of trauma every time may be lacking.
3. Inflammatory process in adipose tissue as seen in Weber Christian disease – this hypothesis is discarded due to lack of histological corroboration.
4. Iatrogenic due to adverse reaction to drugs, e.g. methysergide, practolol. Most cases have no consistent history and withdrawal of drugs does not arrest or regress the fibrosis.
5. Sclerosing malignancy is an unlikely explanation in the absence of residual tumor tissue.
6. Hypersensitivity or autoimmunity correlates with the histopathological features.

Idiopathic mediastinal fibrosis is characterized by bundles of intertwining bands of hypocellular, collagenous, fibrous tissue often with whorled pattern containing an infiltrate of predominantly plasma cells with some lymphocytes, polymorphs and fibroblasts. Medical therapy with diuretics, Anticoagulants,
Steroids and penicillamine has been disappointing. Palliative surgery for SVC obstruction in form of autogenous Saphenous vein graft, woven silicon rubber prosthesis and bovine pericardial conduits for venous reconstruction are recommended along with dilatation of bronchial and esophageal strictures.[7] Stenting of vessels, bronchi and esophagus can be of some help.[5]

The immediate and ultimate risk to life is small. But the inconvenience is considerable and sustained. As the time passes the development of venous collateral circulation mollifies some of the distress, but the clinical state never returns to normal unless the mechanical obstruction can be overcome.

REFERENCES

LETTER TO EDITOR

TUBERCULOSIS OF URINARY BLADDER PRESENTING AS PSEDOURETEROCELE

Sir,
A 35-year-old man presented with recurrent episodes of hematuria, increased frequency of urination and occasional mild dysuria of 1 year duration. There was no fever, weight loss, or loss of appetite. Ultrasound examination showed mild dilatation of intramural portion of the left terminal ureter projecting into the lumen of the bladder (Fig. 1). The wall of the dilated intramural ureter was irregular with few internal echos. There was no change in size of the lesion on real time scanning. No obvious sonological abnormality was seen in the kidneys. Plain radiographs of chest and abdomen were normal. Urine microscopy showed plenty of white blood cells and 10–15 red blood cells. No organism was grown in the routine cultures. Other investigations, including haemoglobin, erythrocyte sedimentation rate, leucocyte count, blood sugar, and serum creatinine, showed normal results. On cystoscopy, bladder wall was erythematos and edematous with involvement of left ureteric orifice. Inflammatory exudate was seen at ureterovesical junction. Biopsy from the bladder wall adjacent to the left ureteric orifice revealed chronic granulomatous inflammation consistent with tuberculosis (Figure 2). He was given antituberculous therapy. Follow-up ultrasound examination done after 6 months showed resolution of the pseudoureterocele and the patient was asymptomatic.

Ureteroceles are obstructive cystic dilatations of the intravesical or intramural portion of the ureter that result in ballooning of the distal ureter into the bladder.[1] Ureteroceles were one of the common incidental observations at sonography on asymptomatic patients. On sonography, they appear as a well-defined...