Retroperitoneal fibrosis: Radiological-pathological features

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A 42 years old male presented to a general physician with complaints of progressive symptoms of lower back ache, fatigue, anaemia, weight loss and mild fever. Initial diagnosis on clinical grounds was abdominal tuberculosis but on ultrasonography (USG), a hypoechoic mass encircling the aorta, inferior vena cava (IVC) and ureters at the aortic bifurcation was observed; with associated mild bilateral hydroureteronephrosis. His C-reactive protein (CRP) and erythrocyte sedimentation rate (ESR) level was raised. Retroperitoneal fibrosis (RPF) was suspected and a contrast enhanced computerised tomography (CT) scan was performed. To differentiate between retroperitoneal fibrosis and lymphoma, CT guided biopsy was carried out and it confirmed our top impression on USG and CT of retroperitoneal fibrosis. Histology examination showed infiltration of plasma cells, macrophages, lymphocytes and eosinophils accompanied by fibrosis.

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INTRODUCTION

The French urologist Albarran first described Retroperitoneal fibrosis (RPF) in 1905, as ureteral obstruction secondary to fibrotic changes in the retroperitoneal space but with Ormond's publication in 1948, the disease became an established clinical entity (Vaglio et al., 2006). RPF is a rare disease which can be idiopathic or secondary to some pre-existing factor and is characterized by the presence of chronic inflammation and marked fibrosis of retroperitoneal tissue. It commonly entraps the ureters, great vessels and other abdominal organs at the lower lumbar region (Figure 1). Ureretal involvement is reported in 80-100% of cases at presentation. This condition may presents low back and/or abdominal pain with or without flank tenderness (Albarran, 1905; Ormond, 1948).

CASE PRESENTATION

A 42 years old male presented with lower back ache, fatigue, weight loss, general malaise and mild fever. He was referred to our department for Ultrasonography (USG) to rule out any abdominal evidence of common disease in our society. On scanning, a bilateral mild to moderate degree of hydroureteronephrosis was observed and at the level of aortic bifurcation, a well-defined smooth-marginated, hypoechoic retroperitoneal soft-tissue mass encasing the aorta and inferior vena cava (IVC) was captioned (Figure 2). Distal extension beyond the sacral promontory and the absence of lobulation suggested a

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Figure 1. Typical location of the retroperitoneal fibrosis. (A) Coronal view: Note that plague usually does not extend to lateral borders of psoas muscles. (B) Transverse view: Great vessels are enveloped but not elevated from the spine.

Figure 2. Hypoechoic rind of soft tissue mass encircling the aorta and IVC.

benign etiology. To further confirm our impression of RPF, computerized tomography (CT) scan was advised. On contrast enhanced computed tomography (CECT) an isodense (isodense to muscles) homogenous mass was observed without any vascularity (Figure 3). We were sure on our USG suspicion but on the request of referring physician CT guided biopsy was carried out to make the final diagnosis on histo-pathological grounds to further differentiate between lymphoma and RPF. Histopathology report confirmed our diagnosis of RPF. The patient was referred to urologist because of progressive renal failure and increasing renal parameters (blood urea and creatinine) due to bilateral ureteric obstruction. Double-J-stents were advised to relieve
obstruction. Initially intravenous (IV) and then oral steroids were administered; and the patient gained dramatic recovery with regression of the retroperitoneal mass. He was now on follow-up visits to our department and was still on low dose steroids.

DISCUSSION

RPF is a rare inflammatory disease without an obvious etiology and hard to diagnose because of its non-specific symptoms. RPF is characterized by replacement of the normal tissue of the retroperitonium with fibrosis and or chronic inflammation. However, etiology, clinical and radiological presentation varies in many cases. Up to 15% of patients have additional fibrotic process outside the retroperitonium. Macroscopically, RPF appears as a grey–white fibrous plaque usually arising between the level of aorta and iliac arteries in the retroperitoneal space. Microscopically at an early stage, mixtures of eosinophils, polymorphonuclear leukocytes and other inflammatory cells is present within a matrix of collagen (Figure 4). As the disease progress, the process can become cellular with diffuse fibrosis as the only findings (Warakaulle et al., 2004; Tang et al., 2002). Various terms have been used to designate this disorder, including Ormond's disease, periureteritis fibrosa, periureteritis plastica, chronic periureteritis,
sclerosing retroperitoneal granuloma and fibrous retroperitonitis (Tang et al., 2002). In recent years, the condition has been known as retroperitoneal fibrosis, a term that more closely describes the actual pathologic process. Etiologically, retroperitoneal fibrosis is classified as a primary idiopathic disease or as a disease secondary to known causes. All identifiable causes of fibrosis must be excluded before the process can be considered idiopathic.

Primary or idiopathic retroperitoneal fibrosis

This is considered to be autoimmune in response to ceroid (an insoluble lipid) that can leak through arterial walls from atherosclerotic plaques and leads to a vasculitis.

1. Associations with other autoimmune mechanism have been suggested, like primary biliary cirrhosis, fibrosing mediastinitis, glomerulonephritis, rheumatoid arthritis, ankylosing spondylitis, polyarteritis nodosa, systemic lupus erythematosus (SLE) and hashimoto thyroiditis (LeBlanc et al., 2002).

Secondary retroperitoneal fibrosis

1. Drugs like methysergide, beta-adrenergic blockers, lysergic acid diethylamide (LSD), methyldopa, amphetamines, phenacetin, hydralazine and cocaine have also been suggested.
2. Desmoplastic response to malignancy like lymphoma, carcinoid and retroperitoneal metastases (breast, lung, thyroid, GI tract, GU organs).
3. Retroperitoneal fluid collection as in trauma, surgery or infection.
4. Aneurysm of the aorta or iliac arteries (desmoplastic response).
5. Radiation therapy (LeBlanc et al., 2002; Wolfgang, 2011).

Imaging features

Ultrasonography and CT scanning of the abdomen have major role in initial evaluation, diagnosis and assessing the response to therapy of retroperitoneal fibrosis. However, neither the appearance of the mass nor localization permits definitive differentiation between benign lesions and malignancies such as lymphoma, sarcoma or metastatic adenopathy. Thus the invasive radiological procedure like CT guided biopsies of the mass can be very helpful but still in some cases the diagnosis is not firmly established until surgical exploration is performed.

On intravenous urogram (IVU)

1. Medial deviation of the ureter beginning at the level of the lumbar three vertebra (L3) or lumbar four vertebra (L4) with gradual tapering of the ureters distal to mass with proximal hydronephrosis is most commonly observed, though it is not a constant feature (Wolfgang, 2011).

On Computerized tomography (CT) scan

1. Rind of soft tissue around aorta and inferior vena cava between level of kidney and sacrum.
2. Spreads to involve the ureters, causing varying degrees of obstruction.
3. Fat plane between the mass and the psoas muscle may be obliterated.
4. Unlike adenopathy, RPF tends not to displace aorta anteriorly.
5. Mass may show varying degrees of enhancement depending on the stage of the disease.

Differentiating benign versus malignant masses

1. Mass in RPF is less bulky than most neoplastic lesions
2. Malignancy produces enlarged mesenteric nodes and displacement of the aorta from the spine
3. Most retroperitoneal neoplasms displace the ureters laterally (LeBlanc et al., 2002; Wolfgang, 2011).

Ultrasonography (USG)

1. Hypoechoic homogeneous mass.

Magnetic resonance imaging (MRI)

1. Low to medium homogeneous signal intensity on thoracic one vertebra (T1)
2. Heterogeneous high signal intensity on thoracic two vertebra (T2) (inflammatory stage)
3. Low signal intensity on T2 (dense fibrotic stage).

Positron-emission tomography (PET)/Nuclear scan

(1) Gallium uptake during active inflammation
(2) Increased para-aortic uptake of 18F-FDG, consistent with active inflammation of retroperitoneal fibrosis. Bilateral ureteral obstruction can also be observed on PET image 7.

Treatment

Treatment of primary retroperitoneal fibrosis consists of
relieving ureteral obstruction, restoring renal function and avoiding infection. This is achieved by stent insertion and administration of steroids. If steroids do not respond, then anti-estrogen and immunosuppressive therapy should be considered. Even in some cases, surgical intervention may be required. Treatment of malignant RPF is dependent on the primary tumor (Dedeoglu et al., 2001; Kardar et al., 2002)

Conclusion

Immediate diagnosis of RPF is vital for preserving renal function, avoidance of hydroceles or edema due to obstruction or compression of the retroperitoneal vessels and vena cava. Differentiating from conditions that mimic RPF symptoms such as retroperitoneal abscess, infections and malignancies is important for appropriate treatment (Aziz et al., 2006). Although ultrasonography is the first diagnostic modality, it is not the best imaging modality for assessing RPF. Unilateral or bilateral hydronephrosis, a hypoechoic and well-defined mass are characteristics of RPF on ultrasonography. Assessing the periaortic region may be difficult, particularly in obese cases and unfilled bowel loops by gas or fluid that may result in misdiagnosis. Contrast-enhancement of soft tissues around the infrarenal aorta or iliac vessels without aneurysmal evidence of the infrarenal aorta on CT scan and no blood signals are diagnostic for RPF (Aziz et al., 2006; Rottenberg and Sandhu, 2008).

REFERENCES
