Pulmonary Arteriovenous Malformation Presenting with Severe Hypoxemia

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Abstract
\textbf{Objective:} To report a case of right pulmonary arteriovenous malformation (PAVM) affecting the right upper lobe, following the incidence of empyema thoracis in the contralateral lung. \textbf{Clinical Presentation and Intervention:} A 19-year-old, previously healthy male presented with acute respiratory distress, left pleuritic chest pain, fever and hypoxemia. Clinical findings, laboratory and radiological examinations including pulmonary angiogram were consistent with the diagnosis of left pneumonia complicated with parapneumonic pleural effusion and right upper lobe PAVM. The patient was intubated and ventilated because of persistent hypoxemia. He was successfully treated by percutaneous transcatheter embolization. \textbf{Conclusion:} This case shows that percutaneous transcatheter embolization is a safe and effective first option for the treatment of PAVM.

Introduction

Pulmonary arteriovenous malformation (PAVM) is an abnormal communication between the pulmonary arteries and the pulmonary veins. It occurs twice as often in women as in men, and is usually congenital [1]. It may occur as a primary disease in the lung or in association with hereditary hemorrhagic telangiectasia (HHT) in 70\% of the cases [1]. It is classified as a simple PAVM when there is a single feeding artery and a single draining vein (80–90\% of cases), and complex PAVM when there are 2 or more feeding arteries or draining veins (10–20\% of cases) [2]. It affects the lower lobes of the lung in 50–70\% of cases [1]. It is a unilateral disease in 75\% of patients, and multiple lesions are seen in 36\% of patients. Individual PAVM is typically 1 to 5 cm in diameter but can be as large as 10 cm [1]. Lesions smaller than 2 cm are asymptomatic. PAVM associated with HHT is inherited and is present at birth, but it seldom manifests clinically until adult life, after the vessels are subjected to pressure over several years [1]. It is present between the fourth and sixth decades of life with epistaxis, dyspnea, hemoptysis, polycythemia or a stroke. Clinical examination may show superficial telangiectasia on the face, mouth, chest and upper extremities, finger clubbing, cyanosis and murmur or bruit can be heard over the site of PAVM [3, 4]. We report a case of PAVM affecting the right upper lobe.
Case Report

A 19-year-old male patient, previously healthy and symptomless before admission, presented with a 4-day history of increasing breathing difficulty, left pleuritic chest pain and fever. Physical examination showed reduced air entry in the left lower chest, absence of bruit or murmur over the right chest wall and no evidence of clubbing or cyanosis. Clinical diagnosis of pneumonia in the left lower lobe was made. Chest X-ray and computed tomography (CT) showed the left lower lobe collapse consolidation, left pleural effusion and a shadow in the right upper lobe suggestive of varix (fig. 1, 2).

Our patient was started on broad-spectrum intravenous antibiotics, and a chest tube was inserted to drain the pleural fluids on the left side. Remarkable results on laboratory work-up included low arterial blood O₂ saturation of 85% in 10 l of O₂, PaO₂ 7.8 kp, PCO₂ 3.39 kp and hemoglobin of 11.8 g/dl. In spite of supportive treatment, he continued to deteriorate and required intubation and ventilation because of worsening of breathing and persistent hypoxemia. Oxygen saturation was 70% with FiO₂ of 80–90% and positive end expiratory pressure of 15 cm H₂O (table 1). Echocardiogram was done, which indicated the possibility of right to left intrapulmonary shunt. Preoperative contrast chest CT scan suggested PAVM in the right upper lobe. Digital subtraction pulmonary angiography was performed to determine the site and size of PAVM (fig. 3). Brain CT scan showed no intracranial malformations. The patient underwent percutaneous transcatheter embolization of a large single feeding artery draining to the left atrium using amplatzer ductal occluder 6–8 and ASD amplatzer occluder 7 mm (fig. 4). Following embolization, blood gases showed a dramatic improvement from baseline (table 1). The patient was extubated shortly after embolization and was discharged 6 days later. Follow-up visit at 4 weeks showed no dyspnea on exertion and arterial blood gas oxygen tension at room air was normal with O₂ saturation of 98% (table 1).

Discussion

PAVM is a benign disease with potentially life-threatening complications from paradoxical embolization leading to cerebral vascular accident, cerebral abscesses, massive hemoptysis and hemothorax [4]. It can occur in the form of single or multiple lesions affecting one or both lungs, causing right to left shunt of the circulating blood without participating in gas exchange and leading to hypoxemia [1–5]. The severity of presentation depends on
the size and number of PAVM and the degree of shunt. It can vary from an absolutely asymptomatic state to severe hypoxia and heart failure. The presence of a bruit on chest auscultation that increases with inspiration, history of epistaxis and family history of HHT are key factors in suspecting the diagnosis, as the PAVM is found in 33% of cases with HHT [5]. In our case, the patient was symptomless before admission until he developed left parapneumonic effusion, which led to sudden deterioration in his condition. PAVM was discovered incidentally. In the majority of cases, chest radiograph will show a uniformly rounded mass, lobulated and well defined in the lower lobes, and the feeding vessel can be traced all the way to the hilum [5]. Contrast echo is a useful noninvasive and very sensitive procedure to detect right to left shunt and is used to screen families with HHT for PAVM [6]. It shows a delay of three to eight cardiac cycles before contrast is visualized in the left atrium [1]. Contrast CT scan is highly sensitive in detecting PAVM with its exact location. Pulmonary angiography remains the gold standard tool that identifies the pulmonary architecture very well and is mandatory prior to any treatment.

Once PAVM is recognized, it should be treated to prevent its complications. The other indications for treatment are progressive PAVM enlargement and symptomatic hypoxemia. The two modalities of treatment are surgical resection or embolization. Prior to 1977, surgical resection was the procedure of choice; ligation, local excision, segmentectomy, lobectomy or pneumonectomy were performed in most cases [1]. Interventional radiology has led to the successful use of percutaneous transcatheter embolization by coil or balloon for multiple PAVM as in our case. This technique is less invasive than surgery and has a lower complication rate [4]. Embolization therapy is the preferred method of treatment as it avoids major surgery, general anesthesia, loss of pulmonary parenchyma and is the treatment of choice in patients with high operative risk. It is useful for patients with multiple or bilateral PAVM and can be repeated if necessary. Although surgical resection in PAVM is associated with negligible mortality and minimal postoperative complications, it still requires longer hospitalization, and expenses associated with it are high. Moreover, thoracotomy and lung resection carry a risk of morbidity. Hence, surgical resection is used mainly in cases where an experienced interventional radiologist is not available or the patient has an allergy to the contrast media [1–4].

Patients with PAVM should be followed up routinely by arterial blood gases measurements at 1 month and 1 year after embolization therapy and spiral CT scan of the chest every 3–5 years to assess growth of small PAVM, which can be a source of new strokes or cerebral abscesses.

Fig. 3. Digital subtraction pulmonary angiography showing right upper lobe PAVM.

Fig. 4. Digital subtraction pulmonary angiography showing the amplatz occluding right upper lobe PAVM (arrow).
es [1]. The long-term results of both modalities of treatment are very good. To date no mortality associated with embolization of PAVM has been reported, and the complications have been infrequent and self-limited, with pleuritic chest pain being the commonest one.

**Conclusion**

This case shows that percutaneous transcatheter embolization is a safe method of treatment for complex PAVM.

**References**