Idiopathic Retroperitoneal Fibrosis

Candace Derenge, MD

INTRODUCTION

Idiopathic retroperitoneal fibrosis (IRF) is a rare and unusual etiology of abdominal pain that is becoming more frequently diagnosed with the use of CT imaging for abdominal pain. Retroperitoneal fibrosis is characterized by the development of extensive fibrosis throughout the retroperitoneum that can encircle the aorta, iliac arteries, and ureters. About two thirds of retroperitoneal fibrosis cases are idiopathic, thought to be due to an autoimmune process of antibodies stimulating desmoplastic reaction. One third of cases are secondary to drugs (methysergide, beta-blockers, hydralazine, ergotamine, LSD) or diseases the stimulate desmoplastic reaction (tumors, infections, radiation, and Erdheim-Chester disease). Histology of IRF shows abundant fibrosis and chronic inflammation. There is limited data in the literature regarding idiopathic retroperitoneal fibrosis.

CASE PRESENTATION

A 50 yo African-American man presented for several months of severe abdominal pain wrapping around his flanks, associated with chills and 30-pound unintentional weight loss. He was not taking any medications, had no clinical signs of infection, and had no history of malignancy or radiation. On exam he had left-sided abdominal tenderness. His ESR was elevated to 114 with a normal CRP. A non-contrast CT abdomen to identify possible malignancy showed infiltrative peritoneal soft tissue encasing the common iliac artery, common iliac vein bifurcation, and the left ureter, causing mild left hydronephrosis.

DIFFERENTIAL DIAGNOSIS

This diagnosis of idiopathic RPF is made on the basis of CT or MRI and upon exclusion of secondary retroperitoneal fibrosis. Secondary causes of retroperitoneal fibrosis were excluded with negative screening for TB, hepatitis B, hepatitis C, HIV, IgG 4 and syphilis, and biopsy showing fibroblastic proliferation with inflammation.

OUTCOME & FOLLOW-UP

The patient was started on high-dose steroids (1mg/kg prednisone daily for 4 weeks), which significantly improved his pain.

DISCUSSION

Idiopathic retroperitoneal fibrosis is a rare diagnosis of exclusion, and our patient provides a classic example. Retroperitoneal fibrosis is 2-3x more common in men and the mean age at onset is 55 to 60. Clinical presentation is back, abdominal, and/or testicular pain, systemic symptoms (e.g. fatigue, weight loss, chills), oliguria from obstructive uropathy, resultant volume overload, and/or claudication. Flank and abdominal pain is the most common symptom of idiopathic retroperitoneal fibrosis, and systemic symptoms are the second most common. Diagnosis of retroperitoneal fibrosis is made on CT or MRI of the abdomen, and diagnosis of idiopathic retroperitoneal fibrosis is made through excluding possibly etiologies through bloodwork and biopsy..

KEY POINTS

Idiopathic retroperitoneal fibrosis is a rare cause of abdominal pain, and it is a diagnosis of exclusion.

REFERENCES

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