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Leiomyosarcoma Involving Main and Left Pulmonary Artery Treated Surgically With Homograft Replacement and Concomitant Left Pneumonectomy

Sak Lee, MD; In-Kyu Park, MD; Sang-Ho Cho, MD; Do-Kyun Kim, MD

A 47-year-old woman presented with progressive dyspnea on exertion and generalized weakness for 6 months. On physical examination, a grade-III ejection systolic murmur was heard on her upper sternal border. Her chest x-ray showed mild cardiomegaly with multiple, variable-sized nodular opacities in the right lung (Figure 1). Echocardiography revealed severe supralvalvular pulmonary stenosis (peak and mean pressure gradient 76 mm Hg and 45.6 mm Hg, respectively) due to diffuse tubular supralvalvular stenosis associated with increased right ventricular pressure (93.9 mm Hg) and left pulmonary artery narrowing. Multislice computed tomogram showed an intravascular mass lesion at the suprapulmonic valvular area, narrowing the lumen and extending into the left pulmonary artery and upper and lower lobar arteries, with near total occlusion of the left pulmonary artery. Left lung volume loss and hypovascularity with left pleural effusion were also noted (Figure 2). A lung perfusion scan showed no perfusion in the left lung and uniform distribution of the radiotracers in the right lung (Figure 3). A positron emission computed tomographic scan was performed to evaluate the character and extent of the mass, which revealed a malignant mass of intense FDG (18F-2-fluoro-2-deoxyglucose) uptake along the main pulmonary artery and nodules and consolidations in the lungs (Figure 4). The positron emission computed tomographic scan revealed no uptake in any other organ suggesting metastasis.

The patient underwent surgical resection of the main pulmonary artery and left pulmonary artery along with concomitant left pneumonectomy under total circulatory arrest using conventional cardiopulmonary bypass technique. The main pulmonary artery, from the pulmonic valve to the proximal right pulmonary artery, was reconstructed with a homograft valved conduit, and pleural biopsy was performed on the nodules suspected of pleural seeding. The pathological diagnosis confirmed leiomyosarcoma confined to the excised pulmonary artery, and left lung without pleural metastasis. The patient suffered from bronchopleural fistula in the

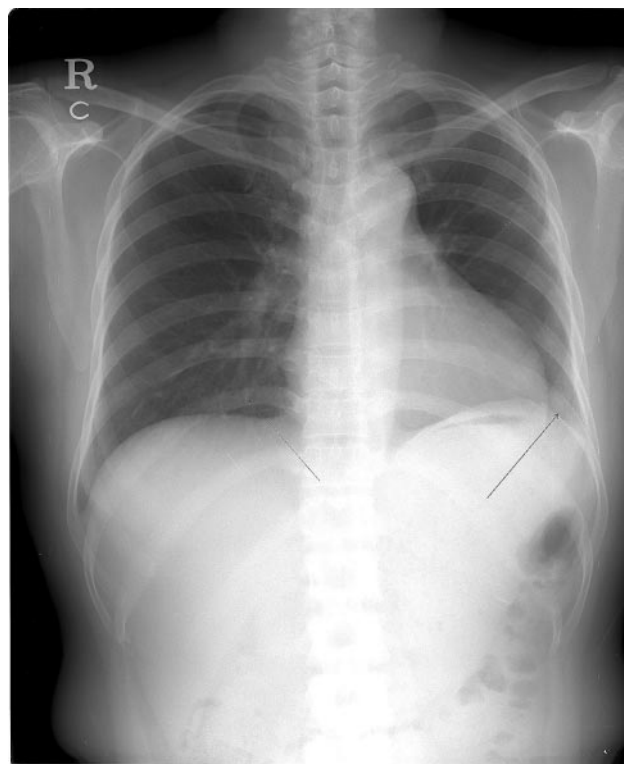


Figure 1. Preoperative chest x-ray shows mild cardiomegaly with multiple, variable-sized nodular opacities in right lung (arrows).

postoperative period, which was eventually occluded with the application of glue. Afterward, she made an uneventful recovery and was referred for chemotherapy. A postoperative chest computed tomographic scan taken 2 months after the operation shows patent pulmonary homograft with no evidence of tumor recurrence (Figure 5).

Disclosures

None.

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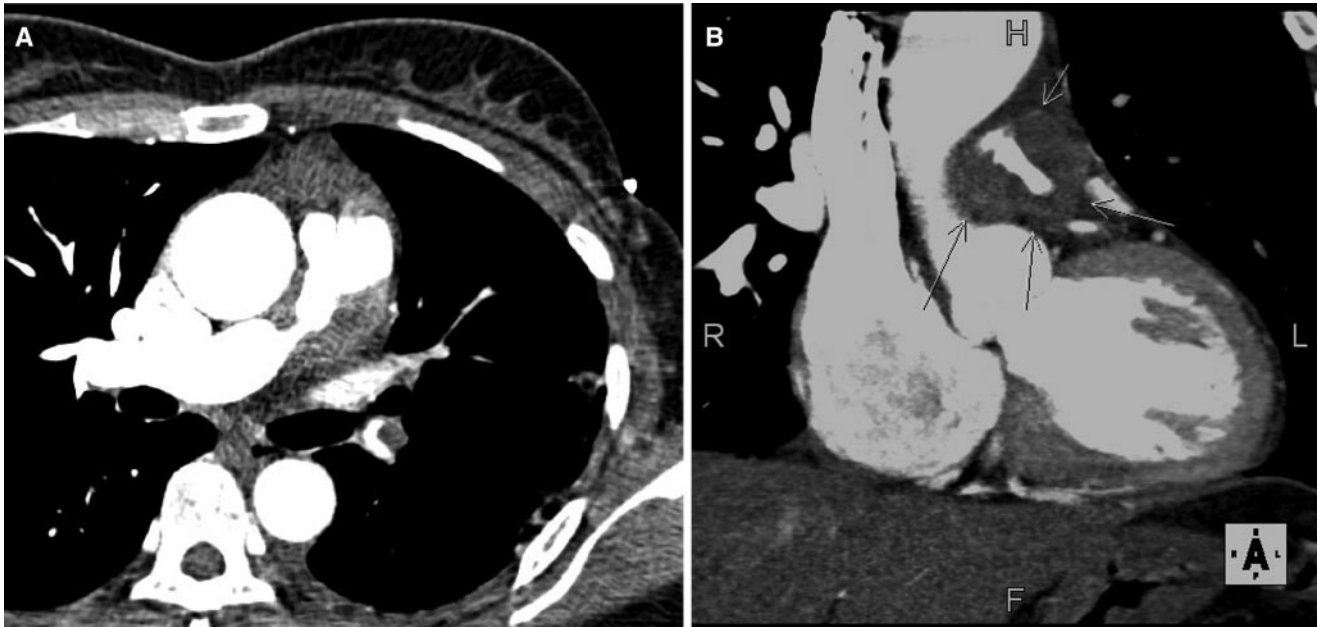


Figure 2. Preoperative multislice chest computed tomogram shows intravascular mass lesion at suprapulmonic valvular area, narrowing the lumen (arrow) and extending into left pulmonary artery and upper and lower lobar arteries with near total occlusion of the left pulmonary artery. H indicates head; R, right; L, left; and F, feet.

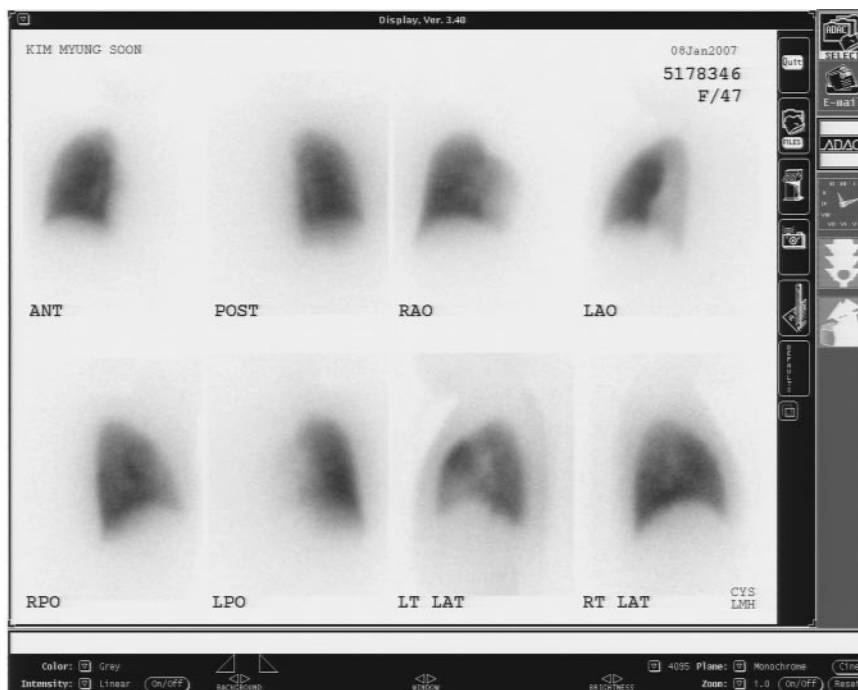


Figure 3. Preoperative lung perfusion scan shows no perfusion in the left lung and uniform distribution of the radiotracers in the right lung. ANT indicates anterior; POST, posterior; RAO, right anterior oblique; LAO, left anterior oblique; RPO, right posterior oblique; LPO, left posterior oblique; LT LAT, left lateral; and RT LAT, right lateral.

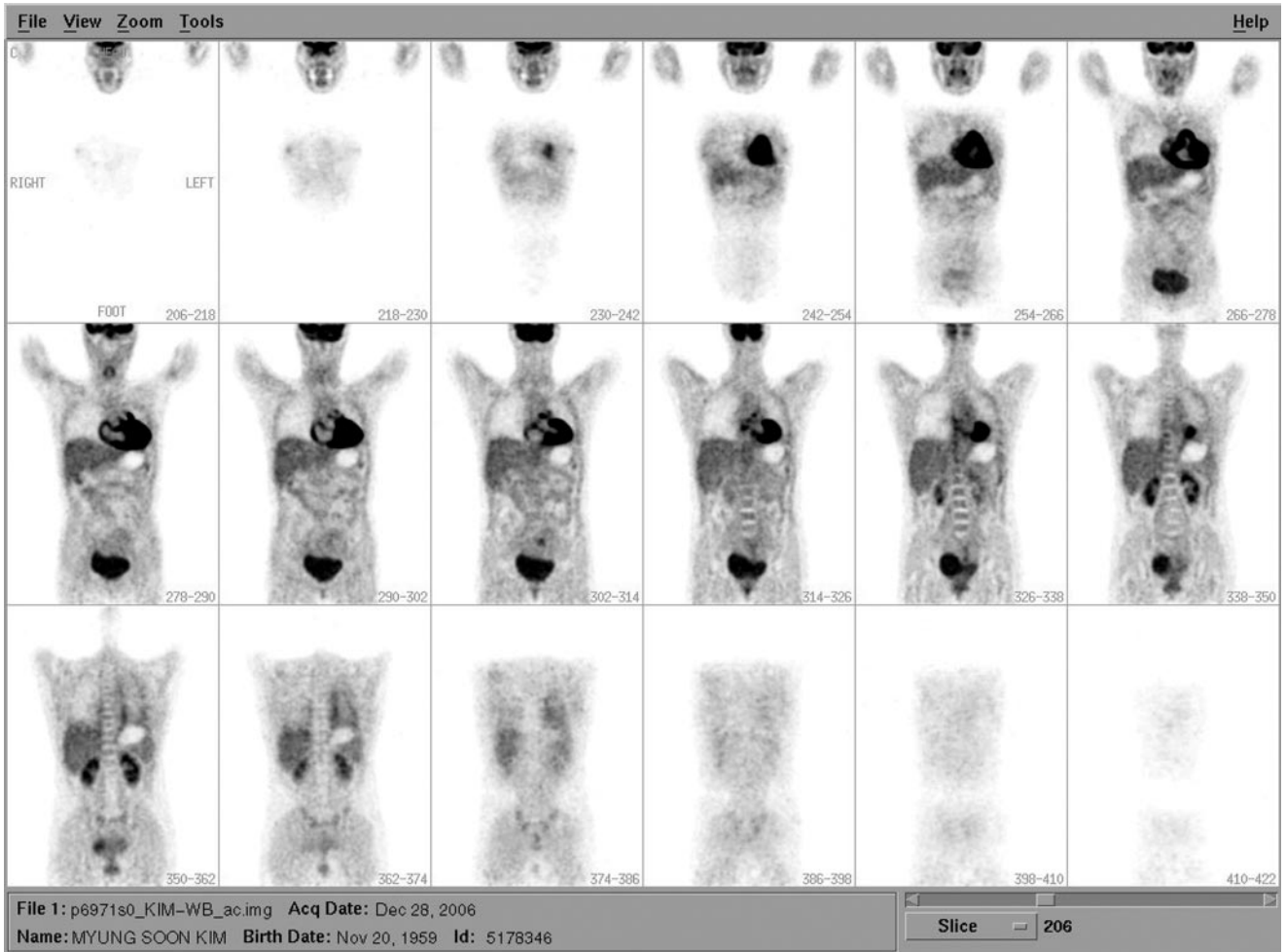


Figure 4. Preoperative positron emission computed tomogram reveals a malignant mass of intense FDG uptake along the main pulmonary artery, and nodules and consolidations in the lungs.



Figure 5. Postoperative multislice chest computed tomogram taken 2 months after the operation shows patent pulmonary homograft with no evidence of tumor recurrence.