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Case report

An atypical cause of retroperitoneal fibrosis: Case report and literature review

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KEYWORDS
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Abstract
Introduction: Retroperitoneal fibrosis (RPF) is a rare inflammatory disease resulting in the development of a retroperitoneal mass, which may encase the aorta and its branches, as well as the ureters.
Observation: The systemic inflammatory response causes constitutional symptoms. The mass itself leads to symptoms of non-specific back pain and abdominal pain, as well as ureteric obstruction and subsequent renal insufficiency. Immunoglobulin 4 (IgG4) has recently been found to be involved in the pathogenesis of multiple autoimmune and inflammatory disorders, including RPF. Blood work-up, imaging and biopsy of the mass remain the mainstay of diagnosis.
Conclusion: RPF remains a diagnosis of exclusion. Treatment hinges on corticosteroids, but other immunosuppressants and disease-modifying agents are also being used more commonly. Surgical intervention is only carried out when conservative measures have failed or are contraindicated, and in order to preserve renal function.

Introduction

Retroperitoneal fibrosis (RPF) is a systemic inflammatory disease resulting in retroperitoneal plaque formation. This mass is initially inflammatory in nature, and later evolves into a fibrotic mass [1]. The estimated incidence varies between 0.1 in 100 000 and 1 per 200 000 in different studies, but little epidemiological data exists to confirm or refute this [1]. Much controversy surrounds the diagnosis and treatment of this disease. With this case report, we wish to highlight a patient with immunoglobulin-mediated RPF confirmed on histology.

Case report

A 56-year-old male, with no known comorbidities, presented with right flank pain. The patient was found to have microscopic

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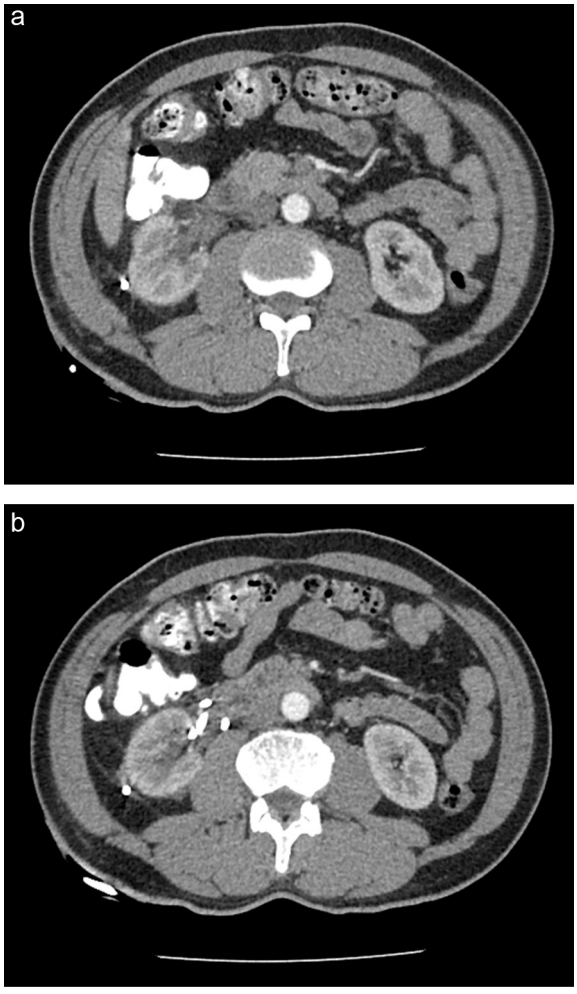

Fig. 1 (a) Mass at the level of the renal hilum. (b) Mass at the level of the renal hilum – nephrostomy visible in proximal ureter.

haematuria on urine dipsticks on admission, as well as abnormal renal function, with a creatinine of 134 μmol/L. The erythrocyte sedimentation rate (ESR) was 60, with a white cell count (WCC) of 7.2×10^9.

On renal ultrasound, the patient was found to have right-sided hydronephrosis and proximal hyroureter. The patient then underwent an uncontrasted computer-tomography (CT) scan of his abdomen and pelvis in order to assess for a cause for the unilateral hydronephrosis. The CT scan revealed a mass extending from the level of the renal hilum on the right and encasing the right ureter as noted in Fig. 1.

A right percutaneous nephrostomy tube was inserted as an emergency, and the patient's renal function subsequently decreased to 109 μmol/L. A double-J (DJ) ureteric stent was placed (antegrade) while pending further work-up. The patient was also started on high-dose oral glucocorticoid therapy due to the CT features and raised ESR suggesting RPF. Despite ureteric stenting and glucocorticoid therapy, it was not possible to remove the percutaneous nephrostomy tube. During attempted clamping of the nephrostomy tube, the patient experienced pain relieved only by unclamping of the tube. The cause was thought to be related to extrinsic compression by the mass.

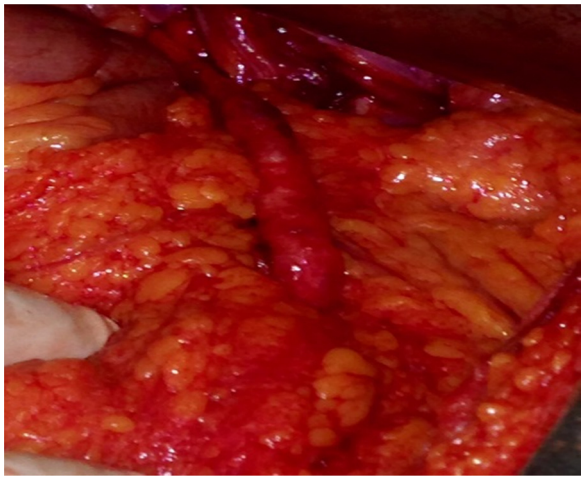

Fig. 2 Right ureter mobilised with omentum to be used for wrapping ureter.

Diagnostic and therapeutic laparoscopy was performed in view of the ongoing obstructive uropathy despite ureteral stenting and glucocorticoid therapy. The mass was noted to resemble an inflammatory plaque, and frozen section specimens were sent intra-operatively. The results of frozen section revealed inflammatory tissue, with no features of malignancy. Ureterolysis was performed, whereby the ureter was released from the inflammatory plaque and wrapped in omentum, as seen in Fig. 2. Unfortunately, it was necessary to convert to laparotomy due to challenges with tissue planes and the proximity of vital structures to the obstructed portion of the ureter.

The percutaneous nephrostomy tube, as well as the DJ stent were both removed within 6 weeks of the ureterolysis procedure. The patient has normal renal function and is currently asymptomatic. Formal histology confirmed the diagnosis of IgG4-related retroperitoneal fibrosis with extensive collagenous fibrosis, dense lymphoplasmacytic chronic inflammation, obliterative phlebitis (Fig. 3a), and >30 $IgG4^+$ plasma cells per high power field (Fig. 3b).

Discussion

RPF was first described by Ormond in 1948 [2]. The classical description is that of a retroperitoneal plaque encasing the great vessels and the ureters from the level of the renal hilum up to the pelvis [1]. Seventy percent of these cases are idiopathic in nature, with the other 30% having an underlying precipitating cause [1,3]. The most important of these underlying causes that a treating physician must rule out is a malignant process. Malignancy has been found in 8–10% of cases of patients with RPF [1]. Other precipitants are documented in Table 1 [1,4].

Other disease processes that may be associated with RPF, but not necessarily precipitating causes, include [1,3]:

- Sclerosing cholangitis
- Pancreatitis
- Riedel's thyroiditis
- Ankylosing spondylitis
- Uveitis
- Systemic lupus erythematosis