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Idiopathic Retroperitoneal Fibrosis: Case Report with Review of the Literature

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Abstract: Background*: Idiopathic retroperitoneal fibrosis (RPF) is a subtype of chronic periaortitis (CP). CP is a rare disorder characterized by the presence of a fibro-inflammatory mass usually surrounding the abdominal aorta and the iliac arteries.* ***Case Presentation****: We report the case of A 45-year-old woman who was admitted to the hospital because of abdominal pain, fever, and weight loss. And the diagnosis of retroperitoneal fibrosis was made by CT.* ***Conclusion:*** *The clinical picture of idiopathic RPF is characterized by abdominal, back, or lumbar pain, usually associated with constitutional symptoms such as fever and weight loss, imaging findings can confirm the diagnosis.*

Keywords: retroperitoneal, fibrosis, idiopathic

INTRODUCTION

Idiopathic retroperitoneal fibrosis (RPF) is a rare disease and is considered a subtype of chronic peri aortitis (CP). CP is a rare disease characterized by the presence of a fibroinflammatory mass usually surrounding the abdominal aorta and iliac arteries [1]. This mass can entrap adjacent structures such as the ureters and inferior vena cava. There are three main subtypes of CP, namely inflammatory abdominal aortic aneurysms (IAAA), perianeurysmal retroperitoneal fibrosis (PRF), and idiopathic PRF. In idiopathic RPF, the aorta is often not dilated [2]. In AAPI and PRF, aneurysmal dilation of the aorta is present, and both forms of PC are referred to as aneurysmal RPF. In IAAA, the periaortic mass does not affect adjacent organs, whereas, in PRF, it causes obstructions. [1,3]

We report a case of a woman presenting nonspecific signs such as abdominal pain, fever, and weight loss and whose diagnosis was made by abdominal CT scan**.**

CASE PRESENTATION :

A 40-year-old woman was admitted to the hospital because of abdominal pain, fever, and weight loss Routine laboratory tests. Routine laboratory tests performed on admission revealed elevated Erythrocyte Sedimentation Rate [ESR] - 73 mm), C-reactive protein [CRP] – at 127 mg/l). Other values, including creatinine and BUN, were within normal limits.

Abdominal ultrasound showed dilatation of the pyelocaliceal system of the left kidney.

The abdominal CT revealed a concentric retroperitoneal mass beginning 23 mm below the origin of the right renal artery. Hence, a diagnosis of retroperitoneal fibrosis was made

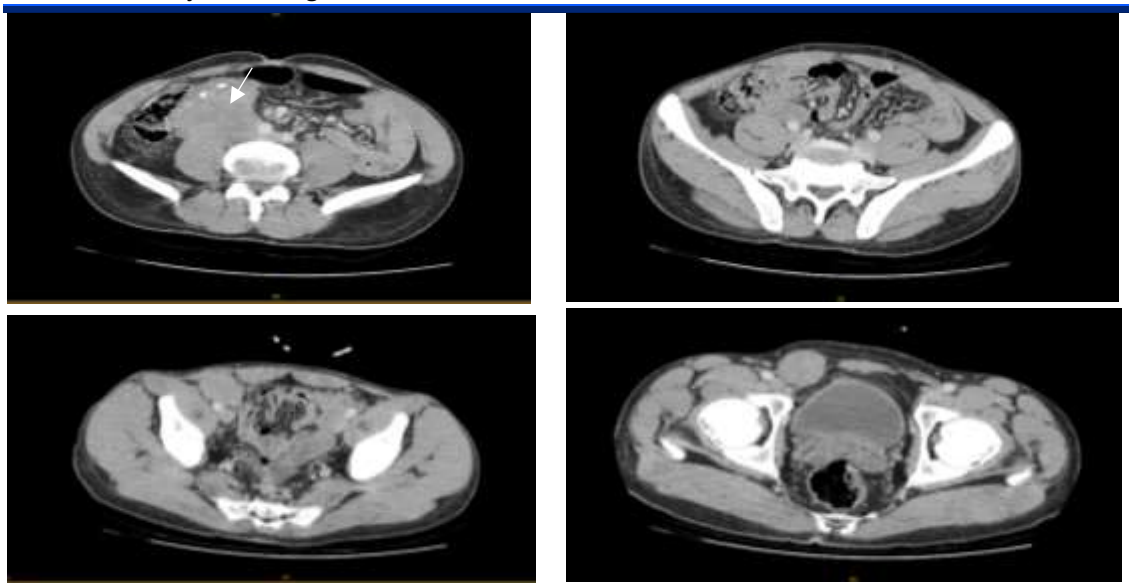

FIGURE 1: INITIAL CT SCAN.

. A JJ catheter was placed in the left ureter and treatment with corticosteroid steroids (prednisone - 60 mg/day) and azathioprine (150 mg/day) was started. The JJ catheter was removed after 6 months. A follow-up ultrasound performed after the removal of the ureteral catheter showed a normal left urethra.

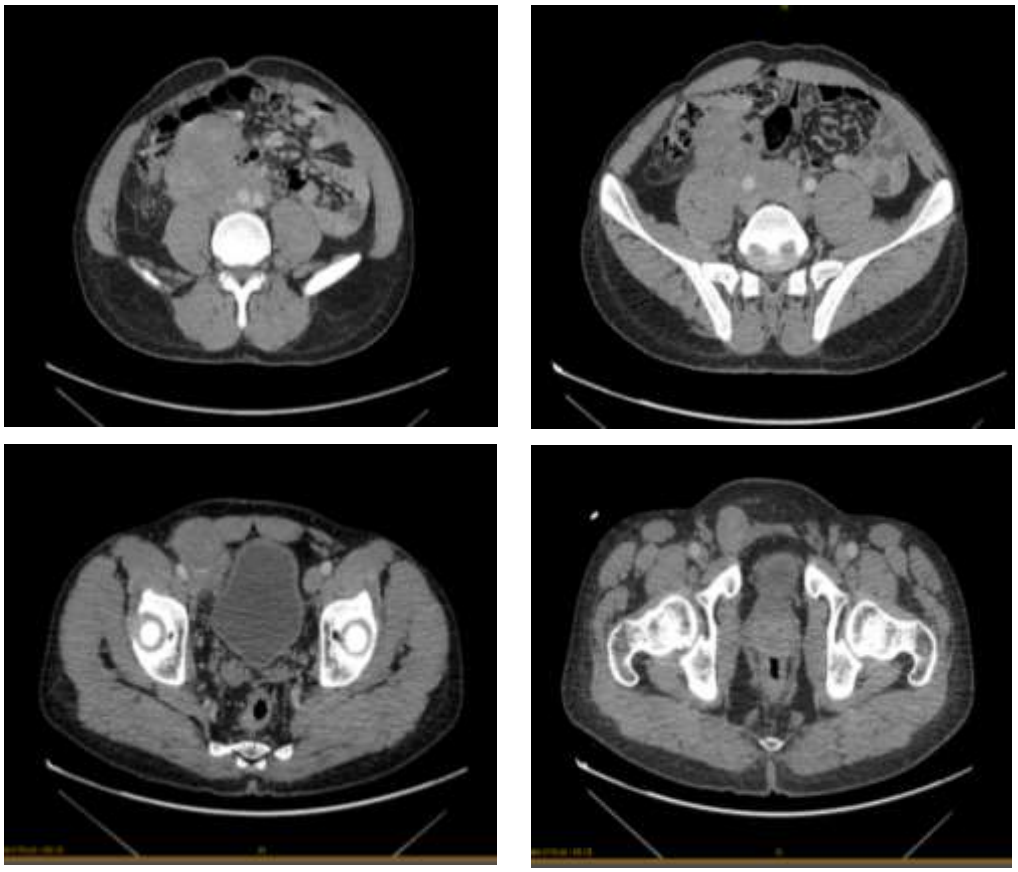

Figure 2 : follow up ct scan

DISCUSSION :

The clinical picture of idiopathic RPD is highly variable and can be characterized in many cases by abdominal pain, back pain or even back or lumbar pain, usually associated with other general symptoms such as fever and weight loss. Our patient had most of these symptoms. [4] RPD can also present with complications. The most common serious complications of idiopathic RPD are hydronephrosis and renal failure [4,5]

It is estimated that about 80% of patients with RPD develop renal failure early or late in the course of the disease. The management of idiopathic RPD relies mainly on corticosteroids and immunosuppressive drugs. In the case of ureteral obstruction, procedures to relieve the obstruction are performed, including the most common method which is nephrostomy, also [Ureteral stenting usually remains sufficient. Another surgical method, ureterolysis, may be necessary in cases of advanced stenosis. Given the lack of trials

of randomized Controlled Trials, recommendations for the treatment of idiopathic PSI have not yet been developed. [7]

Imaging features :

Contrast-enhanced abdominal CT SCAN is the mail diagnosis modality, retroperitoneal fibrosis is presented classically as a soft mas density , it develops around the aortic bifurcation and extends proximally where it may envelope the kidney hilum. It surrounds but does not involve or obstruct the vascular or ureteral vessels. Ureteral obstruction and venous thrombosis may occur, however. In early or active stages, variable enhancement may be seen with intravenous contrast, whereas no enhancement may be seen in the quiescent disease.

MRI: MRI is as sensitive as CT in assessing retroperitoneal fibrosis, with the added advantage of a high contrast resolution between closely spaced retroperitoneal structures. It can evaluate the urinary tract using fast T2 spin-echo weighted sequences without requiring intravenous contrast in patients with compromised renal function. The soft tissue mass is generally dark on T1W and T2W, except in cases of active inflammation, where T2W images may be hyperintense.

CONCLUSION :

Idiopathic retroperitoneal fibrosis (RPF), is a rare fibro-inflammatory disease that develops around the abdominal aorta and the iliac arteries, and spreads into the adjacent retroperitoneum, where it frequently causes ureteral obstruction and renal failure. The diagnosis is generally made by cross-imaging and the management is based on corticosteroids and immunosuppressive drugs.

ABREVIATIONS :

RPF : Idiopathic retroperitoneal fibrosis

CP : chronic periaortitis

IAAAs : inflammatory abdominal aortic aneurysms

CRP : C reactive protein

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