Mediastinal Fibrosis Treated with Endovascular Stent Placement

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Abstract
We report an interesting case of mediastinal fibrosis causing significant narrowing of both pulmonary arteries (right > left) which led to progressively increasing dyspnoea and pulmonary hypertension. This was treated with endovascular stenting of the right pulmonary artery with good clinical outcome.

Introduction
Mediastinal fibrosis is a common entity that causes slowly progressive encroachment of mediastinal structures. Venous structures (Superior Vena Cava and Pulmonary veins), due to thin walls and low intraluminal pressure, tend to be compressed earlier than are the arteries, tracheobronchial tree and oesophagus. Patients typically present with superior vena cava (SVC) syndrome.¹,² We report an interesting case of mediastinal fibrosis causing significant narrowing of both pulmonary arteries (right > left) which led to progressively increasing dyspnoea and pulmonary hypertension. This was treated with endovascular stenting of the right pulmonary artery.

Case Report
A 28 year old female patient came with dyspnoea on exertion which was grade 1 on the Medical Research Council (MRC) scale. It progressed to grade 3 dyspnoea over a period of 3 years with occasional dry cough, pedal oedema and facial puffiness which decreased on medications. There was also history of occasional bluish discoloration of skin and thinning of hair with hair loss.

Chest X-ray (Figure 1) was done which revealed mediastinal widening and prominence of the pulmonary bay. Trans-thoracic echocardiography had demonstrated a left ventricular ejection fraction of 54% with abnormal septal motion consistent with right ventricular pressure overload.

Contrast enhanced CT (Figure 2a and b) showed a nonenhancing soft tissue in the mediastinum encircling the proximal right and left pulmonary artery causing significant narrowing of these vessels, especially the right pulmonary artery. There was stenosed right descending pulmonary artery with no intraluminal abnormality. There was mild dilatation of right ventricle, right

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atrium, inferior vena cava and hepatic veins. Main pulmonary artery was dilated to approximately 3 cm.

Ventilation perfusion (V/Q) scan showed mismatch with reduction in perfusion in multiple segments of right lung. PET-CT revealed active disease in the pre-carinal and subcarinal region which needed biopsy to rule out infective/neoplastic etiology. CT guided core biopsy of the lesion showed lymphoid tissue and fragments of lung parenchyma; the fragments of lung parenchyma showed thickened interstitial septae.

Patient was then referred to us for endovascular management. Digital subtraction angiography (Figure 3) of the pulmonary arteries performed through right transfemoral venous access revealed short segment stenosis of the right pulmonary artery and mild stenosis of the left pulmonary artery. The mean pressure gradient across the right pulmonary artery stenosis was 24 mm of Hg. An AES guide wire (Cook) was passed across the stenotic lesion. A long (90 cm) 7 Fr brite tip sheath was parked in the main pulmonary artery. A 14x40 mm self-expanding stent (ev3 Protégé GPS) was placed across the lesion. Post stenting angioplasty was done using a 12x40 mm angioplasty balloon catheter (ATB, Cook). Post stenting, there was good antegrade flow across the lesion. Post stenting angioplasty revealed good flow of contrast through right upper lobe branches.

Discussion

Mediastinal fibrosis may be idiopathic or associated with tuberculosis, histoplasmosis, sarcoidosis, silicosis, or other fungal infections. The first and most common structure affected is the SVC, but involvement of the pulmonary artery, phrenic nerve, recurrent laryngeal nerve, and pulmonary veins have also been documented. In our case, the mediastinal fibrosis was involving both the pulmonary arteries. But since the stenosis was more significant and treatable in the right pulmonary artery and the V/Q scan showed a larger area of ventilation perfusion mismatch in the right lung, stenting of the right pulmonary artery was done.

In patients with mediastinal disease and pulmonary hypertension, evaluation should include careful examination of the mediastinum by CT scan or MRI and V/Q scanning for possible pulmonary artery compromise. Endovascular management is a good treatment option in patients with mediastinal fibrosis where surgical role is not feasible.

References