Case Report Of A Patient With Bilateral Hydronephrosis Secondary To Retroperitoneal Fibrosis

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Citation

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Abstract

Retroperitoneal fibrosis is a rare condition. It is characterised by inflammation and formation of fibrous tissue in the retroperitoneum. We describe the case of an 85-year-old Caucasian male who presented with acute renal failure and bilateral ureteric hydronephrosis, secondary to retroperitoneal fibrosis. The initial management of his condition is also described.

INTRODUCTION

Retroperitoneal fibrosis is a rare condition with associated medical and surgical complications. The diagnosis of this condition requires a high index of suspicion, as the clinical features of retroperitoneal fibrosis is often non-specific. Most cases of retroperitoneal fibrosis are idiopathic, but there are other specific causes, including drugs and malignancy.[1]The main diagnostic tool is imaging, and the detection rate of this condition is further enhanced with the advent of cross-sectional imaging. We report the case of an 85-year-old male who presented with acute renal failure and bilateral hydronephrosis secondary to retroperitoneal fibrosis.

CASE REPORT

An 85-year-old Caucasian male presented to the emergency department with 6 months history of non-specific symptoms, which included weight loss, anorexia and lethargy. This gentleman was known to have various co-morbidities, including ischaemic heart disease, hypertension, hypercholesterolaemia and gout. On admission, he was found to be in acute renal failure with a creatinine of 250 µmol/L.

Apart from an enlarged prostate on digital rectal examination, his physical examination was otherwise unremarkable.

His renal tract ultrasound showed bilateral hydronephrosis, a distended bladder and an enlarged prostate. With the initial working diagnosis of prostatic enlargement causing obstructive nephropathy, a urinary catheter was inserted to relieve the obstruction. However, despite catheterisation, there was no significant diuresis.

He subsequently underwent a non-contrast CT, which showed a soft tissue mass with significant fat-stranding surrounding the abdominal aorta, compressing both ureters.(Fig. 1)

Figure 1

Fig 1. Cross-sectional image on CT-scan showing retroperitoneal mass enveloping the abdominal aorta

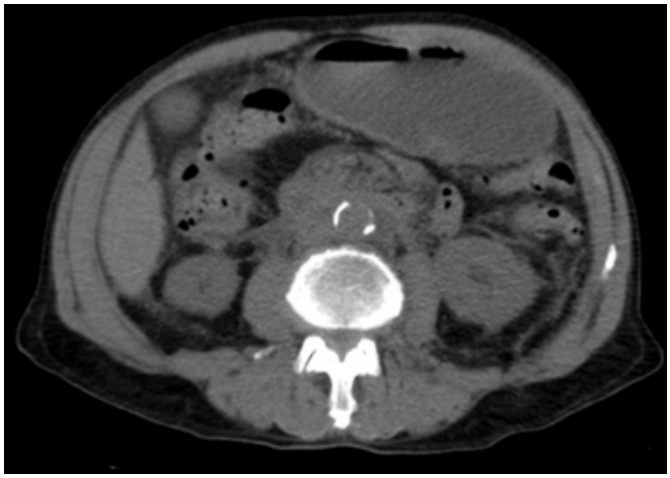
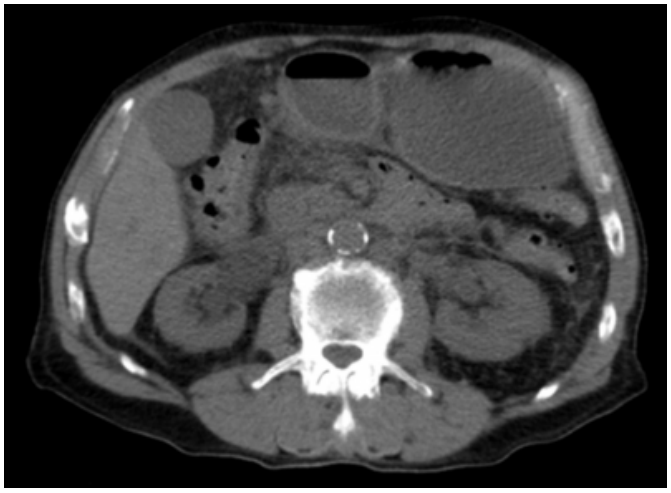


Figure 2



To ensure ureteric patency and to protect kidney function, he had ureteric double-J stents inserted bilaterally. This procedure achieved good clinical results, as he consequently had adequate diuresis with demonstrable improvement in renal function.

Further investigations including tumour markers were performed to identify a primary cause for the retroperitoneal fibrosis. These revealed elevated tumour markers in our patient, with Ca 12-5 of 9494 and a CA19-9 of 11018. These results were highly suspicious of a pancreatic carcinoma.

Ideally, the next step of investigation where there is a suspicion of malignancy is to perform a biopsy. However, due to the existing co-morbidities of the patient, a consensus was made to respect the patient's wishes and not pursue for a tissue diagnosis of pancreatic cancer. He was nevertheless commenced on long-term oral steroids.

DISCUSSION

Retroperitoneal fibrosis is a rare condition characterised by an inflammatory mass with fibrosis, usually centred around the fourth and fifth lumbar vertebra[2], with the potential of obstructing retroperitoneal structures. It has an estimated prevalence ranging from 1.38 in 100,000 to 1 in 200,000[3,4]and has a male preponderance (approximate male to female ratio of 2 – 3 :1).[1,5]It usually affects patients ranging from 40 to 60 years of age.[6,7,8]

Most cases of retroperitoneal fibrosis are idiopathic, with a specific cause or association identified in only about a third of all cases[1,9]>, and may include malignancy, drugs,[1]occupational exposure to asbestos[7], or chronic inflammatory disease/autoimmune disease[10,11], among others.

There are various causes for retroperitoneal fibrosis. A malignant process must however be specifically pursued and ruled out first.

Patients with uncomplicated retroperitoneal fibrosis often present with non-specific symptoms. These symptoms may include back and abdominal pain, weight loss, malaise or pyrexia.[1,12]If there is associated structural compression from the condition, patients may present with acute renal failure if the ureters are obstructed, varicoceles if the gonadal veins are affected, and lower limb oedema.[13]

Diagnostic imaging is the primary modality of diagnosis, with CT and MRI being the current modalities of choice. On CT, the retroperitoneal fibrosis appears as a mass with similar density to muscle, enveloping the aorta and inferior vena cava, and has variable contrast enhancement depending on the stage of the disease process.[12]Hydronephrosis can also be observed in cases of ureteric obstruction. MRI imaging can be of assistance in providing further characterisation of the dimensions and extension of the plaque.[14]

For optimal care in retroperitoneal fibrosis, a multidisciplinary approach combining non-surgical and surgical therapies should be employed. The aims of management are to firstly rule out an underlying malignant process, and subsequently to prevent secondary organ damage, typically renal failure from ureteric obstruction.

Patients who have retroperitoneal fibrosis often present with some degree of ureteric obstruction and hydronephrosis, which may require ureteral stenting to ensure patency and preservation of renal function. This less invasive approach is a suitable option for the high-risk and elderly patients. Open ureterolysis, although highly effective[15], is associated with a significant morbidity.[16]

The surgical options above can be complemented with the medical therapy of prednisolone[17,18]which would serve to prevent ongoing inflammatory process and inhibit maturation of fibrotic tissue. Azathioprine can be used as a steroid-sparing agent, in combination with steroids or can be used as a substitute when steroid treatment has failed.[19,20]Other agents, such as tamoxifen[21]and mycophenolate mofetil can also be considered.[22,23]

Given the potential for end-organ damage such as renal failure, it is advisable to get early involvement of the nephrologist in the care of these patients. Further, the disease progression should be monitored with regular blood tests,

including an ESR (Erythrocyte Sedimentation Rate) and creatinine levels.[8]Repeat radiologic assessments with a CT or MRI should also be performed[8], with the prolongation of imaging intervals once the disease process has stabilised. Given the potential for late ureteric stenosis, long-term follow-up of these patients is necessary.

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