Retroperitoneal Fibrosis

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A 50-year-old man with type 2 diabetes mellitus, nonalcoholic fatty liver disease, and sarcoidosis presented to the emergency department with lower abdominal pain for the past 3 months. Physical examination revealed suprapubic discomfort, back pain, stress-induced incontinence, abdominal distention, and scrotal swelling. A computed tomographic angiogram of the abdomen and pelvis showed progressive, circumferential, periaortic internal iliac soft tissue swelling (image A, arrow). Laboratory test results confirmed elevated levels of C-reactive protein and serum complement C3 and C4. A venacavogram showed high-grade infrarenal inferior vena cava stenosis (image B, arrow). The patient was admitted to the hospital and given 1 mg of intravenous methylprednisolone for 3 days. A stent was placed, and the patient was discharged 6 days later. Oral prednisone was prescribed, beginning at a dose of 60 mg and decreasing by 10 mg every 3 weeks. One month after discharge methotrexate was prescribed (5 mg every 12 hours, 1 time weekly) and the patient's symptoms improved.

Retroperitoneal fibrosis is rare, and more than two-thirds of cases are idiopathic.[1] Computed tomographic and magnetic resonance images can reveal soft-tissue swelling surrounding the abdominal aorta and iliac arteries, with possible encasement of neighboring structures.[2] Management consists of immunosuppressants aimed at inactivation of acute-phase reactants and relapse prevention.[2,3] Relapses and progression to end-stage renal disease can be minimized by close follow-up and patient adherence to medication.[3] (doi:10.7556/jaoa.2017.067)

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