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A Rare Case of Retroperitoneal Fibrosis Extending into the Peritoneum: Case Report

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Abstract **Case Report**

Retroperitoneal fibrosis (RPF) is a rare disease with an incidence de 0-1/ 100000 inhabitants per year and is associated with chronic inflammatory fibrosis of the retroperitoneum. It may be idiopathic or secondary to neoplasic or multisystemic diseases. It usually develops around the abdominal aorta and the iliac arteries, and spread into the adjacent retroperitoneum, where it frequently causes ureteral obstruction. We report in this work a rare case of retroperitoneal fibrosis extending through the superior mesenteric artery into the peritoneum. To our knowledge, no similar case is reported in the literature.

Keywords: Retroperitoneal fibrosis (RPF), retroperitoneum, peritoneum, multisystemic diseases.

INTRODUCTION

Retroperitoneal fibrosis (RPF) is a group of diseases characterized by the proliferation of abnormal fibroinflammatory tissue, usually surrounding the infrarenal portion of the abdominal aorta, the inferior vena cava (IVC) and the iliac vessels. This can spread to neighboring structures, frequently resulting in entrapment and obstruction of the ureters and usually leading to renal failure [1]. It is typically seen in people 40–60 years old [2], and men are two to three times more likely to develop retroperitoneal fibrosis than women. Most (> 70%) cases are thought to be idiopathic [3]; the remainder occur in association with inflammatory disorders, malignancies, or medications [1, 4]. Diagnosis and management of RPF represent a challenge for clinicians. Because of the nonspecific nature of the symptoms and the lack of sensitive and specific laboratory tests, RPF is frequently detected at imaging and multidetector computed CT plays a key role in the diagnosis and ultimately management by helping distinguish benign and secondary forms of RPF [1, 5].

In this article, we report a case of peri aortic fibrosis originating in the sus renal abdominal aorta, and extending into the peritoneum by encircling the coeliac trunk and superior mesenteric artery. To our knowledge, no similar case has been described in the literature.

PRESENTATION OF THE CASE

A 65-year-old male with no particular medical or surgical history presented with mild abdominal pain that was slowly increasing over a period of three months and was poorly responsive to analgesic treatments and a weight loss calculated at sixteen kilograms over the same period. At examination the patient was cachectic but both hemodynamically and respiratory stable and presented an epigastric tenderness. Laboratory tests revealed an elevated CRP count and normal renal function. The patient was then referred to our radiology department for an abdominal CT scan that revealed a circumferential mass surrounding the abdominal sus renal aorta and its visceral branches, extending into the peritoneum following the path of the superior mesenteric artery. It appeared as a well-defined circumferential soft tissue mass with irregular margins, with no arterial displacement spontaneously hypodense and poorly enhanced after contrast acquisition in the venous phase, the mass appeared to be originating from the retroperitoneum, circling the aorta in its hepatic portion, the coeliac trunk, and followed the path of the superior mesenteric artery into the peritoneum (Figures 1 and 2). The abdominal CT revealed no enlarged lymph nodes or splenomegaly or other features suggesting a malignant etiology.

An ultrasound was later performed and revealed a circumferential mass surrounding the aorta

extending from the origin of the coeliac trunk and following the path of the superior mesenteric artery into the peritoneum. It was hypoechoic with little to no vascularization on the color doppler. The wall of the aorta and its dividing branches appeared normal without intimal thickening or atheromatous calcifications.

A thorough examination was later performed; tumor markers and IgG/Total IgG ratio were analyzed and came within the normal margins. A thoracic CT was also prescribed to rule out a neoplasic origin and revealed no significant findings. The diagnosis of RPF was retained in view of the negativity of the etiological work-up and the suggestive appearance on imaging. The patient was put under steroids with a noticeable improvement in clinical condition and weight gain at the two month follow up. Considering the clinical evolution, we decided there was no need for a biopsy of the lesion.

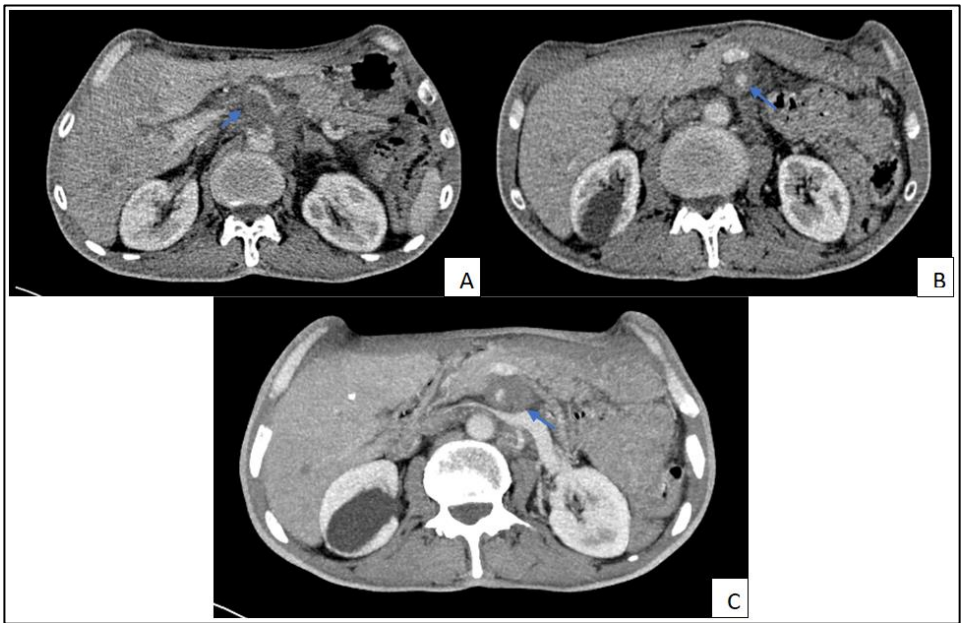

Figure 1: Axial images of abdominal CT with contrast showing a hypodense soft tissue mass with irregular margins (arrow), circling the coeliac trunk (A) and superior mesenteric artery(B and C)

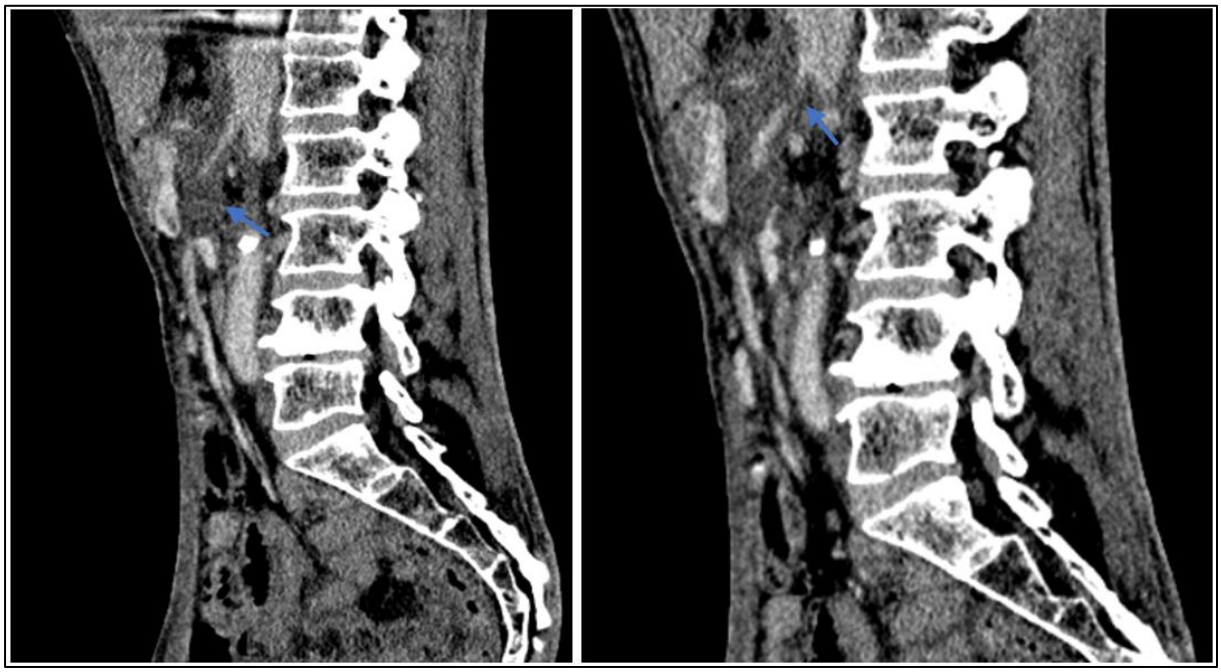

Figure 2: Sagittal images of abdominal CT with contrast showing a hypodense a circumferential mass (arrow) surrounding the abdominal sus renal aorta and its visceral branches, extending into the peritoneum following the path of the superior mesenteric artery

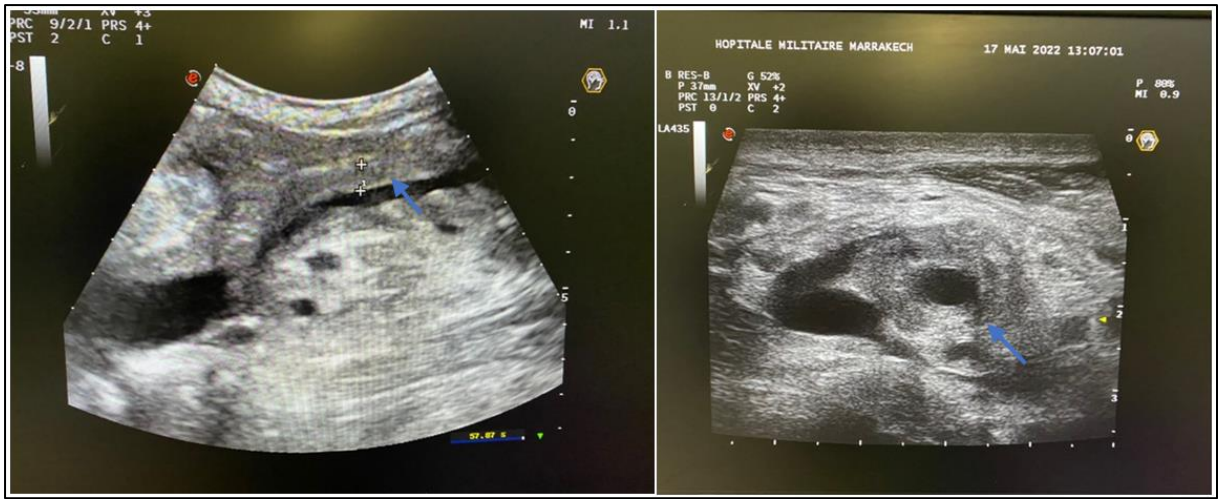

Figure 3: Axial (A) and sagittal (B) ultrasound images showing a hypoechoic a circumferential mass (arrow) surrounding the superior mesenteric artery

DISCUSSION

RPF refers to a group of diseases characterized by the abnormal proliferation of inflammatory tissue fibers surrounding the abdominal aorta, inferior vena cava, and iliac vessels [6]. It can be idiopathic or secondary to malignancy, radiotherapy, medication, or infection. Idiopathic RPF is a rare disease, usually starts from retroperitoneum surrounding the abdominal aorta and spreads caudally to involve the iliac vessels and laterally to involve the inferior vena cava. Fibrosis spreading to the adjacent retroperitoneum frequently reaches the ureters and causes obstructive symptoms with subsequent renal failure if presented late [7].

Disease pathogenesis is still unclear, as most of the literature reviewing RPF is made up of case reports and case series with no clear guidelines. The hypotheses advanced include genetics, immunology IgG4 deposition, and atherosclerosis [7].

The disease typically manifests between the ages of 40 to 60 years old [8] and presents with nonspecific symptoms such as weight loss, malaise, anorexia, and low-grade fever [9]. As the disease progresses, obstructive uropathy and subsequent renal failure, lower extremity edema, and claudication may develop [10]. Laboratory findings reveal a high erythrocyte sedimentation rate and C-reactive protein level, and anemia. Because of the nonspecific symptoms and signs of this disease and the lack of sensitive and specific laboratory tests, imaging studies have played an important role in the diagnosis of RPF [11, 12] and the diagnosis of retroperitoneal fibrosis and the differentiation between RPF and other inflammatory and malignant processes has remained a challenge. Many authors initially emphasized the need for open biopsies during surgery [3, 13]. Currently this attitude has been shifted in favor of typical imaging features of CT or MRI, rather than percutaneous biopsy, to make the diagnosis [14]. CT usually reveals a well- defined but irregular para-aortic soft tissue mass extending from the level of the renal vessels to the iliac vessels that often entraps bilateral ureters, IVC, and abdominal aorta. The degree of contrast enhancement depends on the stage of the disease [15]. Obvious contrast enhancement is noted in the early stage of the disease, but little or absent contrast enhancement is noted in the late or inactive stage [16]. while MRI shows a low signal intensity in the para-aortic soft tissue mass on T1-weighted images and high signal intensity on T2-weighted images with obvious contrast enhancement on contrast-enhanced T1-weighted images in the early stage [1, 14]. CT-guided core biopsy of the para-aortic mass is useful for differentiating between the benign form and the malignant form of RPF. Prompt diagnosis and treatment of most benign forms of RPT usually lead to a good prognosis. Corticosteroids are considered the first line of treatment for patients with idiopathic RPF, while Tamoxifen or other immunosuppressive drugs can be used as steroid-sparing agents or in steroid-refractory cases. Surgery is reserved for refractory cases, and the standard surgical approach involves open biopsy, ureterolysis, and ureteral transposition with omental wrapping of the involved ureter [17, 18]. Surgery is reserved for refractory cases. The standard surgical approach involves open biopsy, ureterolysis, and ureteral transposition with omental wrapping of the involved ureter. Effective management of secondary forms of RPF requires an approach based on the cause, when identified [8].

CONCLUSION

Recent advances in knowledge of RPF have led to the use of multidetector CT and MRI as the mainstay of noninvasive diagnosis. These imaging modalities allow for a comprehensive evaluation of the morphology, location, and extent of RPF and involvement of adjacent organs.

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