

Case Report

Idiopathic retroperitoneal fibrosis presented as abdominal discomfort and low back pain (IgG4-related disease)

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We presented a 45 year-old male patient in a hospital, who had low back pain and non-specific abdominal discomfort for duration of 2 weeks. Abdominal Computed Tomography (CT) showed retroperitoneal Para aortic mass at lower aortic roots before bifurcation and elevated C-RP. Unfortunately, the patient refused to do biopsy, and has a history of amlodipine usage with a very high suspicion of idiopathic retroperitoneal fibrosis (IgG4-related disease).

Key words: Idiopathic retroperitoneal fibrosis, low back pain, abdominal discomfort.

INTRODUCTION

Idiopathic retroperitoneal fibrosis (IRF) is a disease characterized by fibrotic process at the retroperitoneal area around the aorta, and the fibrotic plaques entrap and gradually obstruct retroperitoneal structures such as the ureter, inferior vena cava and aorta.

IRF may present with lower back pain, renal failure, hypertension, deep vein thrombosis and other obstructive symptoms. It is a disease believed to have developed as a result of non-specific inflammatory processes mediated by varying degrees of immune reaction. About 15% of IRF patients have fibrotic processes in areas other than the retroperitoneal space, elevated erythrocyte sedimentation rate (ESR), hypergammaglobulinemia, autoimmune antibody, multiple antibody, and vasculitis, pericardial or pleural effusion are frequently found in IRF. The outlook is usually good, but, if not appropriately diagnosed or treated, the disease can cause severe complications such as end-stage renal failure.

The disease is named after John Kelso Ormond who rediscovered the condition in 1948 (Baker, 1988).

CASE REPORT

A 45 year-old male patient who is a known case of hypertension and hyperlipidemia complained of non-

specific abdominal discomfort with low back pain for 2 weeks and flank pain for several weeks. Pain was gradual at the onset, progressive in course with no aggravating or relieving factors, mild to moderate in severity, radiating to all abdominal region and with no history of trauma. The patient denied ever having any form of fever, weight loss, nausea and vomiting, malaise, polyuria, or anorexia. He also denied having any past history of abdominal injuries or any previous surgery. His drug history is: amlodipine and statin; he denied any other medication such as ergot, beta-blocker, methyl dopa, etc.

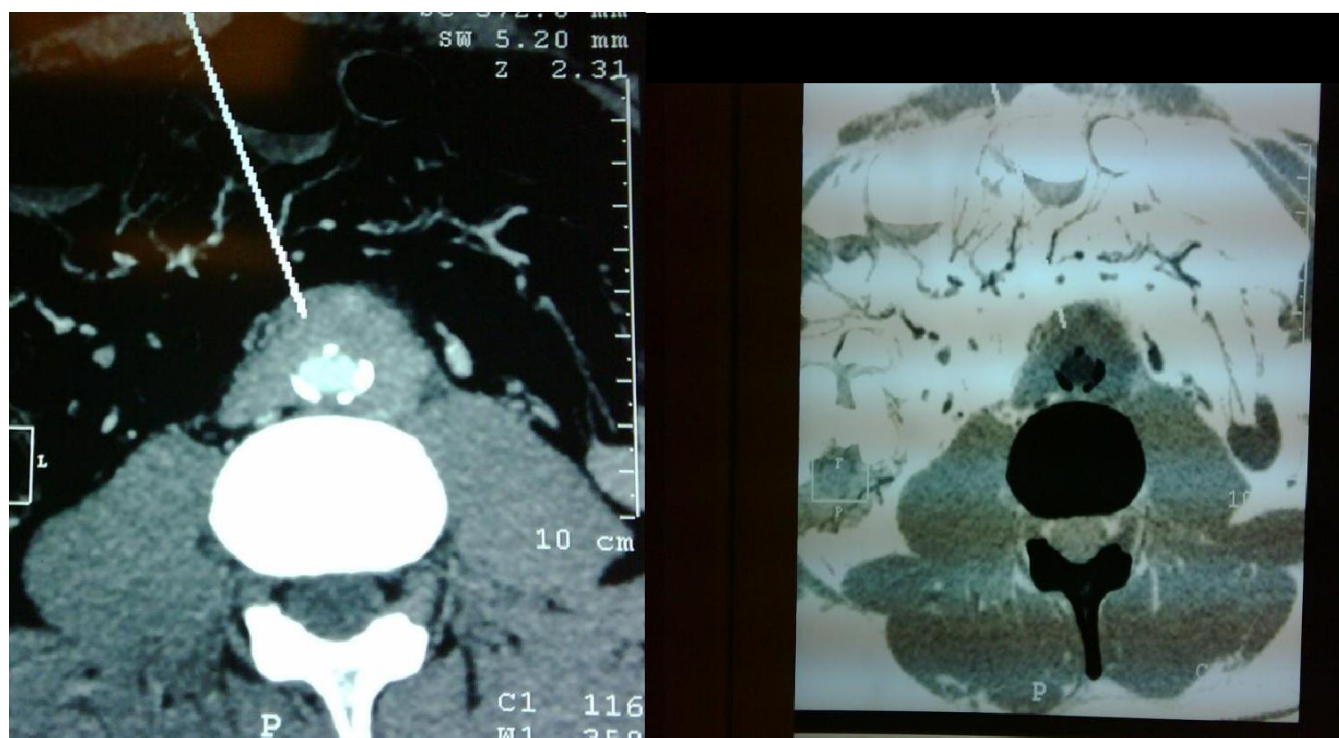
On admission the patient was slightly obese, weighing 75 kg; his blood pressure was 120/70, his temperature was 36.8°C. He was consciously alert, oriented of place, person and time, and physical examination showed nothing significant (cardiovascular, chest and abdominal examinations were normal). Laboratory data on the day of admission are listed in Table 1. He had an elevated C-reactive protein, but normal ESR.

Computed tomography (CT) of the colon (Figures 1, 2 and 3) showed retroperitoneal para-aortic mass at the lower aortic root before bifurcation. It also showed normal deferent part of colon, normal persacral space, no clear evidence of abnormal filling defect, and accidentally distal abdominal aorta with an enhanced soft circumferential structure. The patient was asked to do a biopsy to

Table 1. Laboratory data of the patient at the time of admission.

CBC	HB 14, 2 / WBC 6, 9 / Plat. 195
Bleeding profile	PT 12,7 / PTT 32 / INR 0.97
Chemistry	FBS 92, urea 34, creatinine 0.69, uric acid 5.4. Protein-total 7, 9, albumin 4, 4, globulin 3.5; Bilirubin- total 0.37. Bilirubin-direct 0.17; Alkaline phosphatase 108, ALT 67, AST 35, Na 140, K 3.8, Cl 104, albumin/globulin ratio 1.26
Lipid profile	Cholesterol 181, triglycerides 84, HDL 35. Cholesterol/HDL ratio 5.2 , LDL 129, LDL/HDL ratio 3, 69, VLDL 16,8
Serology	ESR 17, C- reactive protein 7.53
Urine analysis	Specific gravity 1.015, Ph 8, WBC 2/HPF, RBC 2/HPF

Abbreviations: CBC Common blood count, **WBC**; white blood count, **ESR**; erythrocyte sedimentation rate, **AST**; Aspartate aminotransferase, **ALT**; Alanine aminotransferase, **RBC**; Red blood cell, **HPF**; High power field, **VLDL**; Very low density lipoprotein, **LDL**; Low density lipoprotein, **HDL**; High density lipoprotein, **FBS**; fasting blood sugar, **HB**; hemoglobin, **PTT**; Partial thromboplastin time.

**Figure 1.** CT representation of retroperitoneal para-aortic mass at lower aortic root before bifurcation.

confirm the diagnosis of idiopathic retroperitoneal fibrosis (IgG4-RD) as both histological features and IgG4 immunostaining are critical to the diagnosis of IRF IgG4-RD. Unfortunately, though, after explaining pros and cons of the open biopsy, the patient decided not to do one. That notwithstanding, the case was treated as a case of retroperitoneal fibrosis based on the clinical and physical examination as well as the patient's abdominal CT (retroperitoneal mass surrounding the aorta, usual in IgG4-related retroperitoneal fibrosis), so the patient was

given 60 mg of Prednisalone and 20 mg of Mycophenolate mofetil daily.

The retroperitoneal mass was suppressed and the patient's low back pain was decreased, which confirmed our diagnosis of retroperitoneal fibrosis.

DISCUSSION

Retroperitoneal fibrosis is a slow but progressive



Figure 2. Sagittal CT representation of retroperitoneal para-aortic mass.

condition of unknown etiology, characterized by the deposition of fibrous tissue in the retroperitoneal space, which compress the ureters, great vessels and other structures. It is usually benign. Its incidence is 1: 200,000. There are of two types of retroperitoneal fibrosis (Stephen, 1991):

1. Primary (70% idiopathic) – male to female ratio is 3:1; onset is at age 30 to 60.
2. Secondary: 30% of the causes of the disease are related to malignancy, aortic aneurysm, post-irradiation therapy, foreign body reaction, and medications such as beta-blockers methysergide, lysergic acid diethylamide, methyl dopa, amphetamines, pergolide and cocaine.

Few studies have suggested that idiopathic RPF can be classified into cases that are associated with histopathologic features of IgG4-related disease (IgG4-RD) and those that are not IgG4-RD which account for a sizeable subset of patients with RPF. Both histological features and IgG4 immunostaining are critical to the diagnosis of IgG4-RD. Findings on plasma cell infiltration, tissue eosinophilia, and storiform fibrosis could lead

pathologists to perform IgG4 immunostains. The IgG4/IgG ratio may be more important in diagnosis than the absolute number of IgG4-positive plasma cells/HPF, particularly when extensive fibrosis is the paramount finding.

In this study, the patient most likely has idiopathic retroperitoneal fibrosis. Although we did not do a biopsy, the patient's clinical picture most likely represented the clinical picture of retroperitoneal fibrosis such as is common in 92% of cases of IRF present with pain, where the site of the pain may be in the flank, back, lower abdomen, or scrotum. In the patient used in the study, the pain was not associated with fever, weight loss, nausea, anorexia, malaise which if present favors malignancy (Wu, 2002).

The pain of IRF is usually relieved by Non-steroidal Anti-inflammatory Drugs (NSAIDs) due to the inflammatory nature of the fibrosis. Other symptoms of IRF are hypertension, obstructive renal symptoms such as polyuria, frequent hematuria and symptoms of vascular and venous compromise, inferior vena cava obstruction (IVC) obstruction, leg edema, deep vein thrombosis (DVT), etc.

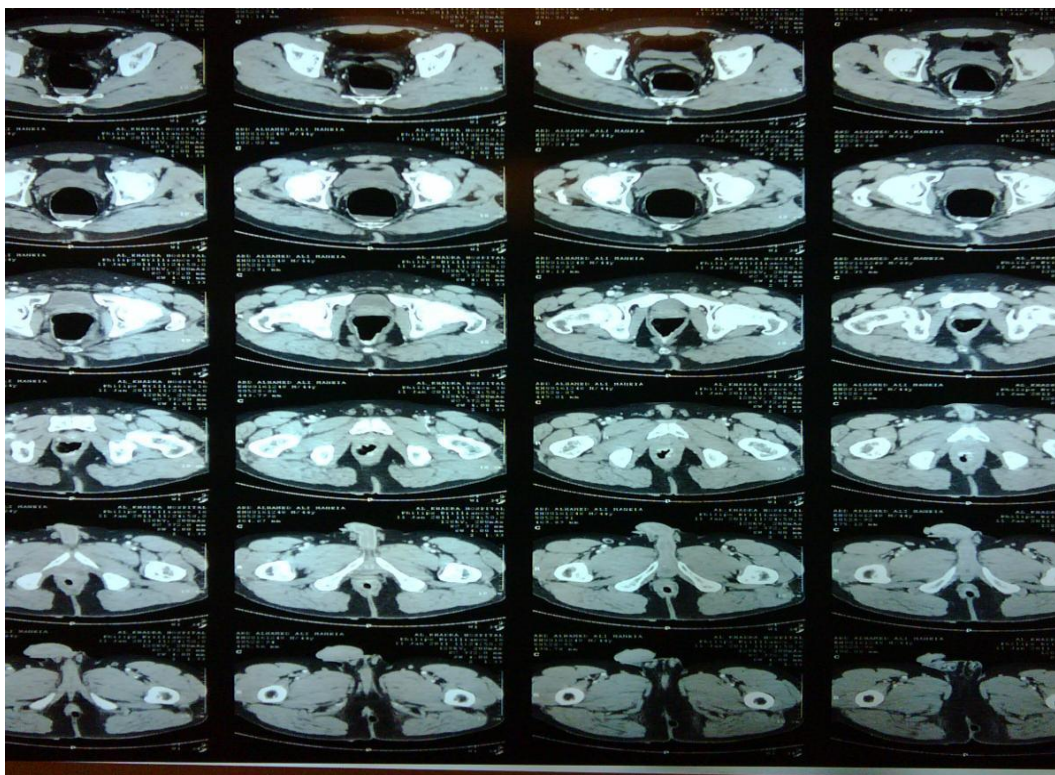


Figure 3. Representation of retroperitoneal para-aortic mass at lower aortic root before bifurcation.

The diagnosis of IRF was made by many steps. First, laboratory results may show high ESR, high C-reactive protein and increased bun/creatinine due to obstructive renal disease, but in this study, the patient had normal ESR but high C-reactive protein. The next step in the diagnosis is CT scan which is a test of choice used to visualize the extent of fibrosis, and to assess the presence of lymphadenopathy and tumor. The mass in retroperitoneal fibrosis may be bulky but not as massive as neoplastic lesions. The presence of enlarged mesenteric nodes and displacement of the aorta from the spine by the periaortic mass favors malignancy. Although some displacements can occur in retroperitoneal fibrosis, most retroperitoneal neoplasms displace the ureters laterally unlike retroperitoneal fibrosis, (Khan et al., 2004).

Magnetic resonance imaging (MRI)

This could be done and may show some findings comparable to those of CT scanning, but signal intensity is good for distinguishing retroperitoneal fibrosis from adjacent fat and psoas muscle. However, neither CT scan nor MRI can distinguish idiopathic retroperitoneal fibrosis from that due to tumor in all patients. Biopsy is used to differentiate the two processes and is recommended in all patients in whom there is no

apparent inciting agent.

Renal ultrasonography may be needed if one suspects any obstructive renal symptom which usually reveals a poorly margined, periaortic mass that may be associated with hydronephrosis. The treatment of idiopathic retroperitoneal fibrosis traditionally, has been surgical, but there may be an evolving medical component of stopping offending drugs – this may result in resolution of symptoms and complications (Tanagho, 1995).

Medical: Patients with systemic or an auto-immune disease may respond better to immunosuppression:

1. Glucocorticoids for months to years (usually high doses, but exact amount not yet established).
2. Glucocorticoids + azathioprine may shorten duration of overall treatment.
3. Glucocorticoids + mycophenolate mofetil (2 g/day) – case report showed regression in 1990s.
4. Tamoxifen – case reports have shown response but pathophysiology is not clear (thought to have promising results) (Diamond, 2004).

Surgical: Here urethral obstruction can be relieved via nephrostomy of tubes and urethral stents, transposition /transplantation of urethra, or wrapping of urethra in omental fat. Other obstructions can be relieved by de-

bulking mass, stenting vessel or by transplanting tissue.

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