CASE REPORT

Pancreatitis Leading to Retroperitoneal Fibrosis and Ureteric Obstruction

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Summary

Two patients who had acute pancreatitis subsequently developed characteristic appearance on urography of smooth extrinsic narrowing and medial deviation of the right ureter suggestive of retroperitoneal fibrosis (RPF) resulting in ureteric obstruction. Both these patients had clinical, biochemical and sonographic evidence of acute pancreatitis. CT scan of the abdomen performed on the second patient also documented acute pancreatitis. Intravenous urograms were consistent with ureteric obstruction due to retroperitoneal fibrosis. Both cases were treated conservatively. They were well after an average of 20 months. These 2 cases illustrate the uncommon association between pancreatitis and RPF.

Key Words: Pancreatitis, Retroperitoneal fibrosis, Ureteric obstruction

Case History

Case 1

A 54 year old Asian man was admitted for gastroscopy to exclude peptic ulcer disease. He had occasional dyspepsia, but no other significant past medical history. Post-operatively, he developed sudden, severe epigastric pain which radiated to the upper back. Physical examination revealed a tender and guarded upper abdomen, with stable vital signs. The laboratory results confirmed the clinical suspicion of pancreatitis with a markedly raised serum amylase level of 4400 U/L and a urinary diastate of 22,000 U/L subsequently. He did not have other biochemical abnormalities such as hypocalcaemia, hypoxaemia or hyperglycaemia. He also had ultrasonographic evidence of pancreatitis which showed a grossly swollen pancreas of low echogenicity. Follow up ultrasound on the 15th day showed interval evolution of acute pancreatitis with peripancreatic fluid collection. The kidneys were sonographically normal. He was managed conservatively and was subsequently discharged from hospital after a month's stay.

He remained asymptomatic till 3 years when he complained of right loin pain. An intravenous urogram (IVU) revealed a short smooth stricture with medial tapering of the right ureter at the lower lumbar region and mild hydroenephrosis (Fig 1). A technetium 99m-labelled DTPA renogram study showed preserved and comparable renal function bilaterally. The right pelvicalyceal system,

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although dilated, was not obstructed. This dilated right pelvicalyceal system was also documented on ultrasound. In view of the above findings, a right-sided retrograde pyelogram was carried out. This demonstrated a persistent 2cm smooth stricture with medial deviation of the right ureter at the level of L4 vertebra. This stricture did not cause significant obstruction and there was unimpeded retrograde passage of ureteric catheter. He was treated conservatively and remained asymptomatic on follow up at 18 months.

Case 2

A 76 year old Indian man and known alcoholic was admitted with complaints of a sudden onset of acute abdominal pain. He was previously seen by a family physician for episodic epigastric discomfort. Physical examination revealed tenderness in the upper abdomen. An electrocardiogram (ECG) performed showed T wave inversions in the lateral chest leads indicative of underlying ischaemic heart disease but there were no hyperacute changes to indicate an acute myocardial event. The other working diagnosis of an acute abdomen was confirmed when the biochemical results showed a significantly raised serum amylase of 4265 U/L and urinary diastase of 24,530 U/L indicative of acute pancreatitis.

Ultrasonographic confirmation was obtained on the third day of illness which revealed an enlarged pancreas with focal hypoechoic areas within. There were no gallstones and the intrahepatic ducts were not dilated. A computed tomography (CT) of the upper abdomen on the 6th day showed diffuse enlargement of the head and body of the pancreas, with stranding of the surrounding fat compatible with acute pancreatitis (Fig. 2). The kidneys were normal in appearance. He was discharged well. However, follow up ultrasound examination revealed moderate right hydronephrosis. No renal stones were visualised. An intravenous urography (IVU) revealed classic findings of a smooth stricture with medial deviation in the upper right ureter causing incomplete obstruction. He was treated conservatively, and was discharged well, after 2 years of follow up.

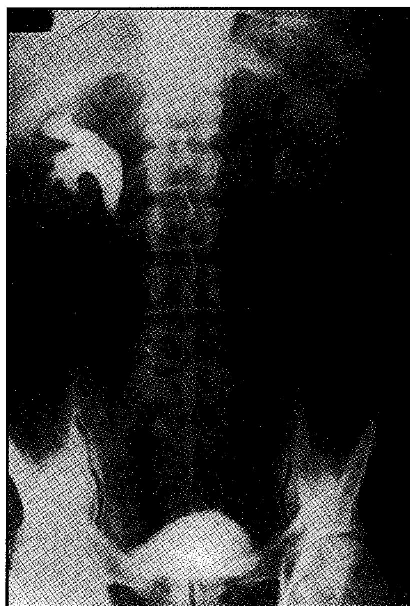

Fig. 1: Intravenous urogram shows smooth narrowing and medial deviation of the upper ureter with resultant moderate hydronephrosis.

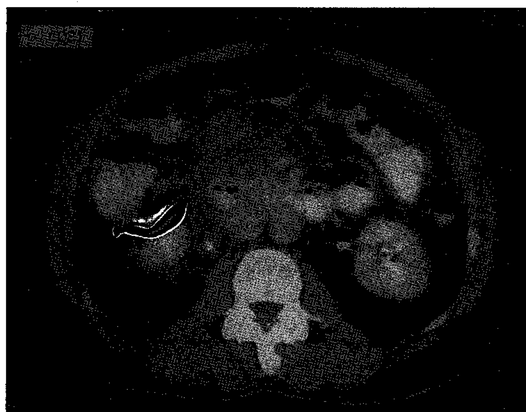

Fig. 2: CT scan shows pancreatic swelling, peripancreatic fluid and inflammatory stranding involving ureter resulting in peri-ureteric wall thickening.

Discussion

Aetiology

Retroperitoneal fibrosis (RPF) is idiopathic in two thirds of cases [1]. Several theories have been proposed as to the cause which included vasculitis alone or as a manifestation of a systemic collagen disease. Another theory suggested possible immunologic response of leakage of ceroid, an insoluble lipid from an aortic atheromatous plaque Bartholomew et al [2] in 1963 described idiopathic retroperitoneal fibrosis, multifocal idiopathic fibrosclerosis (MIF), as a system fibrosing disease involving other sites, manifesting as Riedel's thyroiditis, sclerosing cholangitis, mediastinal fibrosis, and occasionally orbital involvement.

The aetiolgoy in the remaining one third of cases are myriad and include chronic ingestion of methylsergide, an ergot derivative previously prescribed for migraine [3]. Other ergot derivatives implicated are lysergic acid diethylamide and bromocriptine. In addition, beta blockers, methyldopa, hydralazine and various analgesics and antibiotics are also associated with RPF.

Other conditions causally linked to RPF include specific infectious like tuberculosis, syphilis, actionomycosis, histoplasmosis and fungal infections. Inflammatory processes of the gastrointestinal tract such as diverticulitis and appendicitis have also been reported [4]. Pancreatitis as a possible aetiology has also been suggested.

As haemosiderin was detected in some fibrotic plaques, haematomas from both traumatic and spontaneous causes have been implicated. Surgical procedures especially anterior spinal fusion and aortic bypass surgery have been associated with RPF [4]. Yet another association is that of metastatic deposits inciting a desmophlastic response resulting in a fibrotic plaque. The malignancies reported include carcinomas of the breast, lung thyroid, gastrointestinal tract, and genitourinary organs as well as sarcomas and lymphomas. Extravasations of urine and aqueous contrast medium into the retroperitoneal space has also been described as possible associations.

Proposed pathophysiology in Pancreatitis

In pancreatitis, the pancreatic enzymes released cause destruction of the surrounding parenchyma resulting in autodigestion, fat necrosis, inflammation and haemmorhage. If the patient survives the acute insult, healing with focal or diffuse parenchymal fibrosis occurs. The ureters being in close proximity in the retroperitoneal space, then becomes entrapped with extension of this fibrotic process.

In the first case study, the patient developed classic appearance on urography of an "encased" right ureter with a short segment of ureteric narrowing and medial deviation. Also, the retrograde catherisation of the right ureter was performed with ease demonstrating a lack of ureteric obstruction. It has been propounded that RPF by encasement of the ureter inteferes with ureteral dynamics and peristalsis rather than causing a true mechanical obstruction. In addition, the still patent lumen in RPF serves to decompress the upper urinary tract and can cause insignificant caliectasis in some cases [5].

In the nest case study, the patient is a known alcoholic with acute pancreatitis. He also had evidence of underlying ischaemic heart disease. This patient had CT documentation of acute pancreatitis and also radiological evidence of retroperitoneal spread of inflammation to surround the right ureter. He subsequently developed a ureteric stricture one month after the acute presentation.

However, as both these cases were treated conservatively, there has not been any histological confirmation of the diagnosis, both were based on clinical and radiological evidence highly suggestive of retroperitoneal fibrosis.

Treatment

The treatment of retroperitoneal fibrosis ranges from conservative management, oral steroid therapy, ureteric stenting, to open surgery, biopsy with ureteric bypass. Treatment of the underlying cause is also important. Symptomatic treatment of the ureteric obstruction and relief of the obstruction uropathy is paramount.

In conclusion, the first patient develops classic features on both urography and retrograde examination, suggestive of retroperitoneal fibrosis three years after an attack of acute pancreatitis. The second patient, predisposed to alcoholic pancreatitis by his drinking habit, developed signs of extrinsic narrowing and medial deviation of the right ureter. In both of the, no other associated causes of RPF were elucidated. Although RPF and pancreatitis is not a new association, these reports serve to illustrate retroperitoneal and periureteric fibrosis as a potential sequelae of pancreatitis.

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