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Pelvic confined idiopathic retroperitoneal fibrosis mimicking a large tumor

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ABSTRACT

Idiopathic retroperitoneal fibrosis (IRF) entirely confined to the pelvic cavity is an extremely rare clinical entity. Herein, is described the case of a 36- year old male who presented with clinical and imaging manifestations of a large pelvic tumor. Exploratory laparotomy revealed a large mass in the right pelvis originating from the retroperitoneal space, displacing the right iliac vessels, the right ureter and the urinary bladder completely to the left. A laborious resection of the mass measuring 14x10cm was performed. Histopathological examination and detailed immunohistochemistry analysis were suggestive of idiopathic retroperitoneal fibrosis with no evidence of malignancy. This is a very rare case regarding localization and clinical presentation of idiopathic retroperitoneal fibrosis. We conclude that IRF should be included in the differential diagnosis of patients presenting with a pelvic mass even if there is no involvement of the typical para aortic area.

Key words: retroperitoneal fibrosis, idiopathic, pelvis, Ormond's disease, pelvic tumor

INTRODUCTION

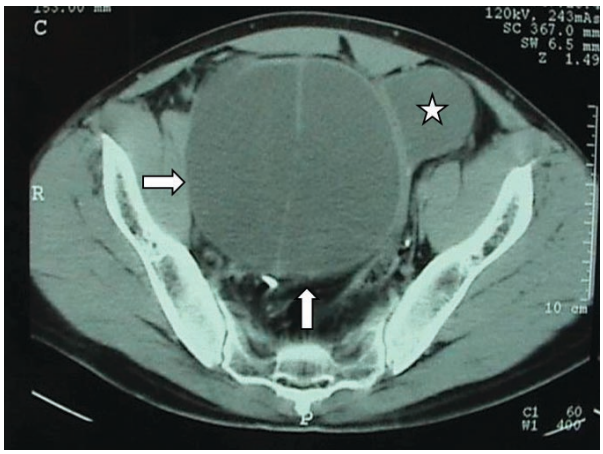
Retroperitoneal fibrosis (RPF) is an uncommon but well described clinical entity, characterized by the replacement of the normal retroperitoneal tissue with fibrosis and / or chronic non specific inflammation [1]. Idiopathic retroperitoneal fibrosis (IRF) localized entirely to the pelvis without involvement of the typical para-aortic area is extremely uncommon. We herein present a case of a pelvic idiopathic retroperitoneal fibrosis in a 36-year old male who presented with clinical and imaging manifestations of a large pelvic mass.

CASE PRESENTATION

A 36-year old man presented with a non tender pelvic mass which he first noticed a week ago. His past medical history was remarkable for nephrolithiasis, but he had not undergone any previous urological procedure. On physical examination he was afebrile, whereas a large firm non tender right pelvic mass measuring approximately 15 cm was palpated with well defined margins. Laboratory data revealed a mild leukocytosis of 10060cells/mm3 with 64% neutrophils, a hematocrit of 44% and a platelet count of 148x10^3/ml. The erythrocyte sedimentation rate was elevated at 48mm/h. Biochemical investigation including serum electrolytes, urea, creatinine and liver function tests were normal. Serum concentrations of carcinoembryonic antigen (CEA), carbohydrate antigen (CA19-9) and alpha fetoprotein were within normal ranges as well.

Contrast enhanced computed tomography (CT) scans revealed the presence of a large mass measuring 14 cm in the right pelvis, arising from the retroperitoneal space completely displacing the urine bladder to the left *(Fig 1)*. No ureter obstruction or hydronephrosis were demonstrated. During exploratory laparotomy, a large thick-walled firm mass measuring 14 cm was found in the lesser pelvis, originating from the retroperitoneal space. The right iliac vessels, right ureter, and the urinary bladder were all completely displaced to the left, but they were not involved in the mass. Intraoperative frozen section of multiple specimens, and tissue imprints showed no evidence of malignancy. A difficult and laborious resection of the whole mass was performed and the ureter was repositioned

Figure 1. **Contrast enhanced computed tomography scan of the pelvis demonstrating a large pelvic tumor with well defined enhanced borders (arrows), displacing urinary bladder completely to the left (star).**



laterally at its anatomic site. Thorough histopathological examination and detailed immunohistochemistry analysis of the whole resected specimen were suggestive of idiopathic retroperitoneal fibrosis with no evidence of malignancy. The postoperative course was uneventful and the patient was discharged 5 days later. He is being followed up every three months. At last follow-up examinations, performed 18 months after the surgical intervention, he was well and asymptomatic without any evidence of recurrence.

DISCUSSION

The incidence of RPF is approximately 1:200000 and predominately occurs in men in the fifth and sixth, decades of life [2]. However, several cases have been reported in children [3]. About two-thirds of all cases are considered IRF whereas the remaining one third has been associated with the use of various medications, surgery, hemorrhage, infections and abdominal aortic aneurysms [4]. Approximately in 8-10% of the cases fibrosis is the result of the infiltration of the retroperitoneal space by metastatic tumors from digestive track, breast, prostate, kidneys, lymphomas and sarcomas [1]. RPF is characterized by the presence of a fibrous dense plaque in the retroperitoneum which starts at the aortic bifurcation and gradually envelops the aorta, inferior vena cava and ureters [2]. Up to 10% of the patients have additional extraperitoneal disease and may present as mediastinal fibrosis, Riedel fibrosing thyroiditis, sclerosing cholangitis and orbital fibrous pseudotumors [4,5].

Signs and symptoms of RPF are non specific, often resulting in a delayed diagnosis and significant morbidity due to progressive loss of renal function [2]. The duration of symptoms prior to diagnosis is four to six months and sometimes more than a year [3]. The most common presenting symptom is a dull poorly localized back abdominal or flank pain whereas other frequent symptoms are weight loss, anorexia, fatigue, anemia and fever [1,5].

Our patient was admitted with an extremely rare presentation. There were no clinical symptoms or signs except the palpable pelvic mass, whereas the disease was entirely confined to the pelvis without involvement of the typical para-aortic area. Both clinical and imaging findings were suggestive of a large pelvic tumor. We were able to find only 14 cases with a pelvic confined retroperitoneal fibrosis. Signs and symptoms in this patients group are different than those of a para-aortic involvement but imaging findings are almost similar.

Computed tomography scans and magnetic resonance imaging are the diagnostic modalities of choice in the diagnosis and follow-up of the patients with RPF [1]. The most common finding is a periaortic soft tissue mass that envelops the aorta and the inferior vena cava between the renal hila and the sacral promontory [2,5]. CT is also used to differentiate RPF from retroperitoneal hemorrhage, primary retroperitoneal sarcomas and metastatic tumors in the region [5]. Magnetic resonance imaging additionally provides excellent assessment of the extent and does not require iodinated contrast material [1].

The goal of treatment in patients with RPF is to relieve the ureteral obstruction and restore the renal function. Additionally, any possibility of malignancy should be ruled out and preventive measures are needed against the progression of the inflammatory process [2]. There are two main therapeutic approaches for RPF [6]. The first consists of open or laparoscopic ureterolysis with or without omental wrapping of the ureter, followed or not by corticosteroids. The second approach consists of ureteric stents placement followed by corticosteroids alone or together with immunosuppressive medication using azathioprine or tamoxifen. Conservative management of RPF in means of relief of ureteric obstruction with or without steroids is effective and is curative in up to two thirds of the patients [6]. Prognosis is excellent and long term success rates exceed 90% in patients without renal compromise and effective ureterolysis, but unfortunately in cases with a malignant fibrosis a poor median survival of 3-6 months has been reported [1]. Long and careful follow-up is mandatory since recurrences of the disease are often asymptomatic [5].

Due to the extremely rare presentation of our patient, in terms of clinical symptoms and location of the disease, the diagnosis of retroperitoneal fibrosis was not clear before surgery. Otherwise, the approach might have been different including image guided fine needle aspiration biopsy of the mass and subsequent conservative treatment. However, the possibility of a malignancy could not be ruled out with certainty without complete surgical resection, in this case of a large pelvic mass.

CONCLUSIONS

We report a very rare case of idiopathic retroperitoneal fibrosis entirely confined to the pelvis, in a young patient who presented with manifestations of a large pelvic tumor. Although extremely rare, IRF should be included in the differential diagnosis of patients presenting with a pelvic mass even if there is no involvement of the typical para aortic area.

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