Clinical Image

IDIOPATHIC RETROPERITONEAL FIBROSIS PRESENTED AS ABDOMINAL DISCOMFORT AND LOW BACK PAIN (ORMOND'S DISEASE)

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SUMMARY: We present 45 old male patient presented to the hospital with low back pain and non specific abdominal discomfort of 2 weeks duration, no urinary or other GI complication, no feature of GI or GU obstruction, abdominal CT show retroperitoneal Para aortic mass at lower aortic root before bifurcation and, elevated C-RP (unfortunately patient has refused to do biopsy), patient has history of amlodipine, patient has very high suspicion of idiopathic retroperitoneal fibrosis.

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 and around the aorta and the fibrotic plaque entraps and gradually obstruct retroperitoneal structures such as the ureter, inferior vena cava and aorta .

IRF may present with It may present with lower back pain, renal failure, hypertension, deep vein thrombosis and other obstructive symptoms.

IRF is believed to develop as consequence of non-specific inflammatory processes mediated by varying degree of immune reaction. About 15% of IRF patient have fibrotic process in area other than retroperitoneal space. elevated erythrocyte sedimentation rate (ESR), hypergammaglobulinema, autoimmune antibody, multiple antibody, multiple 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.

It is named after John Kelso Ormond, who rediscovered the condition in 1948.

CASE REPORT

45 years old male patient known case of hypertension and hyperlipedimia complain of non specific abdominal discomfort of 2 weeks ago and low back pain and flanks of several weeks duration, pain was gradual onset, progressive course, no increase or decrease factors, mild to moderate in severity, radiate to all abdominal region, no history of trauma, patient has denied any of fever, weight loss, nausea and vomiting, malaise,

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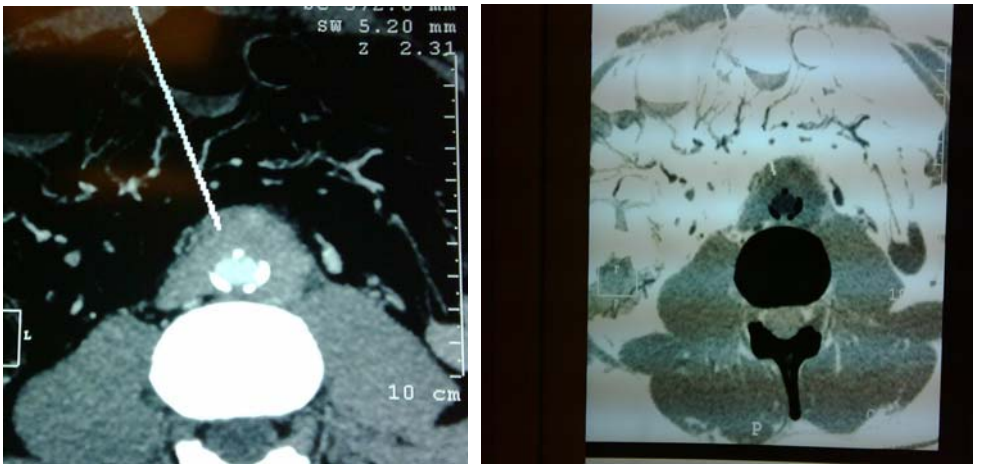

Figure 1: CT show retroperitoneal Para aortic mass at lower aortic root before bifurcation.

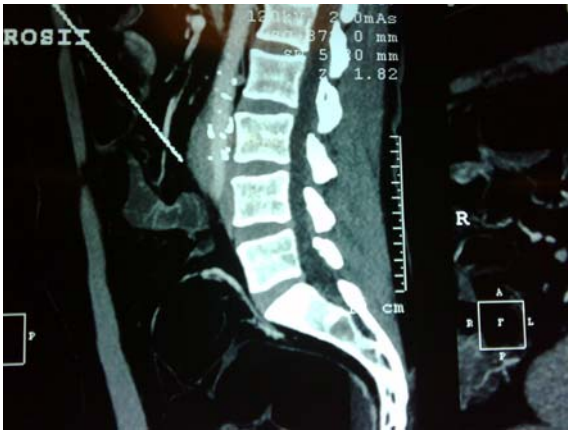

Figure 2: Sagital CT show retroperitoneal Para aortic mass.

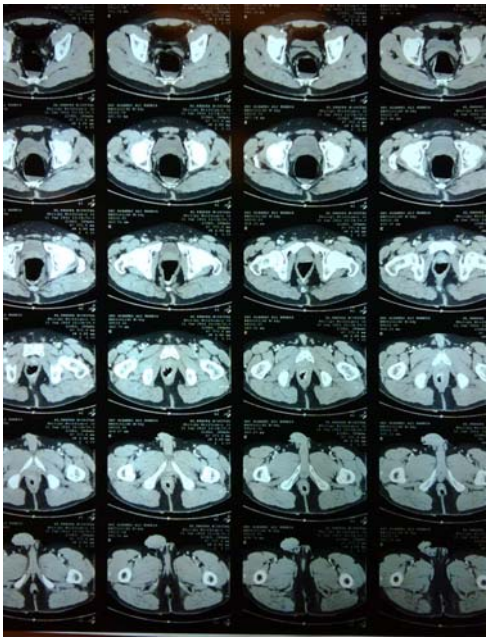

Figure 3: Whole colon CT.

polyuria, or anorexia, Patient also has denied any past history of abdominal injures, or any previous surgery, the patient Drug history is: amlodipine, statin, he denied any other medication as ergot, beta-blockers, methyl dopa etc.

On admission he was slightly obese 75 k , his blood pressure was 120|70, his temperature was 36.8°C, Patient was conscious, alert, oriented of place person and time and, physical examination was not significant (cardiovascular, chest and abdominal examination all was normal).

Laboratory data at day of admission where listed in Table 1, elevated C-reactive protein but normal ESR, otherwise is normal.

Computerized tomography (CT) of the colon Figures 1, 2 and 3 show retroperitoneal Para aortic mass at lower aortic root before bifurcation, CT also showed normal deferent part of colon, normal persacral space, no clear evidence of abnormal filling defect, and accidentally distal abdominal aorta soft enhanced circumferential structure.

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

Patient has non specific abdominal discomfort of 2 weeks duration and low back pain, no urinary or other GI complication, no feature of GI or GU obstruction, patient was asked to do a biopsy to confirm the diagnosed of idiopathic retroperitoneal fibrosis but unfortunately after explaining pones and cons of the open biopsy patient decided not to do one, however the case was treated as a case of retroperitoneal fibrosis based on the clinical and physical exanimation and abdominal CT patient, so the patient was given prednisalone 60 mg, Mycophenolate mofetil 20 mg daily mg.

The retroperitoneal mass has suppress and the patient low back pain has decrease, which confirm our diagnosis of retroperitoneal fibrosis.

Retroperitoneal fibrosis is a slowly progressive condition of unknown etiology, characterized by deposition of fibrous tissue in the retroperitoneal space compressing the ureters, great vessels, and other structures. Its usually benign. The Incidence - 1:200,000, there is two types.

- Primary -- ~70% idiopathic - male:female ratio 3:1, onset at age 30-60

- Secondary -- ~30% some causes are Malignancy, Aortic aneurysm, Post-irradiation therapy, Foreign body reaction, Medications as Beta-blockers Methysergide, Lysergic acid diethylamide, Methyldopa, Amphetamines, Pergolide, Cocaine.

In our case the patient most likely have idiopathic retroperitoneal fibrosis's although we didn't do a biopsy but the patient clinical picture most likely represent the clinical picture of retroperitoneal fibrosis as in 92% of cases of IRF it present with pain but the site of this pain maybe seen in the - flank, back, lower abdomen, and scrotum in our patient the pain has no other association as fever, weight loss, nausea, anorexia, malaise which if present it favors malignancy, Pain of IRF is usually relieved by NSAIDs due to inflammatory nature of the fibrosis.

Other symptom of IRF symptom is Hypertension, obstructive renal Symptoms as polyuria, frequency, hematuria from and symptom of Vascular compromise - venous -- IVC obstruction LE edema, dvt etc..

The diagnosis of IRF is made by many steps first laboratory may show high ESR, and high C-reactive protein and may show increased bun/creatinine due to obstructive renal desiese , but in our case pt had normal ESR but high C-reactive protein.

Next step in mangment is CT scan which is the test of choice 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 displacement can occur in Retroperitoneal fibrosis, Unlike Retroperitoneal fibrosis, most retroperitoneal neoplasms displace the ureters laterally.

Magnetic resonance imaging (MRI) could be done which may show some findings comparable to those with 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 ordrde if u suspect any obstructive reanal symptom usually reveals a poorly marginated, periaortic may be associated with hydronephrosis.

The treatment of idiopathic retroperitoneal fibrosis traditionally has been surgical, but there may be an evolving medical component.

Stop offending drugs - this may result in resolution of symptoms and complications

Medical:

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

1. Glucocorticoids for months to years (high dose usually, but exact amount not estab. yet)

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).

Surgical:

Relieve ureter obstruction - via nephrostomy tubes and ureteral stents.

Or via transposition/transplantation of ureters

Or via wrapping ureters in omental fat.

Relieve other obstruction - by debulking mass, stenting vessel, or transplanting tissue.

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Editor's note: *Recently an article appeared "Vaglio A, Palmisono A, Alberici F, et al. Prednisone versus tamoxifene in patients with idiopathic retroperitoneal fibrosis: an open-label randomised controlled trial. Lancet, 378:338-346, 2011." about the clinical approach to the patients with idiopathic retroperitoneal fibrosis.*