CASE REPORTS

Myocardial Visualization on a Perfusion Lung Scan

Heidi S. Weissmann, Richard M. Steingart, Thomas M. Kiely, Leroy A. Sugarman, and Leonard M. Freeman

Albert Einstein College of Medicine and the Montefiore Hospital and Medical Center, Bronx, New York

Myocardial activity was noted on a lung scan performed following the i.v. administration of Tc-99m macroaggregated albumin. The patient had primary pulmonary hypertension with a right-to-left shunt through a functionally patent foramen ovale.

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The perfusion lung scan is a simple qualitative means of demonstrating the presence of right-to-left intracardiac shunts (*1–3*). The presence of renal or intracerebral activity following the i.v. injection of labeled macroaggregated albumin (MAA) indicates that such a shunt is very likely present. The purpose of this communication is to present a case of right-to-left shunt where myocardial activity was noted following i.v. injection of MAA.

CASE REPORT

The patient was a 21-year-old woman with known cyanotic heart disease who was admitted to the medical center on July 19, 1979, with a chief complaint of pleuritic chest pain and hemoptysis.

The patient was born by Caesarean section after an uncomplicated full-term pregnancy. Her birth weight was 7 lbs. 5 oz. She developed normally until 6 mo of age, when her mother noted dyspnea with feeding, and at 12 mo she was clearly underdeveloped. She was first admitted to the hospital at age 3.5, with failure to thrive and dyspnea and fatigue on exertion. Physical examination at that time revealed her to be underdeveloped and acyanotic at rest, but cyanotic with crying. Cardiac examination revealed a second heart sound that was single and loud. A grade 3/6 holosystolic murmur was heard at the left fourth intercostal space. The chest radiograph revealed cardiomegaly with right-ventricular enlargement and pulmonary vascular overcirculation. The electrocardiogram was compatible with right ventricular hypertrophy. During cardiac catheterization there was no significant oxygen step-up suggestive of a left-to-right shunt. Femoral-artery saturation was 77% and pulmonary artery pressure was 130/64, with no significant systolic gradient between the right ventricle and pulmonary artery. The systemic blood pressure was 80/40. Dye curves revealed early appearance of dye in the aorta on the right atrial injection, indicating a right-to-left shunt at the atrial level. A diagnosis of primary pulmonary hypertension was made.

By age 12 increasing congestive heart failure was noted, and the patient was placed on digoxin. Her course was subsequently characterized by bouts of hemoptysis precipitated by frequent upper respiratory infections, and polycythemia with hematocrits greater than 65, requiring phlebotomy.

The present admission at age 21 was preceded by a 2-wk history of an upper respiratory infection treated with tetracycline and a history of hemoptysis 10 days before admission. Four days before admission the patient complained of left-sided chest pain. Physical examination on admission revealed the patient to be thin, cyanotic, and in respiratory distress. Blood pressure was 86/66, pulse 112 and regular, temperature 37.6°C and respiratory rate 24. Her lungs were clear to percussion and auscultation. Cardiac examination revealed a diffuse apical impulse with a left parasternal heave. S1 was normal, and there was a grade 3/6 harsh systolic murmur at the left lower sternal border, radiating widely. S2 was loud and single. The arterial blood gases revealed a pH of 7.45, pO_2 of 33.5 mm Hg and pCO_2 of 32. The chest radiograph showed a large, wedge-shaped, pleural-based, left upper lobe infiltrate (Fig. 1). A perfusion lung scan was performed on the first hospital day (Fig. 2). It revealed heterogeneous uptake throughout both lungs,

FIG. 1. Postero-anterior chest radiograph shows markedly enlarged cardiac silhouette with infiltrate in upper lobe of left lung.

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For reprints contact: Heidi S. Weissmann, MD, Dept. of Nuclear Medicine, Montefiore Hospital and Medical Ctr., 111 E. 210th St., Bronx, NY 10467.

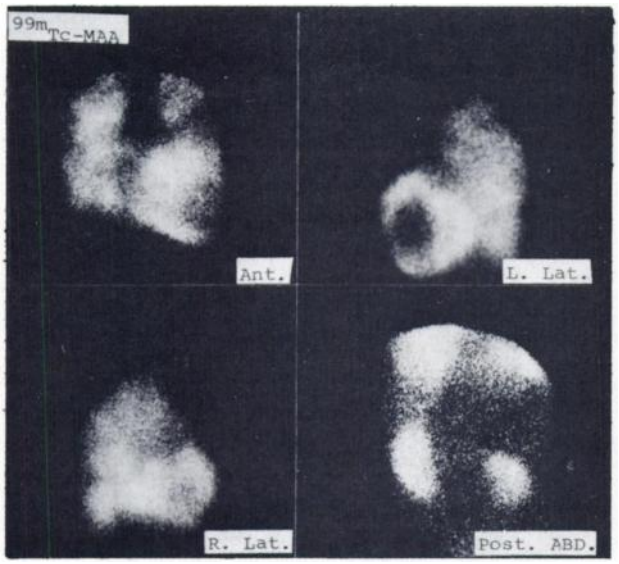

FIG. 2. Perfusion lung scintigrams show diffuse heterogeneous uptake. Myocardial activity is clearly evident, most distinctly in anterior and left lateral views. Significant renal activity confirms presence of a right-to-left shunt. Band-like area of pulmonary ischemia in left upper lung field is seen just above myocardial activity in anterior view. This corresponds to radiographic lung density.

with an oblique band of marked ischemia in the midleft anterior lung field. The most distinctive feature was uptake in the myocardium, which was most strikingly apparent on the anterior and left lateral views. A posterior scintiphoto of the abdomen showed bilateral renal activity, thereby confirming the presence of a right-to-left shunt.

After walking a few steps on the sixth hospital day, the patient complained of extreme dyspnea and lightheadedness. She subsequently sustained a respiratory arrest followed by a cardiac arrest. Despite basic and advanced cardiopulmonary resuscitation, cardiac rhythm could not be restored, and the patient died.

Autopsy was limited to examination of the heart and lungs. There was massive cardiomegaly (560 g), with right atrial dilatation as well as right ventricular dilatation and hypertrophy. The right ventricular wall was 1.7 cm thick (Fig. 3). The ductus arteriosus was probe closed, but the foramen ovale was probe patent and displayed multiple fenestrations, with an overall measurement of 2.0 × 1.5 cm. An area of subendocardial fibrosis, 6.0 × 5.0 cm, was evident at the apex of the right ventricle. Microscopically, marked myocardial fiber hypertrophy was evident, with patchy areas of interstitial fibrosis. The coronary arteries were unremarkable; no congenital anomalies were identified.

The lungs displayed mild edema. Moderately severe pulmonary artery atherosclerosis was evident. An acute pulmonary infarction was evident in the left upper lobe, which was 50% compromised, as well as multiple small healed infarcts bilaterally. Organized and organizing pulmonary emboli were found bilaterally. Patchy septal fibrosis was present, as well as patchy mononuclear cell infiltrates of alveolar septa. There was moderately severe pulmonary vascular sclerosis, primarily involving the larger vessels, with minimal involvement of the smaller arteries.

The final anatomic diagnosis was idiopathic pulmonary hypertension with pulmonary vascular sclerosis, acute pulmonary infarction, massive right ventricular hypertrophy, and a probe-patent foramen ovale.

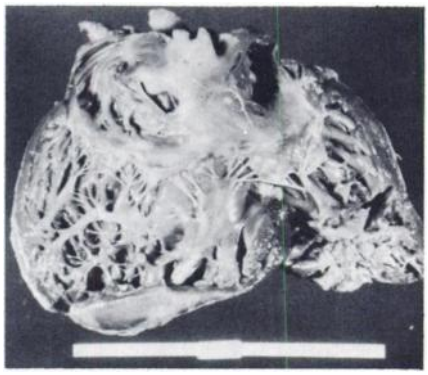

FIG. 3. Autopsy specimen of heart shows massively hypertrophied right ventricular musculature resulting from patient's primary pulmonary hypertension.

COMMENT

Primary pulmonary hypertension is a disease that generally is characterized by an obliterative cellular intimal thickening in the small nonmuscular arterioles and is associated with medial hypertrophy in more proximal vessels (*4*). From a pathologic standpoint, the primary involvement of the larger pulmonary vessels represents an unusual feature of this case.

The marked myocardial and renal perfusion noted on the lung scan must be explained on the basis of right-to-left interatrial shunting via a functionally patent foramen ovale. In cases of pulmonary hypertension, right-sided atrial pressures may exceed those of the left side, and the patent foramen ovale affords a viable route for shunting (*5*).

As indicated earlier, renal activity is the most commonly observed finding on lung scan performed in patients with right-to-left shunts (*1–3*).

Greenfield and Bennett (*1*) used calculations based on the distribution of cardiac output and renal blood flow to estimate that a shunt of greater than 15% is required in order to make the kidneys apparent on a routine lung scan. The relatively small contribution of cardiac output to coronary blood flow (~5%) should generally preclude myocardial visualization in such cases. The hemodynamic cause for the unusual, but obviously significant, increase of coronary blood flow to our patient's massively enlarged right ventricle remains unknown to us.

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