Singapore Med J 1998; Vol 39(9):416-417

Idiopathic Retroperitoneal Fibrosis with Bilateral Lower Ureteric Obstruction – A Case Report with Literature Review

J S B Koh, M Y C Wong, M K Li, K T Foo

ABSTRACT

Whilst prostatic enlargement remains the commonest cause of bilateral hydronephrosis in males greater than 50 years of age, other rarer conditions such as retroperitoneal fibrosis should be borne in mind.

We report a case of bilateral hydronephrosis which was eventually diagnosed as idiopathic retroperitoneal fibrosis and treated with open surgical ureterolysis with omental wrapping.

Keywords: bilateral obstructive uropathy, retroperitoneal fibrosis, open surgical ureterolysis

INTRODUCTION

There are many causes of retroperitoneal fibrosis[1]. The need to exclude possible malignancy with multiple, deep biopsies of the retroperitoneal space cannot be over-emphasised. When malignancy is excluded, treatment is then aimed towards prompt relief of ureteric obstruction.

CASE REPORT

Our patient is a 66-year-old Chinese retired taxi-driver with a medical history of hypertension (on nifedipine and propanolol), as well as ischaemic heart disease. Ultrasonography performed for investigation of asymptomatic renal impairment (creatinine 423 μmol/L) revealed bilateral hydronephrosis.

Cystoscopic examination under local anaesthesia revealed a non-obstructing prostate. Right ureteric catheterisation was performed and showed complete occlusion at the junction of lower and middle third ureter.

As left ureteric catheterisation was unsuccessful, a left percutaneous nephrostomy with antegrade studies was performed, revealing narrowing at the mid-ureter. A right percutaneous nephrostomy was also performed revealing complete obstruction due to extrinsic compression at the level of the right mid-ureter. Bilateral antegrade D-J stenting was performed subsequently.

A CT-scan of the abdomen revealed bilateral hydronephrosis and dilatation of the proximal right and left ureters. A thin mantle of soft tissue was seen anterior to the abdominal aorta and inferior vena cava, suggestive of possible retroperitoneal fibrosis (Fig 1).

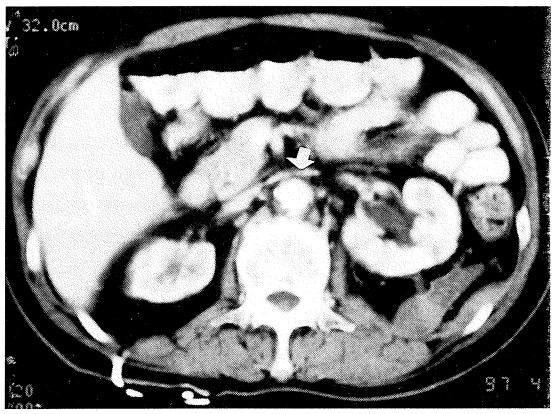

Fig 1 – Arrow showing retroperitoneal fibrosis.

In view of possible retroperitoneal fibrosis, a laparotomy was performed. At laparotomy, liver, stomach and bowels were normal and no para-aortic lymph node was felt. Multiple biopsies of retroperitoneal space with frozen section did not reveal any obvious malignancy. Bilateral ureterolysis was performed with omental wrapping.

Histology confirmed features of retroperitoneal fibrosis with no evidence of malignancy.

The J-stents were removed at 6 weeks post-operatively. The patient recovered well with a reduction in serum creatinine to 152 μmol/L and showed reduced dilatation of bilateral pelvi-calyceal systems on intravenous urogram 6 weeks after ureterolysis.

DISCUSSION

Retroperitoneal fibrosis was first described by Ormond in 1948[2] but two earlier cases may have been included in the French literature by Albarran (in 1905) and Kolischer (in 1922) as suggested by Ormond himself.

The etiology of retroperitoneal fibrosis remains unknown in the majority of cases but it is important to exclude a malignant cause which can only be ruled out histologically. Other known causes of retroperitoneal fibrosis include retroperitoneal injury from trauma, surgery or irradiation, suppurative processes of the abdominal and pelvic organs, urinary tract infections with extravasation urine, and drugs, notably methysergide[1].

However, our patient was not known to have any history of surgery or urinary infections and was only

Department of Urology
Singapore General Hospital
Outram Road
Singapore 169608

J S B Koh, MBBS
Medical Officer

M Y C Wong, M Med (Surg), FRCS (Ed), FAMS
Consultant

K T Foo, MBBS, FRCS (Ed), FAMS
Senior Consultant & Head

Division of Urology
Department of Surgery
National University Hospital
5 Lower Kent Ridge Road
Singapore 119074

M K Li, FACS, FRCSI, FRCS (Glas)
Consultant & Head

Correspondence to:
Dr M Y C Wong

on nifedipine and propanolol which have not been linked to retroperitoneal fibrosis. Our patient had the commonest form of retroperitoneal fibrosis, that is, idiopathic retroperitoneal fibrosis.

Idiopathic retroperitoneal fibrosis has a male pre-dominance of approximately 2:1[3], usually affecting men in their fifth or sixth decades[4] as in this case. The process has a pre-dilection for the lower third of the ureters causing bilateral ureteric obstruction also demonstrated in our patient[5].

The early phase of the disease may have an insidious onset with non-specific flank, abdominal and back pain. Less frequently, patients may complain of weight loss and nausea[6]. Urinary manifestations of oliguria and anuria have also been reported.

Our patient was in fact, asymptomatic and bilateral ureteric obstruction was only suspected when investigations for renal impairment revealed bilateral hydronephrosis on ultrasonography.

Radiological evidence of retroperitoneal fibrosis is characterised by the classical triad of upper ureteral hydronephrosis, medial deviation of the ureters and occasional extrinsic ureteral compression as suggested by Witten[7]. However, the diagnosis can only be confirmed histologically.

CT-scan of the abdomen may occasionally show up the fibrotic plaque. In our patient, the CT showed a thin layer of soft tissue anterior to the abdominal aorta and inferior vena cava, suggestive of possible retroperitoneal fibrosis.

Recently, magnetic resonance imaging (MRI) has been said to be invasive study for differentiating benign fibrosis from that associated with malignancy[8].

Having excluded malignancy, the aim of therapy is directed towards preservation of renal function through prompt relief of obstruction by surgical ureterolysis.

Long-term ureteral stenting is not enough due to high failure rates as shown by Docimo and Dewolf[9]. Open surgical ureterolysis remains the main stay procedure for the relief of ureteral obstruction due to extrinsic ureteral compression by a benign fibrotic process. After ureterolysis, the ureters must be displaced from the area of fibrosis by wrapping them in omentum. Alternatively, lateral or intraperitoneal displacement of the ureters may also be performed.

In some instances, extensive involvement of a small segment of ureter may necessitate excision and re-anastomosis. In the event of a non-functioning kidney, nephrectomy is performed.

The use of steroids has been reported to achieve generally good responses in the early stage of the disease[10,11]. However, in advanced contracted stage, steroids are of minimal value.

More recently, with the advancement of technology, several cases of endourological treatment of retroperitoneal fibrosis via percutaneous balloon dilatation or endoscopic incision, dilatation and stenting have been reported with varying results[12-14].

Endoscopic ureterolysis has also been recently studied as another minimally invasive approach to extrinsic ureteral obstruction. Elashry et al compared the technique with that of open surgery in a series of patients and concluded that laparoscopic unilateral ureterolysis is a less morbid, yet equally effective procedure over conventional open surgery[15]. However, the authors maintain that presently, ureterolysis with omental wrapping remains the first option.

The prognosis for patients with idiopathic retroperitoneal fibrosis is generally good provided the kidney has adequate function at the time of ureterolysis.

CONCLUSION

Retroperitoneal fibrosis is one of the rarer causes of bilateral obstructive uropathy. Having excluded malignancy, prompt relief of ureteric obstruction is the goal. Whilst endourological and laparoscopic techniques have been reported, open surgery remains the method of choice at present.

REFERENCES

1. Lepor H, Walsh PC. Idiopathic retroperitoneal fibrosis. J Urol 1979; 122:1-6.
2. Ormond JK. Bilateral ureteral obstruction due to envelopment and compression by an inflammatory retroperitoneal process. J Urol 1948; 59:1072-9.
3. Ormond JK. Idiopathic retroperitoneal fibrosis: A discussion of the etiology. J Urol 1965; 94:385-90.
4. Koep L, Zuidema GD. The clinical significance of retroperitoneal fibrosis. Surgery 1977; 81:250-7.
5. Hewett AL, Headstream JW. Pericystitis plastica. J Urol 1960; 83:103-7.
6. Pallette EM, Pallette EC, Harrington RW. Sclerosing lipogranulomatosis: Its several abdominal syndromes. Arch Surgery 1967; 94:803-10.
7. Witten DM. Reptroperitoneal fibrosis. I : Clinical Urography. Pollack HM (ed). Philaelphia: WB Saunders Co, 1990:2470-1.
8. Arrive L, Hricak H, Tavares NJ, Miller TR. Malignant versus non-malignant retroperitoneal fibrosis: differentiation with MR imaging. Radiology 1989; 172:139-43.
9. Docimo SG, Dewolf WC. High failure rate of indwelling ureteral stents in patients with extrinsic obstruction: experience at 2 institutions. J Urol 1989; 142:277-9.
10. Longmire WP Jr, Goodwin WE, Buckberge GD. Management of sclerosing fibrosis of the mediastinal and retroperitoneal areas. Ann Surg 1967; 165:1013-22.
11. Ochsner MG, Brannan W, Pond HS, Goodlet JS Jr. Medical therapy in idiopathic retroperitoneal fibrosis. J Urol 1975; 114:700-4.
12. Downey DB, O'Connell D, Smith J, Donohoe J. Percutaneous baloon dilatation of a mid-ureteric obstruction caused by retroperitoneal fibrosis. Br J Urol 1987; 60:84-5.
13. Ward AS, Karanjia ND, Russell AJ. Ureteral obstruction following aortobifermoral bypass: Management by endoscopic baloon dilatation. J Urol 1992; 147:120-2.
14. Mikkelsen D, Lepor H. Innovative surgical management of idiopathic retroperitoneal fibrosis. J Urol 1989; 141:1192-6.
15. Elashry OM, Nakada SY, Wolf JS Jr, Figenshau RS McDongall EM, Clayman RV. Ureterolysis for extrinsic ureteral obstruction: A comparison of laparoscopic and open surgical technique. J Urol 1996; 156:1403-10.