Ormond's disease – case report

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Retroperitonel fibrosis is a rare disease that occasionally faces the general surgeon as a case of retroperitoneal tumour. Ureteric stenting may succeed in relieving urinary obstruction, however, these stents are only for temporal use. For this reason and to exclude presence of retroperitoneal tumours, surgical ureterolysis remains the corner stone for saving the kidneys from the ultimate fate of renal failure. Steroids are of help when added to the surgical therapy. Recently tamoxifen was reported to be the treatment of choice in primary retroperitoneal fibrosis.

Here, we describe the management of 2 cases of idiopathic retroperitoneal fibrosis one in a police officer with diabetes mellitus and the other in a housewife.

Keywords: Bilateral hydronephrosis, retroperitoneal fibrosis, surgical ureterolysis, bilateral hydronephrosis, tamoxifen

Case 1

A 48-years-old police officer presented to our surgical gastroenterology department, at the National Centre of Gastrointestinal and Liver Diseases, with low back ache, persistent bilateral loin pain dull aching in nature, general ill -health, fatigue and loss of weight for six months. This illness disabled him from conducting his work.

On physical examination he was ill looking, not pale, jaundiced or cyanosed. Pulse 88/min, respiratory rate 20/min, afebrile, BP 100/70. Cirvical lymph nodes were not palpable. His chest and cardiovascular systems were clinically normal. Examination of his abdomen revealed vague discomfort in both hypochondria and at the hypogastrium. There was mild tenderness at the renal angles on gentle fist percussion, but his kidneys were not palpable and no other palpable abdominal masses. His femoral pulses were palpable, but rather feeble. Testis and scrotum were normal. Neither lower limb oedema nor varicose veins of lower limbs was detected.

Laboratory data

His Hb was 10.6 gm/dl; WBC 5.3 × 10^9 /L, ESR 112 in first hour. Fasting blood sugar 160/dl, blood urea 95mg/dl, serum creatinine 3.2 mg/dl, sodium 132 mmol/l, potassium 4.2 mmol/l. Glucose tolerance test showed a diabetic curve.

Chest X-ray and ECG were normal. Ultrasonic scan revealed bilateral hydronephrosis and a surapubic mass extending from the pelvis to the pancreas and seems to encase the aorta. CT scan documented a solid mass in the posterior abdominal wall anterior and at both sides of the bodies of the lumber vertebrae extending from the pancreas down to the pelvis surrounding the aorta and the inferior vena cava and their common iliac branches with bilateral hydronephrosis (Fig 1 and 2). There were no para-aortic lymph nodes or liver metastases. IVU was not done because of the high blood urea. Retroperitoneal fibrosis, fibrosarcoma and lymphoma were considered as differential diagnoses.

Hospital course

The patient was put on insulin and an informed consent was taken for surgery. During laparotomy dense fibrous tissue was found, in the retroperitoneal area, surrounding the aorta, inferior vena cava, their iliac branches and both ureters. Debulking of the vascular, tough fibrous tissue mass was done and the excised tissue was taken for histopathology. Ureters were freed from below the pelvic brim up to the pelvis of the kidney.

The walls of the ureters were noted to be very thick being infiltrated by the fibrous tissue. Ureters were then brought into the peritoneal cavity and covered with greater omentum. Common iliac vessels were freed partially from the fibrous tissue.

Fig1: Retroperitoneal mass involving the Aorta, IVC and ureters

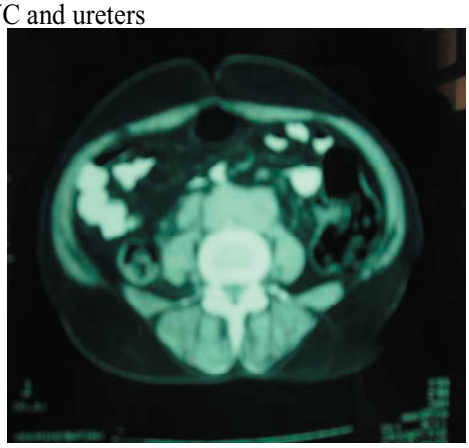

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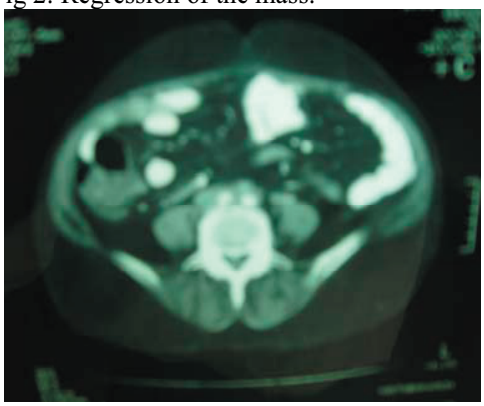
Fig 2: Regression of the mass.



Follow up

The post operative period was uneventful. Histopathology reported benign fibrous tissue, no malignant change was seen. The appearance is consistent with retroperitoneal fibrosis.

Because of his diabetes and the incomplete removal of the fibrous tissue mass the patient was put on prednisolone 60mg daily in divided doses tapered off after 2 months. He was followed up with serial ultrasonic scanning, renal function tests and fasting blood sugar levels. Four months after completion of his first course CT scan was ordered. This showed that hydronephrosis has disappeared, and the size of the mass regressed (Fig 3 and 4)

His blood urea and serum creatinine returned back to normal. He was put on another similar course of prednisolone for a couple of months. He is now followed up for 48 months and is in good condition with normal renal functions and controlled diabetes with 5mg Glibenclamide (Doanil, Wrexham-UK) daily.

Case 2

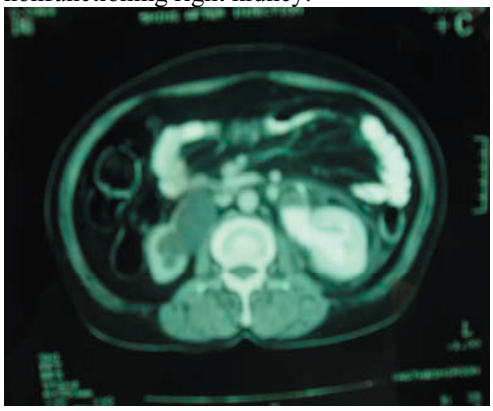
A 55–year-old lady presented with generalized aches and pains, fatigue and vague abdominal pain. On physical examination she was ill-looking, not pale, with normal vital signs and bilateral loin tenderness.

Investigations revealed Hb 10 gm/dl WBC $5.600X^{9}$/ml ESR 120mmfirst hour. Ultrasound reported retroperitoneal mass with bilateral hydronephrosis. CT scan with contrast showed retroperitoneal mass with bilateral hydronephrosis but the right kidney is not functioning (Fig 3).

Management:

She was put on combination of prednisolone 15 mg tid for three months and Tamoxifen 10 mg bid for 6 months.

Fig3: Bilateral hydonephrosis and right nonfunctioning right kidney.



Followup

She showed progressive improvement. However, she complained of feeling of vage generalized swelling. Follow up CT scan showed dramatic regression of the retroperitoneal mass and complete resolution of the hydronephrosis but the right kidney became smaller in size (Fig 4).

Fig 4 Regression of the retroperitoneal mass.

Discussion

Retroperitoneal fibrosis was first described by Ormond in 1948[1] but two earlier cases were reported in the French literature one by Albarran in 1905 and the other by Kolischer in 1922 as suggested by Ormond himself[2]. The definitive cause of the idiopathic retroperitoneal fibrosis is not known. However, lar g ebowel strictures[3], mediastinal fibrosis[4], orbital pseudo tumours[5], uveitis[6] and sclerosing cholangitis[7] were reported with idiopathic retroperitoneal fibrosis. None of these conditions are present in our cases however; In the first case retroperitoneal fibrosis was associated with recent onset of diabetes mellitus. Although the retroperitoneal fibrosis in our case is extending up to the pancreas, unfortunately we didn't take a biopsy from the tail of pancreas.

Therefore we are not sure whether his diabetes is due to infiltration of the pancreas by fibrosis or this association is a mater of chance only. Yet, factors known to play a definite roll in retroperitoneal fibrosis are drugs like methysergide[8], infections extending from diveticular disease of the colon or other pelvic organs, extravasation of urine, or healing after surgical procedures or traumatic injuries at the retroperitoneal area. Also, none of these was present in the second case.

Retroperitoneal fibrosis affect male as twice as females[9]. Our patients are 42 and 55 years of age, this is consistent with the reported age of this disease which predominantly occurs in the forties and fifties[10]. Our patients presented with almost typical clinical features reported[11] as backache, loss of weight, anorexia and general illhealth which is most likely attributed to the slow impairment of the renal function. Retroperitoneal fibrosis has predilection to obstruct the lower third of the ureter[12], but each of our two patients seems to have presented with extensive disease involving the whole length of both ureters.

IVU shows the diagnostic radiological features of retroperitoneal fibrosis as hydronephrosis due to extrinsic compression of the ureters which appear to be medially deviated[13]. In our first case we didn't request IVU because of the renal impairment. Yet, because of fear of malignancy biopsy is the corner stone for the correct diagnosis. However, magnetic resonant imaging (MRI) is reported to differentiate between benign fibrosis and fibrosis infiltrated with malignancy[14]. MRI was introduced in this country two years after establishing the diagnosis of the first case. Histological confirmation of the diagnosis could be done by tru-cut needle biopsy[15], however since surgical ureterolysis is essential we feel open biopsies and freeing of ureters and major vessels is more convenient.

To relief the ureteric obstruction ureteric stenting has high failure rate[16]. Therefore, open surgical ureterolysis remains the most important part of the treatment. It has withstood the test of time; although recent reports have shown that unilateral laparoscopic retroperitoneal ureterolysis carries less morbidity[17]. We have been doing laparoscopic cholecystectomy for more than 10 years, yet we feel that laparoscopic retroperitoneal dissection requires special training. Open surgical ureterolysis does not completely excise the retroperitoneal fibrous mass, but it does free the ureters. Therefore a problem of unilateral recurrence and obstruction of retroperitoneal vessels resulting in lower limb oedema remain a challenge. In addition some patients may present too ill to with stand open bilateral ureterolysis. For these reasons and for all patients with retroperitoneal peritoneal fibrosis the use of steroids gave good results[18-20]. The duration of steroids is suggested to be between six months to two years. Because steroids are known to elevate the blood sugar, and in our patient the disease is associated with resent on set of diabetes we opted to give prednisolone in pulses every two months. However, it is of needless importance to stress in the fact that steroids should mot be given before biopsies were taken because of fear of missing cancer. However, the second case showed good response to combination of prednisolone and tamoxifen but she developed symptoms of salt and water retention which is expected to improve after withdrawal of the drugs.

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