Pulmonary Artery Obstruction due to Giant Cell Arteritis*

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Giant cell arteritis is often referred to in the context of polymyalgia rheumatica with temporal artery involvement. There are, however, more malignant forms of presentation of this necrotizing arteritis involving either the great vessels of the aorta or, occasionally, the pulmonary arteries. Our case relates to giant cell arteritis presenting as pulmonary artery obstruction in a patient without polymyalgia rheumatica or extensive aortic or proximal great vessel involvement.

While the association of giant cell arteritis (GCA) with polymyalgia rheumatica is well described, more unusual, life-threatening presentation of this idiopathic arteritis include acute myocardial infarction, aortic arch involvement, stroke syndromes, aortic aneurysm and dissection and even aortic insufficiency. We report a case of giant cell arteritis presenting as pulmonary artery obstruction in an elderly woman without extensive great vessel arteritis.

CASE REPORT

A 77-year-old caucasian woman was admitted for evaluation of progressive dyspnea, fatigue, weakness and right-sided chest pain without fever over approximately six months, with no history of chronic headaches or muscle pain to suggest polymyalgia rheumatica. Physical examination disclosed an elderly woman in no acute distress, blood pressure 90/60 mm Hg, pulse 110 bpm and regular, mild jugular vein distention to 12 cm water, a two-over-six systolic ejection murmur at the lower left sternal border with a normal second sound and no peripheral edema.

Laboratory examination results were normal with the exception of mild anemia, a hemocrit of 36, and a Westergren sedimentation rate of 46 mm/hr. Electrocardiographic examination showed right bundle branch block and a vertical QRS axis. Ventilation lung scan showed a posterior segment of right upper lobe defect and a questionable right lower lobe wedge-shaped defect consistent with pulmonary embolism.

Echo cardiographic examination showed right ventricular enlargement with a right ventricular volume overload pattern and three plus tricuspid regurgitation by Doppler study. Left ventricular ejection fraction was calculated in the 70 to 80 percent range. The pulmonary artery was not well visualized and the aortic, mitral, and tricuspid valves were normal. There was a small pericardial effusion.

Because of worsening symptoms, the patient underwent cardiac catheterization which showed a right atrial pressure a wave of 15 mm Hg, mean pressure of 10, RV equal to 74/17, pulmonary artery pressure of 10/7 with a pulmonary capillary wedge pressure of 6, cardiac output of 1.8 L/min with a cardiac index of 1.1 L/min. There was a 64 ml gradient between the pulmonary artery and RV just above the pulmonary valve. Pulmonary artery saturation was 34.8 percent with an aortic saturation of 88.5 percent. Left heart pressure was normal, as was coronary arteriography. Pulmonary arteriogram disclosed marked narrowing of both the right and left pulmonary arteries with what appeared to be extrinsic compression of the main pulmonary artery and both of the proximal pulmonary artery segments. CT scan of the chest was essentially unremarkable with

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the exception of some small mediastinal lymphadenopathy and no evidence of extrinsic mass lesion in the region of the pulmonary arteries.

Because of severe right heart outflow tract obstruction, the patient underwent surgery for a definitive diagnosis and a palliative procedure in an attempt to increase pulmonary blood flow. At the time of surgery, the right pulmonary artery was adherent to the ascending aorta and was dissected. Then, right pulmonary arteriotomy was performed and organized thrombus was removed from the proximal right pulmonary artery. The pulmonary artery was markedly thickened and the walls of the main pulmonary artery appeared infiltrated. A right pulmonary artery endartarectomy was performed to re-establish blood flow to the right lung. However, at the time of surgery significant bleeding was encountered at the arterial cannulation site and a biopsy sample disclosed diffuse lymphocytic infiltration. Hemostasis was very difficult to achieve and the patient would not come off cardiopulmonary bypass.

A necropsy study was performed the same day and was limited to the thoracic area at the family's request. Marked thickening of the main pulmonary artery and both proximal pulmonary arteries was noted, with some adhesions of the right pulmonary artery to the ascending aorta. Histologic section of the aorta disclosed a necrotizing arteritis with round cell and giant cell infiltration involving the adventitia and the media, with sparing of the intima. The pulmonary artery was extensively infiltrated with the same process resulting in subtotal obstruction of the main pulmonary artery. There was extension of this process 4 cm into the left pulmonary artery and 2 cm into the right pulmonary artery without distal infiltration of the pulmonary arteries. The pulmonary arteries were supple without evidence of infiltration beyond the proximal thickened regions.

Moderate right ventricular hypertrophy was noted with a 1 cm right ventricular free wall thickness. Histologic sections are enclosed (Fig 1 and 2).

**DISCUSSION**

Giant cell arteritis is an idiopathic inflammatory process involving the large and medium-sized arteries. Involvement of the pulmonary artery has been reported, but pulmonary artery obstruction due to giant cell arteritis has not been previously described.

Symptoms occurred late in our patient and thromboembolism was most likely due to severely-reduced pulmonary blood flow. Our case is somewhat unusual in that extensive involvement of the pulmonary arteries without extensive aortic involvement has not been previously described. Recognition of this form of arteritis is important as significant improvement or palliation often can be obtained with steroid or anti-inflammatory therapy.

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