A 45-Year-Old Man With Left Lung Hypoperfusion and Possible Pulmonary Embolism*

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A 45-year-old man presented with left-sided pleuritic chest pain and shortness of breath that developed suddenly while playing golf. The man had been in his usual state of health when these symptoms developed. His medical history was significant only for atrial fibrillation that was successfully treated 8 months previously via circumferential radiofrequency ablation. The patient was a nonsmoker and worked as a painter, but otherwise his history was noncontributory.

Physical Examination and Laboratory Data

Physical examination on presentation was unremarkable, with a BP of 148/90 mm Hg; pulse rate, 84 beats/min; respiratory rate, 20 breaths/min; and oxygen saturation, 98% on room air. Cardiac enzymes were negative, with a creatine phosphokinase level of 95 U/L and a troponin I level of < 0.03 ng/mL. ECG showed sinus rhythm with a left anterior fascicular block. Findings of subsequent chest radiography were normal, but a ventilation/perfusion (V/Q) scan was read as high probability for pulmonary embolism.

Hospital Course

The patient was started on IV heparin and admitted to the hospital with a diagnosis of pulmonary embolism. Results of subsequent studies including lower-extremity duplex ultrasound and a d-dimer were negative. In addition, an echocardiogram demonstrated normal right ventricular size and function. Further review of the V/Q scan revealed global hypoperfusion in the left lung with a second defect in the right upper lobe. The discordance between the patient’s clinical condition, specifically the lack of tachycardia or tachypnea, and the striking global hypoperfusion of the left lung prompted further evaluation with a CT angiogram. The CT angiogram did not reveal any pulmonary emboli, but there were multiple small peripheral infiltrates seen on the initial review (Fig 1). A second review of the CT angiogram suggested that although there were no emboli evident, there was a paucity of blood vessels in the left lung. Three-dimensional reconstruction of the CT angiogram was done to further evaluate this possibility (Fig 2) and confirmed the finding of left lung oligemia. A quantitative perfusion scan demonstrated markedly asymmetric perfusion, with 70% of blood flow going to the right lung.

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Figure 1. CT angiogram demonstrating relative oligemia of the left lung as compared to the right. Also note left upper lobe ill-defined opacities.
What is the diagnosis and how would you treat it?
Diagnosis: Pulmonary vein stenosis secondary to atrial fibrillation ablation

One goal in the treatment of atrial fibrillation is restoration of sinus rhythm. This is sometimes accomplished with electrical cardioversion, but may also be attained with medical therapies such as ibutilide, propafenone, or flecainide. When these therapies fail or atrial fibrillation recurs, ablative therapies can be used.

In most instances, atrial fibrillation arises from reentrant circuits in the atria, but more recent studies of patients with spontaneous atrial fibrillation have demonstrated that the pulmonary veins are often a source of ectopic beats in a large percentage of these patients. The circumferential radiofrequency ablation procedure utilizes a transeptal approach, which allows catheters to be placed in the left atrium for defining anatomic relationships between the atrium and the pulmonary veins. Mapping procedures allow identification of the areas within the pulmonary veins that activate local endocardium. The pulmonary veins are subsequently ablated in a circumferential fashion within 5 mm of the ostia. The procedure end point is electrical isolation of all pulmonary veins from the left atrium. A second approach used for radiofrequency ablation called the “maze” procedure involves linear energy delivery to both the right and left atria in an attempt to decrease the atrial mass within which reentrant wavelets cause atrial fibrillation. Ablation therapy has proven effective in eliminating atrial fibrillation in 62 to 86% of patients undergoing these ablative procedures.

Complications of these ablative procedures are varied and can include pericardial effusion, systemic emboli, pulmonary dysfunction, and bleeding. An additional and potentially severe complication is pulmonary vein stenosis. This entity is likely related to thermal injury of the venous wall and may manifest clinically with dyspnea, orthopnea, cough, hemoptysis, pulmonary infections and, at the most extreme level, pulmonary hypertension and cor pulmonale. Its incidence has been reported to range from 3 to 42% depending on the ablative procedure used and the method of pulmonary vein assessment.

What constitutes significant pulmonary vein stenosis varies between studies depending on the diagnostic methodology used. There is currently no single widely accepted standard. Options include CT with three-dimensional reconstruction, magnetic resonance angiography (MRA), and transesophageal echocardiography (TEE). The definition of significant stenosis varies with the method of testing. Using CT and MRA as the reference, significant stenosis has been defined as a 50% reduction in diameter between preablation and postablation measurements of the pulmonary veins. Severe stenosis was defined as a ≥ 70% reduction in diameter.

When using TEE, pulmonary vein Doppler flow velocity can be used to establish the diagnosis of pulmonary vein stenosis. A Doppler flow ≥ 110 cm/s and a minimal flow between systolic and diastolic peak flow > 60% of the mean of both peaks, or a peak flow velocity 2 SDs above the preablation flow rate have been used as criteria for identifying pulmonary vein stenosis using TEE.

There are limited data comparing techniques for diagnosing pulmonary vein stenosis. Comparisons of conventional venography to three-dimensional MRA, CT, and intracoronary ultrasound revealed a high concordance. Other reports have shown TEE to be inferior to other modalities in defining pulmonary venous anatomy, although one study demonstrated concordance between three-dimensional MRA and TEE.

In this case, three-dimensional CT angiogram was superior to axial imaging alone in recognizing pulmonary oligemia. Although vascular asymmetry is noticeable with axial imaging, this finding is subtle and may easily be overlooked (Fig 1, 2). Three-dimensional reconstruction makes asymmetric vascularity more obvious to the physician looking for pulmonary oligemia. However, focused axial imaging, paying attention to the entrance of the pulmonary veins into the left atrium, is important and can confirm the diagnosis of pulmonary vein stenosis. In this case, review of the CT angiogram revealed narrowing of the ostia of the left superior and inferior pulmonary veins (Fig 3). In the setting of suspected pulmonary vein stenosis, the combination of CT with three-dimensional reconstruction and focused axial imaging of the pulmonary veins likely increases sensitivity in diagnosing pulmonary venous stenosis (Fig 2, 3).

The number of veins with stenosis is related to the probability of developing clinically significant disease in the future. Although pulmonary vein stenosis may occur in 3 to 42% of patients, if there is only single vein involvement it is often asymptomatic. However, if there is stenosis of more than one pulmonary vein then it is frequently associated with clinically significant pulmonary hypertension.

In those with clinically significant pulmonary vein stenosis, presenting symptoms vary. As many as 44% may be asymptomatic, while others present with shortness of breath, cough, hemoptysis, and pleuritic chest pain. The exact time course for development of pulmonary vein stenosis and the development of long-term clinical sequelae are not precisely defined. Depending on the definition, the onset of pulmonary...
venous stenosis has been estimated to occur anywhere from 5 to 24 months after ablation.

Radiographic abnormalities are present in approximately one half of patients, with the most common radiographic abnormalities reported being lung consolidation and pleural effusions. Unfortunately, the symptoms and radiographic abnormalities are non-specific and are often attributed to other causes, resulting in delays in treatment as well as unnecessary diagnostic and therapeutic procedures.

Optimal treatment for patients who acquire postablation pulmonary vein stenosis is an area of ongoing debate. The two most frequently used modalities are balloon angioplasty and stent placement, although there is significant potential for restenosis. In addition, although a case report has demonstrated success as far out as 1 year following angioplasty with stenting, large-scale longitudinal studies with long-term follow-up data are lacking.

The present patient had acute pleuritic chest pain and shortness of breath diagnosed originally as a pulmonary embolism based on a high-probability V/Q scan. He was treated initially with IV heparin. This was discontinued when a CT angiogram failed to show pulmonary emboli but instead demonstrated left-sided oligemia and peripheral infiltrates. Three-dimensional reconstruction of the CT angiogram confirmed the paucity of blood vessels on the left and also demonstrated narrowing of the left superior and inferior pulmonary veins. Based on the patient’s history of circumferential radiofrequency ablation and the CT findings, it was determined that the patient had acquired pulmonary vein stenosis with pulmonary infarction secondary to his ablative procedure. As the echocardiogram showed no evidence of pulmonary hypertension or cor pulmonale, the patient was discharged to home when his symptoms abated on day 4 of hospitalization. The patient remains asymptomatic, now 15 months after ablation.

Clinical Pearls

1. Potential complications of atrial fibrillation ablation therapy include bleeding, pericardial effusions, systemic emboli, and pulmonary vein stenosis.

2. The importance of recognizing pulmonary vein stenosis lies in the morbidity and mortality associated with its long-term sequela of pulmonary hypertension and cor pulmonale, especially since there are therapeutic interventions available.

3. Pulmonary vein stenosis may present anywhere from 5 to 24 months after ablation with shortness of breath, cough, hemoptysis, and pleuritic chest pain, and often mimics pulmonary embolism.

4. Diagnostic modalities include CT angiogram, MRA, and TEE.

5. A CT angiogram with three-dimensional reconstruction significantly enhances the physician’s ability to detect pulmonary oligemia and is available in many medical centers, making it very useful in the diagnosis of pulmonary vein stenosis.

Suggested Readings


Figure 3. CT angiogram with narrowing of the left superior pulmonary vein (large arrow) as it enters the left atrium. Compare to normal right superior pulmonary vein (small arrow).