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DIFFICULTY IN DIAGNOSING RETROPERITONEAL FIBROSIS

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ABSTRACT

The retroperitoneal fibrosis (RPF) is a rare disease with multiple etiologies. The management of this entity is not standardized. We report a case illustrating the diagnostic difficulties of this disease. A 21 year-old male patient presented with persistent fever and poor general condition without associated signs. There was a biological inflammatory syndrome. A urinary test strip showed the presence of hematuria and proteinuria. The infection inquiry was negative; as well as the immunological tests. A CT scan showed a retroperitoneal mass. The pathological result was consistent with an inflammatory pseudo tumor. Due to an unexplained proteinuria and hematuria renal biopsy was made and showed post-infectious glomerulonephritis. The patient was prescribed immunosuppressive therapy including anti-TNF therapy. After improvement, we noted a resurgence of fever, with biological inflamatory syndrome. There was an increase in the size of the tumor without any mass effect or intra thoracic extension. The surgical resection was undertaken and pathology findings reported a retroperitoneal fibrosis mass surrounding abdominal vessels with a peri lymph node involvement. This case illustrates the diagnostic and therapeutic difficulty of the RPF due to non specific clinical features and points out a differential diagnosis which is the inflammatory pseudo tumor.

KEYWORDS: retroperitoneal fibrosis, inflammatory pseudotumor, diagnosis.

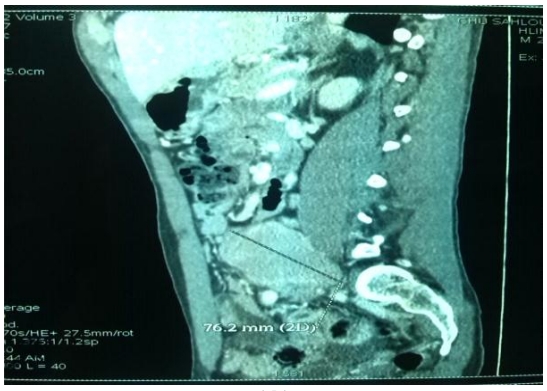
INTRODUCTION

Retroperitoneal fibrosis (RPF) is a rare disease characterized by the proliferation of fibroinflammatory tissue in the retro peritoneum that entraps the ureters, the abdominal aorta, the inferior vena cava and the iliac vessels.[1] It may have an idiopathic form or a secondary form, which may result from certain drugs (derivatives of ergot alkaloids), neoplasms, infections, radiation therapy, major trauma, major abdominal surgery and retroperitoneal hemorrhage, or urine leakage. The idiopathic form accounts for more than 70% of cases.[2] Men are affected twice to three times more often than women; the mean age at presentation is 50–60 years. Pediatric cases are rare, with up to 30 patients described in the literature.[3] Because of the non specific symptoms and signs of this disease and the lack of sensitive and specific laboratory tests and imaging, studies have played an important role in the diagnosis of RPF.

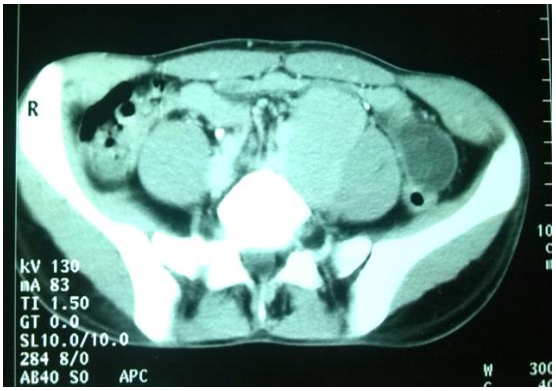
OBSERVATION

A 21 year-old male patient with no particular medical history presented with the main symptom of persistent fever with poor general condition without associated signs. There was a biological inflammatory syndrome. Liver and kidney functions were normal. A urinary strip test showed the presence of hematuria and proteinuria. The infection inquiry was negative (blood cultures, TST, sputum culture for TB, Wright serology test, card test, HIV, CMV, EBV, HBV, HCV and leishmaniasis serological tests).The transthoracic and transesophageal echocardiography as well as the myelogram showed no abnomalies. Serum complement levels were normal and the ANA serum levels were negative.

A CT scan showed a 50 * 31 mm irregular retroperitoneal soft tissue mass extending from the left iliac vessels along side the ipsilateral psoas muscle with left periaortic lymph nodes (Figure 1). Imaging of the liver, pancreas, spleen and kidneys was normal.



(A)



(B)

Figure 1: Sagittal (A) and axial (B) CT scan showing a mass extending from the left iliac vessels alongside the ipsilateral psoas muscle.

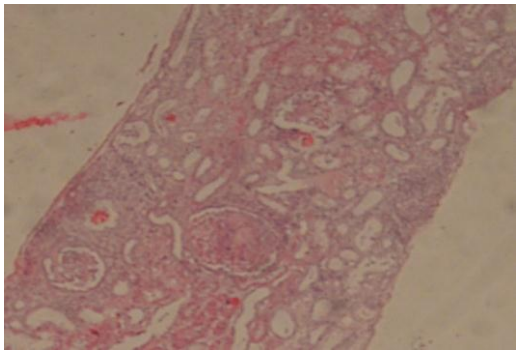
The mass's biopsy revealed a very suggestive histological appearance of a retroperitoneal inflammatory pseudotumor: It's a lymph node tissue formed by hyperplastic follicular structures with expanded reaction center surrounded by dilated inter follicular sinuses containing lymphocytes and plasma cells.

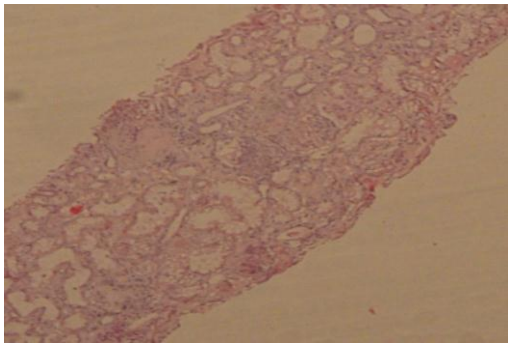
This lymph node joined a lesion formed by fibroblasts with abundant collagen tissue formation associated with lymphocytes, plasma cells and some eosinophils.

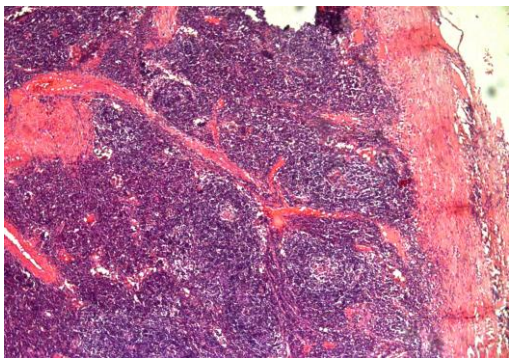
Corticosteroids and immunosuppressors (Azathioprine) were administred for 2 years which were then replaced by anti alpha-TNF therapy (infliximab) for 6 months due to the biological and clinical relapses (weight loss, BIS) with a stable CT scan aspect of the tumor.

The reappearance of the fever, the BIS as well as the recurrence of hematuria and proteinuria with impaired renal function marked the follow-up under anti TNF therapy.

The CT scan revealed an increase in the size of the tumor without mass effect or extension to the thoracic region.
The Surgical resection was undertaken and pathology findings reported a retroperitoneal fibrosis mass surrounding abdominal vessels with a peri lymph node involvement suggesting the diagnosis of retroperitoneal fibrosis (Figure2).







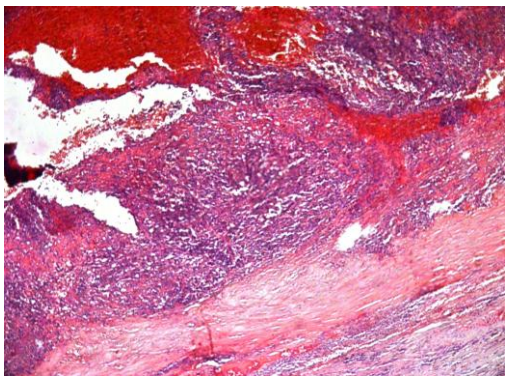


Figure 2: Histopatholgy of the retroperitoneal mass. Many lymphnode structures with a preserved architecture; some of those lymphnodes and the perinodal tissue are thickened by a high active fibrosis containing fibroblasts and numerous inflammatory cells (lymphocytes and mostly plasma cells). Cortiosteroids and immunosuppressors were withdrawn and there have been improvements in the follow-up(over the course of two years) including a preserved renal function.

DISCUSSION

All the forms of RPF (idiopathic, secondary, and IgG4-related) have similar clinical manifestations, thus their clinical presentation may not be of help in the differential diagnosis.

The initial symptoms and signs of RPF are often nonspecific, such as body weight loss, malaise, anorexia, poorly localized pain over the flank, lower back, or abdomen and low-grade fever.[4-8] As the disease progresses, obstructive uropathy and subsequent renal failure, lower extremity edema, deep vein thrombosis and claudication develop.[4,6] Renal failure due to a

bilateral ureteral obstruction is seen in about 42–95% of the cases; when the thoracic aorta or the peri-aortic arteries are involved, patients may suffer from hoarseness, secondary to recurrent laryngeal nerve paralysis, dry cough or upper limb claudication.

Laboratory findings reveal a high erythrocyte sedimentation rate, C-reactive protein level and anemia.[4-7] Because of the non specific 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. Multidetector CT and MRI have become the most useful tools in the diagnosis of RPF.[9] All patients with newly diagnosed RPF should undergo an age-appropriate cancer screening; in doubtful case, a tissue biopsy should be performed.

The microscopic observation of idiopathic RPF samples reveals the presence of two components: a fibrous tissue and an inflammatory infiltrate.[10]

In the absence of randomized trials, the treatment of idiopathic retroperitoneal fibrosis is empirical. Corticosteroids are the mainstay of therapy, other immunosuppressive drugs (mycophenolatemofetil, azathioprine).[11] have been used successfully together with corticosteroids, but the superiority of these combinations over corticosteroids alone is still unproven.[12]

When a contraindication to corticosteroids therapy is present, a good alternative maybe the use of tamoxifen. Recently, there have been some reports of a successful use of biologic agents in case of refractory idiopathic RPF (rituximab, infliximab).

The pseudo inflammatory tumor, also called myofibroblastic inflammatory tumor, are a heterogeneous group of lesions that can affect multiple organs especially the lungs, which is the most common site, liver, spleen, pancreas, digestive tract, the mesentery, mediastinum, retroperitoneum, kidneys.

They are characterized by a chromosomal translocation involving the ALK gene (anaplastic lymphoma kinase) on chromosome 2p23. Simultaneous multifocality in several organs is rare.

Histologically, the inflammatory pseudotumor is as dense and diffuse infiltration of inflammatory cells, predominantly plasma cells within a fibrous tissue. The plasma cell population is polyclonal (Immunohistochemistry). There is no identifiable microorganism by staining or culturing. This explains the diagnosis difficulty with retroperitoneal fibrosis illustrated by the case of our patient.

Because of the nonspecific symptoms and signs of this disease and the lack of sensitive and specific laboratory tests, RPF poses a differential diagnosis dilemma especially with the Pseudo-infalammtory tumor.

We emphasize that low-grade fever and weight loss with elevated acute phase reactants should prompt an evaluation for retroperitoneal fibrosis.

CONFLICT OF INTEREST

The authors declare that there is no conflict of interest regarding the publication of this paper

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