A 78-year-old woman was referred for evaluation of a chest mass. She described progressive dyspnea on exertion over the past several years that she attributed to deconditioning. She had an occasional cough productive of a minimal amount of clear to yellow sputum. She denied wheezing, paroxysmal nocturnal dyspnea, hemoptysis, fever, chills, or weight loss. She also denied chest pain and had no history of coronary artery disease. She had been admitted to the hospital in March 1996 for cataract surgery and was noted to be hypoxemic. She had normal findings from an evaluation for coronary artery disease, including an echocardiogram, which did not show an intracardiac shunt. In January 1997, she was admitted to the hospital for evaluation and management of an embolic cerebrovascular accident. She did not have carotid artery disease. She was a former smoker but quit 30 years ago.

**Physical Examination**

**General:** The patient appeared well and in no acute distress. Vital signs were as follows: respiratory rate, 14 breaths/min; heart rate, 88 beats/min; temperature, 37°C. Chest was clear to auscultation and percussion. Cardiovascular examination showed regular rate and rhythm without murmurs, gallops, or rubs. Her extremities showed mild acral cyanosis without clubbing or edema. Neurologic examination showed mild left-sided weakness, a residual of prior stroke.

**Laboratory Findings**

Hematocrit was 52%. Room air arterial blood gas values were as follows: pH, 7.42; \( \text{PaCO}_2 \), 35 mm Hg; and \( \text{PaO}_2 \), 53 mm Hg. Results of coagulation studies and chemistries were normal. Pulmonary function test results were as follows: FVC, 1.87 L (68% of predicted); FEV\(_1\), 1.42 L (75% of predicted); FEV\(_1\)/FVC ratio, 76%; and diffusion of carbon monoxide, 66% of predicted. Chest radiograph is shown in Figure 1.

**Figure 1.** Chest radiograph showing mass-like abnormality in right lower lobe.

What is the diagnosis?

What tests would confirm the diagnosis?

What are the available modes of therapy?

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Diagnosis: Large solitary pulmonary arteriovenous malformation (PAVM) with probable secondary paradoxical emboli causing stroke.

DISCUSSION

PAVM, also known as congenital arteriovenous fistulas, represent abnormal vascular communications between pulmonary arteries and pulmonary veins. This is an uncommon congenital defect, and although present at birth, the malformation typically does not become symptomatic until adulthood. It is estimated that approximately 30 to 50% of these lesions are associated with Osler-Weber-Rendu disease (OWRD) (hereditary hemorrhagic telangiectasia). Conversely, approximately 15% of patients with OWRD have PAVMs, and are reported to have increased symptoms with an increased incidence of multiple arteriovenous malformations, a more rapid rate of progression, and a higher morbidity.

PAVMs are most frequently found in the lower lobes, usually near the visceral pleura. They occur as single lesions in 40 to 60% of patients, multiple lesions in 30 to 50%, and are bilateral in 8 to 20%. Symptoms and abnormal physical findings are associated more with the size of the lesion rather than the number. Single lesions with a diameter of < 2 cm rarely cause symptoms.

The most common symptoms at the time of presentation include dyspnea on exertion, palpitations, hemoptysis, or chest pain. Patients presenting with epistaxis, GI tract bleeding, hematuria, or neurologic symptoms should be evaluated for coexisting OWRD. As many as 75% of patients may have cyanosis, clubbing, pulmonary vascular bruits, or systolic murmurs on physical examination. Careful auscultation can detect murmurs that can be intensified by inspiring against a closed glottis (Müller's maneuver) and alleviated by the Valsalva maneuver.

Ear, nose, and throat examination may confirm the diagnosis of OWRD by demonstrating the presence of nasopharyngeal or oral mucosal telangiectasias. Neurologic symptoms, including headaches, confusion, dizziness, syncope, and cerebral vascular accidents may arise from the associated hypoxia and secondary polycythemia or paradoxical emboli. Brain abscesses, probably caused by paradoxical septic emboli, have also been reported.

The primary physiologic abnormality is a right to left shunt from pulmonary artery to pulmonary vein, leading to hypoxemia and in extreme cases high output cardiac failure. The most common laboratory finding is reduced arterial oxygen tension, occurring in > 80% of cases. Hemodynamic measurements, including cardiac output, intracardiac, pulmonary arterial, and pulmonary capillary wedge pressures are usually within normal limits. Chest radiographs are abnormal in approximately 98% of patients with PAVM. The most common finding is a well-circumscribed, peripheral, noncalcified nodule(s) connected by blood vessels to the hilum of the lung. Pulmonary angiography remains the gold standard for diagnosing PAVM, providing information regarding size of the lesion, the location of the lesion, and the vessels involved in the malformation. Other modes of investigation include CT, contrast echocardiography, fluoroscopy, perfusion lung scan, and shunt evaluation. Figure 2 shows a CT scan illustrating the PAVM with accompanying pulmonary arterial feeder and venous draining vessels.

The treatment is either surgical excision or vascular embolization. As data accumulate, it is becoming more apparent that conservative management, even in asymptomatic patients, is associated with significant mortality and morbidity. Complications resulting from untreated PAVM include disabling cerebro-
vascular accidents, rupture and hemorrhage into the lung parenchyma, pneumonia occurring in atelectatic areas of the lung, and cardiovascular effects of chronic hypoxia. Prior to coil and balloon embolization, the mainstay of therapy involved surgical resection entailing local excision, segmental resection, lobectomy, or pneumonectomy. However, beginning in the late 1970s, percutaneous catheterization of the femoral vein and embolization of the malformation through a catheter positioned in the feeding pulmonary artery has become the treatment of choice. The advantages of this procedure arise from the avoidance of surgery, general anesthesia, and loss of lung tissue and the fact that patients can undergo repeated procedures for recurrent or multiple PAVMs. This approach, however, is not universally available.

In the case presented herein, six coils were placed in the solitary feeding pulmonary artery resulting in complete occlusion of the artery and obliteration of the malformation on subsequent dye injections. Figure 3 shows the pulmonary angiogram and subsequent embolization. Before the patient left the angiography table, her oxygen saturation had increased from 85 to 95%. In follow-up, the patient has remained in stable condition with an oxygen saturation of 96% on 6-month follow-up. She has also had no recurrent neurologic events.

**PEARLS**

1. Pulmonary arteriovenous malformations may be single or multiple, and usually result in hypoxemia; solitary PAVMs < 2 cm rarely cause symptoms.

2. All patients with PAVMs should be evaluated for concomitant OWRD.

3. Definitive workup includes chest radiography, chest CT, arterial blood gas measurements, and pulmonary angiography. Contrast echocardiography and pulmonary scintigraphy are optional.

4. PAVMs should be treated. Observation is associated with increased morbidity and mortality.

5. Pulmonary artery embolization is the treatment of choice.

**SUGGESTED READING**


**Figure 3.** Pulmonary arteriogram showing the PAVM prior to (left) and following coil embolization of the pulmonary artery (right).