Fibrosing Mediastinitis
A Squeeze on Arterial and Venous Segments of the Heart

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A 38-year-old woman developed fibrosing mediastinitis after an initial episode of histoplasmosis. She had history of frequent bronchitis as a teenager and developed symptoms suggestive of pneumonia unresponsive to conventional antibiotics. In addition to right lung infiltrates, she developed mediastinal lymphadenopathy. After transbronchial biopsy confirmation of histoplasmosis, she received therapy with oral itraconazole. Three years after her initial therapy with itraconazole, she presented with worsening dyspnea on exertion. Echocardiography suggested new-onset pulmonary hypertension and moderate tricuspid regurgitation. Cross-sectional imaging of the chest demonstrated thickened pleura and pericardium; calcified, enlarged lymph nodes; and prominent fibrosis in the hilar region. Thorascopic and mediastinoscopic biopsies confirmed the diagnosis of fibrosing mediastinitis. She was treated with courses of antibiotics and pulsed steroids over the years for symptoms of dyspnea, hemoptysis, and pleuritic chest pain associated with radiological evidence of disease progression. Notably, the steroids did not seem to change the disease progression.

Pulmonary function tests revealed a reduction in lung volumes with a severe reduction in diffusing capacity of the lung for carbon monoxide (DLco, 38% predicted after adjustment for hemoglobin). She developed exercise-induced oxygen desaturation and was treated with supplemental oxygen therapy. Evidence of right pulmonary artery stenosis and pulmonary vein stenosis was seen on contrast-enhanced computed tomography scan (Figure 1). The right-sided pulmonary veins were completely occluded, with significant stenosis in the left inferior pulmonary vein. Notably, the stenosis is in the intra-atrial segment and differs from venous segment stenosis caused by ablation procedures. Percutaneous treatment with balloon dilation (Movies I and II in the online only Data Supplement show before and after dilation), long-term anticoagulation, and oxygen therapy were used to open the stenotic segments are often palliative. Steroids and pleurodesis with balloon dilation can lead to obstruction of the hilar components, notably pulmonary veins, arteries, and bronchi. Superior vena cava and esophageal obstruction is often seen in these patients. Postobstructive necrotizing pneumonia and development of tracheo-esophageal fistula have been reported. Interventions to open the stenotic segments are often palliative. Steroids are useful only in the autoimmune form of fibrosing mediastinitis. Development of cor pulmonale or respiratory compromise is usually the terminal event.

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References

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290
Figure 1. A, Computed tomography (CT) scan with contrast showing significant stenosis of the right pulmonary artery. B, Perfusion scan of the lungs showing absent perfusion to the right lung. C, 2-Fluoro-2-deoxy-D-glucose positron emission tomography imaging showing overall minimal enhancement in the mediastinum; however, increased uptake is seen on 1 of the cardiac lymph nodes. D, CT reconstruction of the left atrium (LA) showing completely occluded right-sided pulmonary veins and critical stenosis in the atrial component near the left inferior pulmonary vein. E, Pressure gradient improves after balloon dilatation of the left inferior pulmonary vein (PV).

Figure 2. Autopsy specimens of the right lung from patient with fibrosing mediastinitis after histoplasmosis. Note the narrowing of artery (A, red arrow) and vein (A, white arrow) with a close-up view in B. B, Stenosis is also noted in the bronchus, which is to the right and inferior to the pulmonary vein.
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