

Retroperitoneal Fibrosis: Diagnostic Challenge and Imaging Features

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Abstract

Retroperitoneal fibrosis (RPF) is a rare condition with an incidence of 1.3 per 100,000 population with a peak age of onset at 40 – 65 years old. Almost two-thirds of the retroperitoneal fibrosis cases are idiopathic and the rest are secondary to multiple factors such as drug use, malignancies, inflammations, and infections. The idiopathic form of retroperitoneal fibrosis is also known as Ormond's disease. Here, we are presenting a case of a woman who was referred to our center from the clinic, due to worsening serum creatinine. After a series of laboratory investigations and imaging findings, the patient was diagnosed with retroperitoneal fibrosis. We would like to discuss this case due to the rarity of the disease and the imaging dilemma that often arises in its diagnosis. We have also discussed methods to differentiate between primary and secondary retroperitoneal fibrosis specifically based on CT findings. This is to provide guidance for the diagnosis and prompt management of cases, especially in district hospitals with no advanced diagnostic equipment such as MRI or biopsy.

Keywords

Retroperitoneal fibrosis, idiopathic, imaging, diagnostic

Introduction

RPF was initially described in 1905 by a French urologist named Albaran, who reported a surgical treatment causing retroperitoneal fibrotic process leading to ureteric obstruction. The condition was then established by Ormond when he published 2 cases in 1948 (3). It is characterized by tissue proliferation and chronic inflammation of a vast range of causes that could be idiopathic or secondary to drug, infection, inflammation, and malignancy (2). Diagnosis of this disease has always been a challenge both in clinical and radiological aspects, especially in district hospital settings.

Case Report

A 62-year-old lady with a background history of hypertension was referred to our center for KUB ultrasound due to frothy urine and worsening serum renal profile. Initial ultrasound revealed moderate to

gross right hydronephrosis and mild left hydronephrosis. No renal or ureteric calculus was noted. The proximal ureters were dilated bilaterally. However, the assessment of the distal ureters was obscured by bowel gas. A computed tomography (CT) urography performed revealed a homogeneously enhancing periaortic soft tissue density which appeared isodense to the surrounding muscle. The mass was encasing the aorta from the L1/L2 level down to the aortic bifurcation and also to involve the right common iliac, internal, and the proximal external iliac arteries. There was associated mild stenosis of the distal right internal iliac artery. However, the vessel remained patent. Distally, the mass also encased the inferior vena cava and extended to the right common iliac vein with loss of fat plane in between. This soft tissue mass also involved the adjacent bilateral ureters distally causing medialisation and obstruction of the ureters.

She was negative for all the connective tissue screening markers including anti-mitochondrial antibody (AMA), anti-smooth muscle antibody (SMA), anti-nuclear antibody (ANA), rheumatoid factor (RF), anti-thyroid peroxidase (TPO), C3, C4, pANCA, cANCA and serum protein electrophoresis. However, her immunoglobulin (IG) assays (IgA, IgG, IgM) were slightly raised above the normal limits. Subsequently, a percutaneous nephrostomy tube was inserted on the right side and the patient was started on a tapering dose of prednisolone for 3 months. Repeated post-treatment CT abdomen showed a significant reduction in the size of the retroperitoneal soft tissue density suggestive of treatment response. There was also evidence of improvement in bilateral obstructive uropathy, however the medialisation of bilateral ureters persisted.

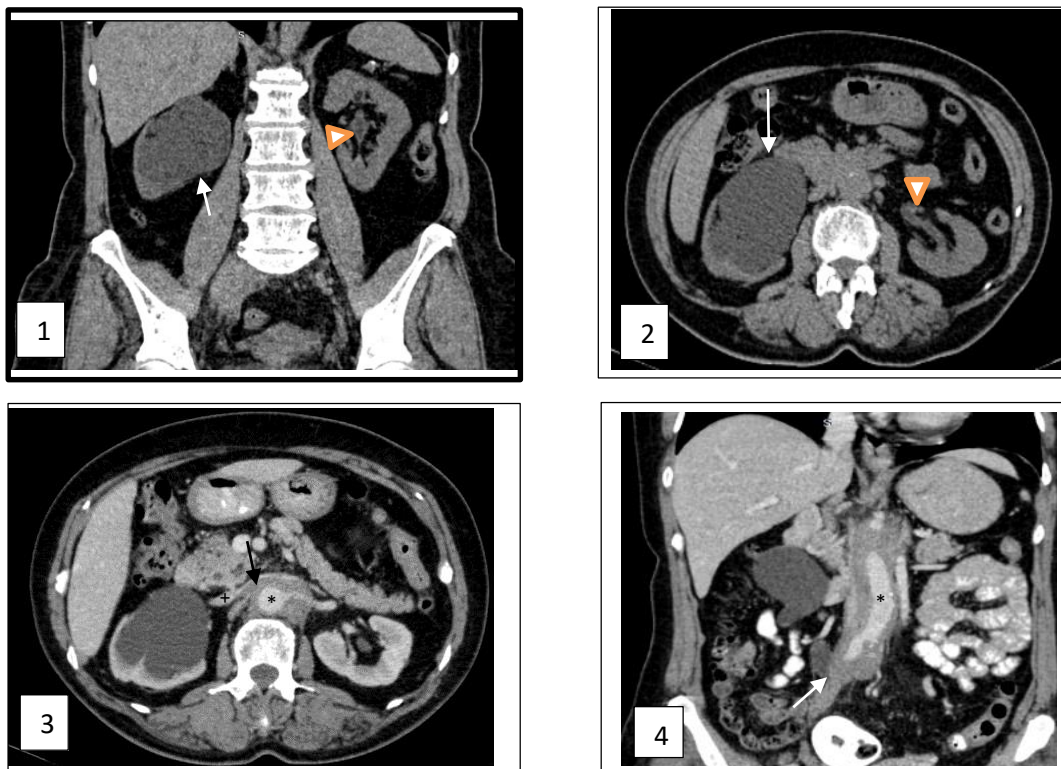


Figure 1 & 2 showed plain CT abdomen in axial and coronal views.

Presence of bilateral obstructive uropathy, moderate (white arrow) on the right and mild (arrow head) on the left.

Figure 3 & 4 showed contrasted CT in coronal and axial views. The images are to show homogeneously enhancing mass (black arrow) encased the aorta (*) and IVC (+). On figure 3, the mass shown encasing the right ureter (white arrow).

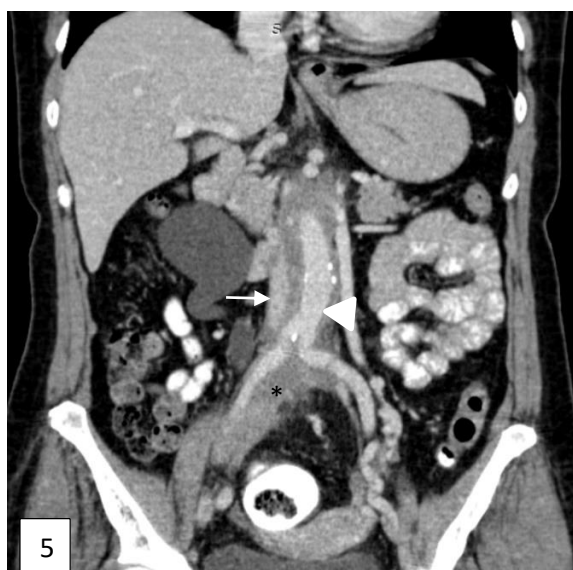


Figure 5: Contrast-enhanced CT Abdomen in coronal views at the time of diagnosis.

Figure 6: Post-treatment showing reduction in the size of the mass (*), IVC (white arrow), Aorta (arrowhead).

Discussion

Idiopathic RPF is a rare entity with an incidence of 1.3 per 100 000 population (1). It has a peak incidence between the ages of 40 – 60 years old with predominance in male patients. The estimated male-to-female ratio is approximately 2:1 or 3:1 (4).

RPF is characterized by the atypical fibroinflammatory proliferation of the retroperitoneal soft tissue commonly involving the infrarenal abdominal aorta, IVC, common iliac arteries as well as its branches (1). The causes can either be idiopathic or secondary. The idiopathic disease is characterized by the chronic inflammation of the tunica adventitia of the abdominal aorta, iliac arteries, and the surrounding retroperitoneal tissue (3). Secondary causes of RPF are often due to drugs, neoplasm such as lymphoma, retroperitoneal sarcoma, carcinoid tumor as well as metastatic disease from the stomach, colon, lung, breasts, genitourinary, or even thyroid. Few predisposing factors have been identified which include infections, radiation, major abdominal surgery, retroperitoneal hemorrhage or hematoma and proliferative diseases such as Erdheim-Chester disease (1). There are also identified risk factors in a study which include hypertension, diabetes mellitus, coronary artery disease, dyslipidemia, cerebrovascular disease, tobacco and asbestos exposure (5).

Pathogenesis of idiopathic RPF can be multifactorial. Atherosclerotic aortic disease has been identified as one of the common cause in patients with idiopathic RPF and is associated with chronic periaortitis. In this condition, it has been considered due to an excessive local inflammatory response to antigens such as oxidized low-density lipoproteins and ceroid, which are usually present in atherosclerotic plaques of the aorta (3). Often these patients present with constitutional symptoms, raised acute phase reactants such as ESR and CRP, positive autoantibodies as well as autoimmune diseases involving other organs which suggest idiopathic RPF could also be systemic rather than a local reaction to atherosclerosis (3), (5). There is also another mechanism identified to suggest its relation to genetic condition associated with HLA-DRB1*03 allele (3), (6). In recent years where immunoglobulin G4 (IgG4) related diseases are becoming known,

certain studies have identified the presence of IgG4-bearing plasma cells in the biopsies of patients with idiopathic RPF (7). IgG4 related diseases encompass various immune mediated systemic syndrome with involvement of multiple organ systems which include idiopathic RPF, autoimmune pancreatitis, sclerosing cholangitis as well as pericardial fibrosis (7), (1).

Patients with idiopathic RPF very often present in the later stage. Initial manifestation of the disease is often vague such as malaise, fever, anorexia, weight loss, nonspecific abdominal and flank pain (1). In almost all patients, 56-100% of the cases tend to have obstructive uropathy due to ureteric entrapment which in long term may lead to renal failure (1). Even though idiopathic RPF had been a known disease, diagnosis of the disease is still a challenge in clinical settings mainly due to the absence of any established diagnostic criteria (6). Hence, radiological imaging plays a major role in diagnosing this condition. Ultrasonography is often used as an initial assessment mainly to assess the presence of its complications such as hydronephrosis and aortic dilatation or aneurysm (8). The lesion can be seen on ultrasound as an isoechoic or hypoechoic mass at the lower lumbar to sacral promontorium (1), (3). It is often subtle and early findings may be missed in ultrasound due to the overlying gas or fluid filled bowel which obscures the retroperitoneal structure (1),(9) as noted in this case.

CT and magnetic resonance imaging (MRI) are still the mainstays of diagnostic tools as they are non-invasive. CT features of RPF includes homogenously enhancing well-defined fibrous tissue which surrounds the major vessels and ureters (10). Often it is also associated with medial displacement of the ureters as noted in this study (8). There had been a classification based on the fibrosis distribution which was published only in certain studies (5), (10). The classification is as follows:

Class I: involves aorta and iliac arteries.

Class II: involves inferior vena cava.

Class III: has lateral extension to the ureters.

Class IV: extends to the renal hilum with involvement of the renal artery or the renal vein.

CT can also be used to differentiate between idiopathic and secondary disease based on certain morphological features described. Idiopathic form usually has benign features of being smaller and has plaque-like densities with peripheral infiltration as observed in our patient (8). It is also usually located distal to the renal hilum, anterolaterally surrounding the aorta and does not cause displacement of the aorta. It only causes medialization of the ureters. Secondary form of RPF often present as nodular and lobulated mass displacing the psoas muscles as well as invading the adjacent bony structures. Idiopathic form also often present with localized lymphadenopathies whereas secondary form has enlarged and confluent lymph nodes which often surrounds the vessels (8). Secondary form of RPF, such as lymphoma also shows heterogeneous enhancement pattern or peripheral enhancement with central hypodensity to suggest intranodular necrosis while RPF shows homogeneous enhancement (11). RPF enhancement pattern also enables differentiation between acute and chronic RPF. In acute phase the mass often shows an avid enhancement while in inactive phase it has reduced or no enhancement. This enhancement pattern can also be considered as treatment response (1). CT also has high sensitivity to identify regression in follow up cases and can be suggestive of treatment response (1). These allows CT to be mainstay method of diagnosis. Even though tissue biopsy is the gold standard, however it is not routinely done. Only certain conditions requires histopathologic examination to be mandatory such as when features of the mass suggestive of malignancy, atypical location of the mass (i.e. pelvic, peripancreatic, perirenal) and if there is treatment failure (1).

Conclusion

RPF remains a challenging condition for diagnosis and imaging plays a main role in it. It should be considered as a differential in retroperitoneal mass. The condition should be diagnosed based on the imaging criterions as described.

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