
Idiopathic Peripheral Pulmonary Artery Stenosis: An Unusual Cause of Ventilation-Perfusion Mismatch

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Idiopathic peripheral pulmonary artery stenosis is a rare cause of ventilation-perfusion mismatch in adults. We report a 39-yr-old man with this entity. Pulmonary scintigraphy demonstrated findings indistinguishable from pulmonary embolism, and pulmonary angiography was necessary for accurate diagnosis.

Key Words: pulmonary artery stenosis; pulmonary hypertension; ventilation-perfusion mismatch

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Peripheral pulmonary artery stenosis (PPS) is known to occur in the pediatric population, usually in association with other congenital cardiac anomalies. We report an unusual case of idiopathic PPS in an adult who was initially evaluated and treated for pulmonary embolism. Pulmonary scintigraphy demonstrated findings indistinguishable from pulmonary embolism. Pulmonary angiography was necessary to establish the final diagnosis of PPS and to determine surgical versus medical management.

CASE REPORT

A 39-yr-old healthy nonsmoking man presented with a 3-mo history of progressive dyspnea and vague bilateral pleuritic chest pain. At the time of onset of the dyspnea, the patient was employed as an airline pilot and involved in an intensive fitness program which included 6 miles of jogging per day. The patient had no known history of congenital heart disease or prior pulmonary disease.

His physical examination was remarkable for a systolic ejection murmur over the pulmonic valve. His lungs were clear to auscultation and percussion. Laboratory findings revealed moderate hypoxia by arterial blood gas analysis, which showed a pH of 7.44, $PACO_2$ of 39 mmHg, and PAO_2 of 81 on room air. The electrocardiogram and chest radiograph were unremarkable. Two-dimensional echocardiography demonstrated a decreased pulmonary acceleration time of 50 msec, with moderate right

ventricular hypertrophy, consistent with pulmonary hypertension.

Nuclear medicine was then considered the modality of choice to exclude pulmonary embolism as a possible cause of acute hypoxia and pulmonary hypertension. Pulmonary perfusion scintigraphy was performed following intravenous injection of 4.5 mCi of ^{99m}Tc -macroaggregated albumin. Pulmonary ventilation scintigraphy was performed after inhalation of 11 mCi/min of ^{81m}Kr . The study showed a marked ventilation-perfusion (V/Q) mismatch pattern with absent perfusion in the right upper lobe and segmental perfusion defects in the left upper and lower lobes (Fig. 1). The ventilation scan was normal (Fig. 1).

The patient was treated for presumed pulmonary embolism for approximately 6 wk with anticoagulation therapy with therapeutic doses of heparin, and then coumadin, without significant clinical improvement. Further evaluation using pulmonary angiography using bilateral selective injections of the right and left pulmonary arteries revealed branch stenoses of the right upper lobe pulmonary artery and basilar segmental left lower lobe branches (Fig. 2). Pulmonary artery pressures were elevated and measured 54/16 mmHg in the right and 63/22 mmHg in the left pulmonary artery. Normal systolic and diastolic pulmonary artery pressures are 20 and 12 mmHg, respectively; pulmonary hypertension is considered when the mean arterial pressure exceeds 20 mmHg (1). On the basis of these new findings, the patient was subsequently referred to another hospital for elective percutaneous transluminal balloon angioplasty therapy. Subsequent follow-up at 1 yr post-angioplasty revealed an improved V/Q mismatch pattern on pulmonary scintigraphy. This correlated with an excellent clinical response, with resumption of the patient's baseline level of activity, including 6 miles of jogging per day.

DISCUSSION

The most common causes of V/Q mismatch include pulmonary embolism, bronchogenic carcinoma and radiation therapy, whereas pulmonary hypertension is an uncommon cause (2). Pulmonary artery agenesis and stenosis are considered rare causes of V/Q mismatch (3).

Most cases of PPS occur in the pediatric population and are associated with other cardiovascular anomalies, such as William's syndrome or maternal rubella infection (4). The clinical course of PPS is variable but frequently related to its associated cardiovascular lesions, whereas isolated PPS (both familial and nonfamilial) is generally stable.

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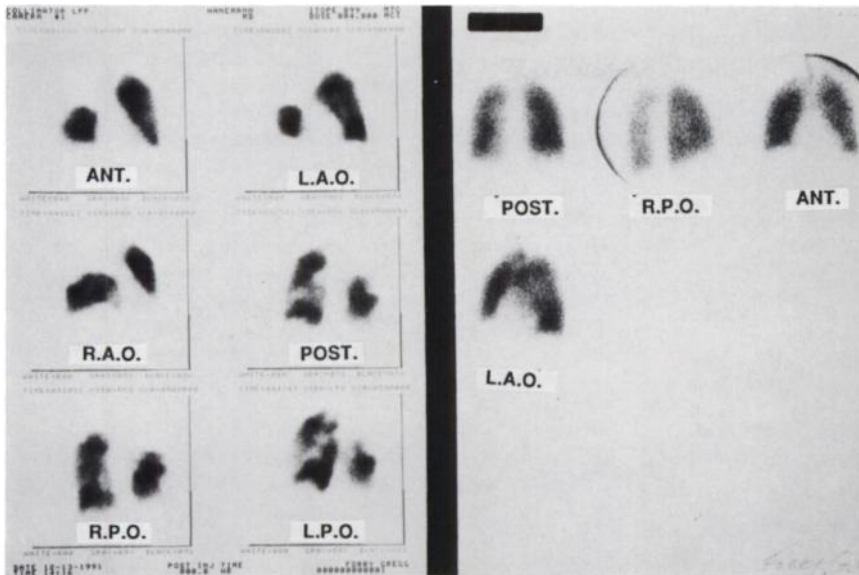


FIGURE 1. Technetium-99m-MAA perfusion scan (left) shows absent perfusion of right upper lobe (arrow) and segmental defects in the left lower lobe (arrowheads); ^{81m}Kr ventilation scan (right) was normal.

Complications such as in situ thrombosis and progression of stenotic lesions leading to pulmonary hypertension have been reported (4).

PPS is rare in adults and can occur as complications of granulomatous disease (5). Marks described two cases of PPS in pregnancy associated with maternal rubella syndrome (6). Takeuch et al. reported a case of PPS in a patient with combined factor VII and protein C deficiency (7). Our case of idiopathic PPS is interesting in that the clinical presentation and findings on pulmonary scintigraphy were similar to pulmonary embolism. The scintigraphic findings involved a marked V/Q mismatch pattern with perfusion defects in the right upper and left upper and lower lobes, and normal ventilation. The scintigraphic features of pulmonary embolism consist mainly of multiple, bilateral perfusion defects in a segmental (usually basilar) distribution (8).

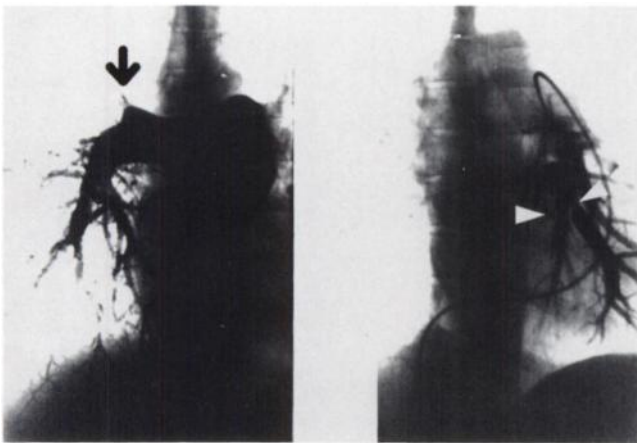


FIGURE 2. Anteroposterior pulmonary angiograms demonstrate stenosis of right upper lobe pulmonary artery (arrow) and multiple stenoses of segmental branches of the left lower lobe pulmonary arteries (arrowheads).

The patient's clinical course, however, was atypical for pulmonary embolism given his age. Typically, there is some evidence of change in the pattern of perfusion defects in the first few days after embolism, with clinical improvement (i.e., higher oxygen saturation). Approximately 50% of patients under 40 yr have complete resolution of symptoms clinically and reversible V/Q mismatch by pulmonary scintigraphy within the first few weeks of a standard 3–6-mo course of anticoagulation therapy (8). Since our patient did not respond to anticoagulation therapy, a complication of pulmonary embolism was considered, such as fragmentation of a larger centrally placed clot with peripheral embolization and possible lung infarction. In retrospect, follow-up pulmonary scintigraphy could have been performed in 1 or 2 wk; if unchanged or worse, a pulmonary angiogram could have been performed sooner to expedite diagnosis and management.

The diagnosis of PPS with pulmonary angiography was essential in the clinical management of our patient. The management of pulmonary embolism involves medical therapy (i.e., approximately 3–6 mo of anticoagulation) (9). Significantly, this therapeutic approach resulted in little clinical improvement in our patient and also delayed the eventual diagnosis, fortunately without significant morbidity. The management of PPS, however, is surgical, either as a staged surgical procedure or using newer interventional techniques such as percutaneous transluminal balloon angioplasty (PTA) (10). Beekman has used PTA successfully with a 50% to 60% hemodynamic benefit in children with PPS associated with tetralogy of Fallot (11). Clearly, failure to diagnose PPS can lead to incorrect patient management, as in our patient, and incur additional morbidity and mortality risk. This is especially critical in young patients who present with clinical and scintigraphic findings of pulmonary embolism but no significant risk factors. In this group of patients, other causes of regional

V/Q mismatching should be investigated and angiography should be considered to exclude lesions potentially amenable to surgical or interventional methods for correction. In addition, idiopathic pulmonary artery stenosis should be included in the differential diagnosis of V/Q mismatch.

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