Epithelioid Hemangioendothelioma of the Thoracic Aorta Resulting in Aortic Obstruction and Congestive Heart Failure

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A 54-year-old white woman was referred for cardiac consultation because of increased dyspnea with exertion and leg pain. The patient’s past medical history was significant for smoking and recent onset of hypertension. On physical examination, she was found to have a blood pressure of 180/90 mm Hg and a heart rate of 110 bpm. The lungs were clear to auscultation. The carotid upstrokes were bounding, and there were bilateral carotid bruits. The central venous pressure was not elevated. The cardiac examination was significant for a loud, harsh systolic murmur (II to III/VI) across the precordium with radiation to the back. There was a left ventricular lift and a third heart sound. The abdominal examination was normal, and stool guaiac was negative. There was no peripheral edema. The femoral pulses were diminished.

The ECG showed normal sinus rhythm, left atrial enlargement, incomplete right bundle-branch block, and voltage and T-wave changes suggestive of left ventricular hypertrophy. An echocardiogram showed a global decrease in left ventricular function, with an ejection fraction of 25%. There was mild left ventricular hypertrophy and mild mitral regurgitation. The chest radiograph showed moderate cardiomegaly with clear lung fields and normal pulmonary vasculature. Significant laboratory findings included a hemoglobin of 10.1 (mean corpuscular volume = 90) and an elevated calcium of 11.6. The patient was referred for bilateral heart catheterization and coronary angiography.

At angiography, her femoral pulses were diminished. A guidewire could not be passed beyond the thoracic aorta; therefore, angiography was performed with a left brachial approach. The proximal aortic pressure was 160/73 mm Hg, and the femoral arterial pressure measured through a 6F sheath was 61/51 mm Hg. Coronary angiography showed only mild to moderate atherosclerotic changes. The left ventricular pressure was 162/8, 33 mm Hg. An aortogram revealed a severe obstruction in the thoracic aorta beyond the left subclavian artery takeoff (Figure 1). A chest magnetic resonance angiogram showed a normal-size aorta without calcification. There was a 10-cm segment of marked circumferential narrowing of the mid descending thoracic aorta, with a lumen of 8 to 10 mm at its narrowest portion (Figure 2). A CT scan of the abdomen and pelvis showed small to moderate pleural effusions. There was borderline enlargement of the hemiazygous and periaortic nodes. A 3-cm lytic lesion involved the left femoral neck, and numerous small lytic lesions involved the iliac bones and left proximal femur and ischium (Figure 3).

The patient underwent successful surgical resection of the thoracic aorta and needle biopsy of the femur. Tissue pathology revealed an epithelioid hemangioendothelioma in both specimens (Figure 4). An echocardiogram performed 5 days after resection of the aortic obstruction showed significant improvement in left ventricular function, with an ejection fraction of 45% to 50%.
Figure 1. Aortogram of descending thoracic aorta demonstrating severe aortic obstruction found at coronary angiography.

Figure 2. Cine magnetic resonance angiogram of thorax demonstrating marked circumferential narrowing of descending thoracic aorta beginning 7 cm distal to origin of left subclavian artery.

Figure 3. CT scan of pelvis showing numerous lytic lesions involving iliac bones, left proximal femur, and ischium. A 3-cm lytic lesion involves left femoral neck (arrow).

Figure 4. Characteristic clusters of atypical epithelioid endothelial cells in a myxoid background. Tumor cells show strong positive staining in immunohistochemical studies with both CD31 and CD34. Biopsies of both aortic mass and bone lesions had similar appearances.
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