Pulmonary Arteriovenous Malformation Treated with Embolotherapy

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Abstract
Pulmonary arteriovenous malformations (PAVM) represent pulmonary vascular anomalies and in majority of cases are congenital in origin. We report a case that presented with predominant complaints of dyspnoea on exertion and cyanosis. Clinical examination revealed a bruit in the left lower interscapular and infrascapular areas which led us to suspect PAVM. The diagnosis was established on the basis of computed tomography (CT) thorax with three-dimensional virtual reconstruction technology (3D VRT) images and pulmonary angiography. Patient was successfully treated by embolotherapy using steel coils, which resulted in 90% exclusion of the aneurysm from the rest of the systemic circulation.

INTRODUCTION
Pulmonary arteriovenous malformations (PVAM) are rare pulmonary vascular anomalies. They can be defined as direct communications between the branches of pulmonary artery and pulmonary veins, without intervening pulmonary bed. These lesions usually represent congenital malformation, with the exception of very rare acquired cases. The fundamental defect in pulmonary arteriovenous malformation, is right to left shunt from the pulmonary artery to the pulmonary vein, the degree of shunt is what determines the clinical effects on the patient. We report a case of pulmonary arteriovenous malformation treated successfully with pulmonary arterial embolisation using steel coils.

CASE REPORT
A 55 year old female, housewife, presented with symptoms of left sided, dull aching intermittent non-pleuritic chest pain and dyspnoea on exertion for the past 10 years. On examination she had a pulse rate of 85 per minute, blood pressure of 150/90 mm of Hg and oxygen saturation of 85% on pulse oximetry. There was grade - III clubbing with central cyanosis. Respiratory system examination revealed ‘bruit’ in the left lower interscapular and infrascapular areas. Cardiovascular, abdominal and neurological examinations were unremarkable.

Investigations showed haemoglobin of 16.8 gm/dl, packed cell volume of 51%; white blood cell count of 13,600 per cu mm; polymorphs 86%, lymphocytes 13%, and eosinophils 1%. Other biochemical parameters were within normal limits. Arterial blood gas estimation showed pH-7.348, PCO₂-41.1, PO₂ - 47.6, and HCO₃ - 22.0 mmols/L. Chest radiograph (Fig. 1) showed a sharply defined rounded opacity of uniform density in the left lower zone.

The presence of bruit on clinical examination and evidence of a well-defined mass on chest radiography raised suspicion of pulmonary arteriovenous malformation and the patient was subjected to computed tomography (CT) thorax (plain and with contrast) with three dimensional virtual reconstruction (3-VRT) images and pulmonary angiography. The computed tomography image revealed presence of an
intensely enhancing soft tissue density lesion in the posterior basal segment of the left lower lobe. There was no evidence of calcification. A feeding vascular channel was also present. The findings were consistent with a vascular malformation, most probably an arteriovenous malformation in the posterior segment of left lower lobe. Two-dimensional contrast echocardiography (2-D Echo) with agitated saline showed air bubbles, which rapidly appeared in the right atrium and gradually dissipated in the pulmonary circulation consistent with PAVM. Pulmonary angiography was later performed. Pulmonary angiogram revealed a large pseudoaneurysm with arteriovenous fistula arising from the left posterior basal and lateral basal pulmonary arterial branches (Fig. 2). Rest of the pulmonary vasculature was normal. The left posterior basal and lateral basal branches were coiled using four steel coils. Post-coiling angiogram revealed 90% exclusion of the pseudoaneurysm from the circulation (Fig. 3). Post-procedure the patient was symptomatically better. The arterial blood gas analysis revealed a pH of 7.430, PCO₂ - 38.5, PO₂ - 64.7, SaO₂ - 87.3 and HCO₃ - 25. The patient continues to remain asymptomatic 3 months following embolotherapy.

**DISCUSSION**

PAVM can be congenital or acquired. More than 80% of pulmonary arteriovenous malformations are congenital and of these 47-80% are associated with Osler-Weber Rendu disease or hereditary haemorrhagic telangiectasia (HHT). Presence of HHT in a patient may be of prognostic value since the patients with coexisting HHT tend to have worse symptomatology, multiple arteriovenous malformations, rapid disease progression and higher complication rate. Our patient did not have HHT. The incidence of PAVM is 2-3 per 1,00,000 population. The male to female ratio varies from 1:15 to 1.8 in several series. The age at the first presentation ranges from newborn to 70 but the majority of cases are diagnosed in the first three decades of life.

Solitary PAVM are commonly seen in lower lobes, the left lower lobe being the most common location (as in our case), followed by right lower lobe, left upper lobe, right middle lobe and right upper lobe.

The patient may be asymptomatic. However, most common presenting symptom is dyspnoea on exertion, which is seen in 31% to 67% of patients. Other symptoms include epistaxis, haemoptysis, palpitation, chest pain and cough. Neurological symptoms like headache, vertigo, paresis, numbness, paresthesia, syncope or confusion may also be found. In addition because the PAVM bypasses the capillary bed, the lung loses its filter function, thus allowing emboli and bacteria to pass directly into the systemic circulation resulting in stroke or cerebral abscess. The most common physical findings are cyanosis, clubbing and pulmonary vascular bruit. Other findings include polycythemia, telangiectasia. Diagnostic investigations include chest radiograph, contrast echocardiography, radionuclide perfusion lung scan, contrast-enhanced computed tomography, magnetic resonance imaging and pulmonary angiography which remains the gold standard in the diagnosis of PAVM. Pulmonary angiography not only confirms the diagnosis, but also defines the angioarchitecture of pulmonary vasculature, which is necessary before therapeutic embolisation.

The morbidity associated with PAVM is up to 50% in untreated patients compared with 3% in patients who receive treatment. The goals of treatment are three fold: 1) improvement of dyspnoea/hypoxemia, 2) prevention of lung haemorrhage and most important, 3) the prevention of neurological sequelae.

The current preferred treatment for the majority of patients with PAVM is percutaneous embolotherapy using coils or...
balloons; this method has largely replaced surgical intervention. Embolotherapy being less invasive and easy to repeat has definite advantages over surgery. The complications of embolotherapy include self-limited pleurisy, air embolism and paradoxical embolisation of the device. Our patient did not experience any “on-table” complication and was symptomatically better post-procedure. Regular follow up is necessary to document resolution and to monitor interval growth of small PAVM. This case report highlights the importance of meticulous clinical examination, which lead us to the suspicion, and subsequently the treatment of PAVM.

REFERENCES


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