A 77-year-old woman was referred to our center for the workup of progressive exertional dyspnea. The patient had a medical history of systemic hypertension and diabetes mellitus. Three years before admission, she was diagnosed with pulmonary tuberculosis confirmed by positive culture of *Mycobacterium tuberculosis* on bronchial aspirate. She received a 6-month antituberculosis regimen, including isoniazid, rifampicin, pyrazinamide, and ethambutol. In the previous year, the patient reported progressive exertional dyspnea. After multiple evaluations, echocardiography suggested pulmonary hypertension with an estimated systolic pulmonary arterial pressure of 60 mm Hg. The ventilation perfusion lung scan (Figure 1) showed multiple nonmatched perfusion defects of the right upper lobe and multiple subsegmental defects and hypoperfusion of the left lung, suggestive of chronic thromboembolic pulmonary hypertension. The patient was referred to the French Referral Centre for Pulmonary Hypertension for hemodynamic evaluation and assessment for operability of chronic thromboembolic pulmonary hypertension by pulmonary endarterectomy. The patient was in New York Heart Association functional class III and had no history of syncope or hemoptysis. Physical examination found an overweight woman (body mass index, 36 kg/m²) with no signs of right heart failure. ECG showed sinus rhythm at 65 bpm. Her chest radiography showed an enlarged mediastinum and cardiomegaly. Her brain natriuretic peptide level was normal. Right heart catheterization showed normal hemodynamics at rest with a mean pulmonary arterial pressure of 19 mm Hg, cardiac index of 2.4 L/min/m², pulmonary vascular resistance at 214 dynes, and normal pulmonary capillary wedge pressure of 8 mm Hg. The right ventricular pressure did not show a dip and plateau pattern evocative of constrictive pericarditis. Flexible bronchoscopy showed distorted bronchi with multiple incomplete stenoses and no endobronchial masses. These extrinsic stenoses did not influence pulmonary function tests, which were normal. Blood gases found mild hypoxemia (Pao₂=70 mm Hg). High-resolution computed tomography of the chest showed a soft-tissue calcified mass diffusely infiltrating the mediastinum and causing encasement of the right intermediate and middle lobe pulmonary arteries and of the left pulmonary artery.

**Figure 1.** Ventilation perfusion lung scan with multiple nonmatched perfusion defects in the right superior lobe and right median lobe and hypoperfusion of the left lung. RPO indicates right posterior oblique view; POST, posterior view; and LPO left posterior oblique view.
artery trunk, but also narrowing of the intermediate and left main bronchi (Figure 2). Pulmonary angiography was potentially misleading for chronic thromboembolic pulmonary hypertension owing to the presence of pulmonary artery intimal irregularities and abrupt vessel narrowing secondary to extrinsic stenoses of the intermediate and proximal left arteries. However, no organized thromboembolic lesion was seen in either pulmonary angiography or high-resolution computed tomography of the chest (Figure 3). Given these radiological and bronchoscopic findings, we concluded that the patient had mediastinal fibrosis, most likely complicating previous pulmonary tuberculosis infection. Heart-lung transplantation was not considered a potential therapy because of the patient’s age and comorbidities. Oral anticoagulation therapy was initiated to prevent clotting at sites of pulmonary arterial stenoses. The condition of the patient was stable on follow-up 10 months after the initial diagnosis.

Mediastinal fibrosis is considered a rare disorder characterized by proliferation of fibrous tissue within the mediastinum. The cause and pathogenesis of this disorder remain unknown. There are several suspected causes of fibrosing mediastinitis, including tuberculosis, sarcoidosis, histoplasmosis, other granulomatous diseases, and radiation therapy.1 There is no established effective treatment besides heart-lung transplantation for this progressive life-threatening condition, which leads to respiratory and cardiac failure by causing bronchial obstruction or pulmonary hypertension.2 Oral corticosteroids are usually not effective in this setting. Recently, the use of percutaneous vascular stent implantation in pulmonary arteries has been evaluated in patients with mediastinal fibrosis and has shown potential; however, the procedure was associated with a high incidence of complications, especially in patients with bilateral mediastinal involvement.3 Further studies are needed to confirm the potential of this approach. Because of the extrinsic obstruction of the pulmonary arteries, mediastinal fibrosis shares many characteristics with chronic thromboembolic pulmonary hypertension, including perfusion defects on V/Q lung scanning and pulmonary angiography and possible precapillary pulmonary hypertension on right heart catheterization. Physicians should be aware of this differential diagnosis of proximal chronic thromboembolic pulmonary hypertension and medical history of granulomatous diseases or radiotherapy, and the absence of typical chronic thromboembolic lesions on high-resolution computed tomography could plead in favor of mediastinal fibrosis, as illustrated by the case presented here.

Disclosures

None.

References

Figure 3. Right (A) and left (B) selective pulmonary angiogram and reconstruction of right (C) and left (D) main pulmonary arteries with a volume-rendering reformatted technique from contrast-enhanced computed tomography. Pulmonary angiogram and reconstruction of main pulmonary arteries show abrupt short-segment stenoses of the proximal right and left pulmonary arteries.
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