Case report - Thoracic general

Mediastinal fibrosis in a patient with idiopathic retroperitoneal fibrosis

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Abstract

Combined idiopathic retroperitoneal-mediastinal fibrosis is rare. We report a case of mediastinal fibrosis that followed the onset of retroperitoneal fibrosis by six years. A 61-year-old asymptomatic woman was diagnosed with idiopathic mediastinal fibrosis in December of 2006 after discovering a 1.4 cm thick prevascular mass encasing the aortic arch. In August of 2001 the patient had been diagnosed with retroperitoneal fibrosis, which was successfully treated surgically. An axillary thoracotomy found dense adhesions that fixed the arch of the aorta to the adjacent lung. Mediastinal biopsies were consistent with idiopathic fibrosis. We describe the imaging of this case and briefly review the literature.

Keywords: Mediastinal tumor; Thoracotomy; Mediastinal fibrosis pathology

1. Introduction

Retroperitoneal fibrosis (RPF) is a rare inflammatory fibrotic process leading to compression of retroperitoneal structures including the ureters. In approximately two-third of the cases it is idiopathic in nature and has the eponym Ormond’s disease. Idiopathic RPF has been found to involve the biliary system, mesentery, and perirenal structures. However, involvement of the mediastinum denotes general systemic involvement and is termed systemic fibrosis or multifocal fibrosclerosis. We present a case of mediastinal fibrosis following the onset of idiopathic RPF by six years.

2. Case report

A 61-year-old asymptomatic woman with a history of idiopathic RPF presented to our clinic in December of 2006 for follow-up of her renal function and lung nodules. A recent computed tomography (CT) of the chest showed stable lung nodules. However, a new prevascular mass was found furrowing the aortic arch (Fig. 1b). A chest CT from six months prior showed no mediastinal mass (Fig. 1a).

The patient’s history was remarkable for six years of idiopathic RPF that led to iliac deep vein thrombosis, bilateral urinary obstruction, hydrenephrosis, and atrophy of her left kidney. A 20-week course of steroids was started in October of 2003 and a right-sided ureterolysis was performed in July of 2004. Recurrent hydrenephrosis due to distal and proximal ureteral obstruction resulted in right-sided ileal ureter creation in March of 2005. At present the ileal ureter is patent and the patient has no voiding symptoms. The history is also remarkable for hypertension, a positive tuberculin test in 2003, and a positive Rheumatoid factor test.

Magnetic resonance imaging (MRI) of the mediastinal mass in January 2007 showed a soft tissue density mass measuring approximately 1.4 cm in thickness encasing the arch of the aorta approximately 270 degrees circumferentially (Fig. 2). The soft tissue density was intermediate in intensity on T1 weighted imaging, slightly hyper intense on T2 weighted imaging, and demonstrated mild diffuse enhancement following intravenous contrast.

A video-assisted thorascopic surgery and lung biopsy on the left was performed. During the procedure, dense adhesions were seen fixing the left upper lung lobe to the periaortic area near the arch. An effort was made to manipulate the left lung off of the aorta under thorascopic guidance. This was ineffective and the surgery was converted to an axillary thoracotomy.

The area of fibrosis was found to be large and extended from the medial aspect of the phrenic nerve out towards the proximal descending thoracic aorta. A wedge resection of the left upper lobe was made where it was adhered to the aorta. The resected lung wedge was then dissected off of the aorta. The lung was then retracted and a difficult dissection was performed to separate the aorta from the pulmonary artery. Multiple biopsies were performed in the periaortic tissue. The biopsies under frozen section demonstrated acute and chronic inflammation consistent with a fibrosclerotic lesion.

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3. Discussion

Fibrosis of multiple organ systems is a rare idiopathic syndrome termed multifocal fibrosclerosis. Comings et al. in 1967 reported different combinations of RPF, mediastinal fibrosis, Riedel’s thyroiditis, orbital pseudotumor, and sclerosing cholangitis in two brothers who were the offspring of a first cousin marriage [1]. Since then, numerous cases have been reported showing two or more of these entities. However, the presentation of mediastinal-retroperitoneal fibrosis is rare. In one series of 491 patients mediastinal involvement was found in only 3.3% of idiopathic RPF cases [2].

The macroscopic appearance of the sclerotic process is that of a grey-white plaque-like tissue. Histological features, which are similar regardless of which anatomic site is involved, include both acute and chronic patterns. Acute fibrosis shows collagen bundles interspersed with an equal or greater volume of inflammatory cells. Chronic fibrosis shows a fibrocollagenous stroma that is relatively avascular and acellular [3]. IgG4-related plasma cells have recently been described in a case of retroperitoneal and mediastinal fibrosis. It has been suggested that IgG4 might become a useful marker for diagnosis of sclerosing lesions and a predictor of corticosteroid sensitivity in these cases [4].

In the case presented, the patient’s idiopathic RPF preceded the mediastinal fibrosis by six years. To the authors knowledge this has not been previously reported in the literature. The patient originally presented with non-specific symptoms of idiopathic RPF — abdominal pain, constipation, and leg swelling. However, the mediastinal involvement was an incidental CT finding, and the patient was asymptomatic. The typical presentation of mediastinal fibrosis is varied and depends on the anatomy involved. Presentations involving numerous mediastinal structures have been described including aortic compression, superior vena cava obstruction, pulmonary artery obstruction with cor pulmonale, stenosis of the pulmonary veins, pericarditis with pericardial effusion, tracheal obstruction with asthma-like symptoms, esophageal stricture, and coronary artery occlusion [1, 5–7].

Therapy for multifocal fibrosclerosis includes a thorough search for an underlying infection or malignancy, removal of any inciting pharmacotherapy, and steroid therapy. Steroid therapy has been found to be effective in about 80% of cases. In refractory cases, other forms of immunosuppres-
sion such as medroxy-progesterone acetate, progesterone, and tamoxifen have been shown to be useful. Surgical intervention is considered for patients who do not respond to immunosuppressants. Long-term follow-up is recommended as recurrences are unpredictable and have been reported to occur from 3 months to more than 10 years after the initial diagnosis [8].

References


