

# Image of the month: Ormond's disease: A diagnostic challenge

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## ABSTRACT

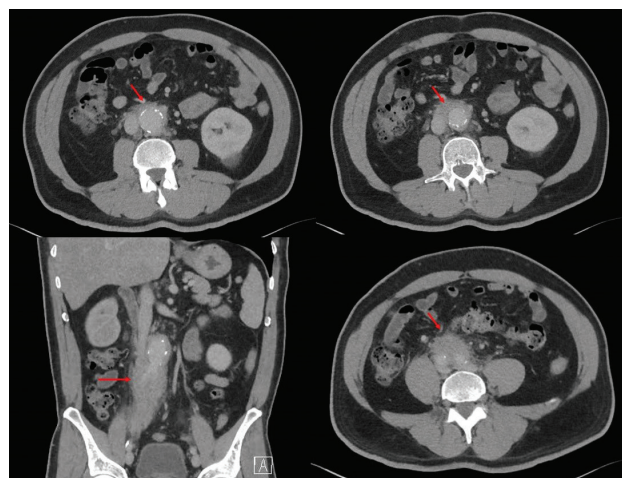
Idiopathic retroperitoneal fibrosis is a rare disease. Symptoms include constitutional symptoms and abdominal/flank pain. Causes of retroperitoneal fibrosis include non-infectious and infectious etiologies. Of the infectious etiologies, tuberculous aortitis is an important differential as it is associated with a high mortality rate. We present a case of a 59-year-old man with a retroperitoneal periaortic mass initially diagnosed as tuberculous aortitis. However following biopsy, the diagnosis was later amended to idiopathic retroperitoneal fibrosis with latent tuberculosis. He was successfully treated with prednisone and methotrexate.

**KEYWORDS:** Tuberculous aortitis, Ormond's disease, idiopathic retroperitoneal fibrosis

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## Case presentation

A 59-year-old man with a known infrarenal abdominal aortic aneurysm and significant smoking history, presented with a 3-month history of dry cough, lethargy, night sweats and 4 kg weight loss. He was pyrexial on admission. Physical examination was unremarkable. Laboratory tests demonstrated an elevated C-reactive protein (42 mg/L), creatinine of 152 µmol/L (baseline 80), QuantiFERON-TB Gold was positive and serum immunoglobulin G4 (IgG4) was elevated at 2.6 g/L (range 0.03–2.01). Whole body computed tomography showed a retroperitoneal mass extending around the infrarenal abdominal aorta (Fig 1), obstructing the right ureter with hydronephrosis, lymphadenopathy adjacent to the mass and a number of lung granulomas. Core biopsy of the mass demonstrated a mixed inflammatory infiltrate comprising macrophages, plasma cells, lymphocytes, neutrophils and a small number of eosinophils, with no evidence of lymphoma, IgG or sclerosing disease. A provisional diagnosis of tuberculous aortitis (TA) was made and he was started on isoniazid, rifampicin, pyrazinamide, ethambutol and 60 mg of



**Fig 1.** Unenhanced computed tomography of the abdomen demonstrating periaortic mass involving the abdominal aorta and iliac arteries.

prednisone (dosed at 1 mg/kg/day). A ureteric stent was inserted to relieve the right renal tract obstruction. Treatment led to a resolution of his constitutional symptoms, reduction in C-reactive protein and creatinine, and size of the para-aortic mass on imaging at 6 weeks. Two months later, the biopsy culture was negative and so the diagnosis was thought to be more in keeping with idiopathic retroperitoneal fibrosis (IRPF), also known as Ormond's disease. IgG4 disease was felt to be unlikely, as there were no features of IgG4-related disease on the biopsy. The patient completed 4 months of antituberculous drugs for latent tuberculosis. Methotrexate (20 mg weekly) was started for treatment of IRPF and the prednisone was weaned by 5 mg every month.

## Discussion

Causes of retroperitoneal periaortic masses include infection (eg histoplasmosis, TA), malignancy (eg lymphoma), sarcoidosis, previous radiotherapy and previous surgery.<sup>1</sup> IRPF is a rare disease, tending to affect males in their fifth decade. The adventitia of the abdominal aorta, iliac arteries and, less commonly, the thoracic aorta can be involved. Symptoms include fevers, weight loss, and pain in the flank, back or abdomen. Ureteral involvement due to ureteral deviation or obstruction is a common complication. C-reactive protein and erythrocyte sedimentation rate is

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usually elevated and can be used to monitor disease activity. Pathologically, the inflammatory infiltrate consists of lymphocytes, plasma cells and macrophages. First-line therapy is glucocorticoid, dosed at 0.75–1 mg/kg/day with gradual tapering to 5–7.5 mg/day over 6–9 months. Response is rapid with remission rates of 75–95% after glucocorticoid therapy.<sup>2</sup> Cyclophosphamide, mycophenolate mofetil and methotrexate have been used effectively as well. Relapse is common and occurs in up to 72%, therefore monitoring with laboratory markers and imaging is required.<sup>3</sup>

TA should also be considered as a differential diagnosis as it carries a mortality rate of up to 50%.<sup>4</sup> It is often difficult to diagnose due to possible inconclusive culture, histology and blood markers. Treatment is with oral antituberculous drugs for 6 months although surgery may sometimes be required.

## Conclusion

IRPF presents with constitutional symptoms and is a rare cause of retroperitoneal fibrosis. TA should also be considered in the presence of high-risk features, such as immigration/travel from

an tuberculosis endemic area, immunosuppression etc as it is associated with significant morbidity and mortality. Both IRPF and TA can be effectively treated. ■

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