

Pulmonary arteriovenous malformation: a rare cause of dyspnoea on exertion

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DESCRIPTION

Pulmonary arteriovenous malformation (PAVM) is caused by the abnormal connection between pulmonary arteries and venous systems. First described in 1897 by Churton, PAVM is a rare disease. In an autopsy study in 1953 at Johns Hopkins, only 3 cases of PAVM were detected out of 15 000 consecutive autopsies.¹ PAVM are present in 15–50% of patients with hereditary haemorrhagic telangiectasia (HHT),² also known as Osler-Weber-Rendu disease; however 70% cases of PAVM are associated with HHT.¹

A 69-year-old woman with a history of hypertension, diabetes, dyslipidaemia and hypothyroidism

presented with a history of progressively increasing dyspnoea on exertion over past 3 months. She denied any history of chest pain, palpitations, presyncope or syncope. At admission pulse oximetry showed 83–90% saturation on room air, correcting to 95–96% on supplemental oxygen. Physical examination was significant for a faint bruit on right infrascapular region. Arterial blood gas (ABG) showed elevated Alveolar-arterial (A-a) gradient of 44.1 mm Hg (normal for age was 17 mm Hg). Chest X-ray (CXR) showed a 3 cm tubular opacity in the right lower lobe (figure 1). CT angiogram confirmed the diagnosis of pulmonary arteriovenous malformation (AVM; figure 1). Two-dimensional

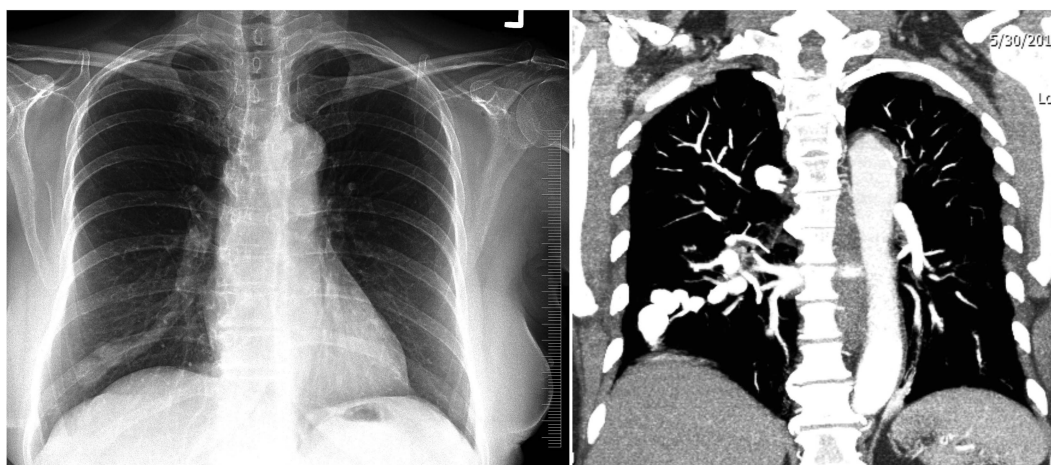


Figure 1 A 3 cm tubular opacity in the right lower lobe seen on the chest X-ray with connecting vessel from the hilum. Findings are replicated on the CT angiogram of the chest.



Figure 2 Magnified view of digital subtraction pulmonary angiogram (left) demonstrating the angioarchitecture of the pulmonary arteriovenous malformation. After embolotherapy using multiple 0.035 inch coils (right) the pulmonary arteriovenous malformation is totally occluded.



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transthoracic echocardiogram showed normal ejection fraction and no anatomical abnormality. She underwent pulmonary angiography and subsequent occlusion of the PAVM with multiple

0.035" Mreye embolisation coils (figure 2), leading to closure of PAVM. Postprocedure her saturation rose to 96–98% on room air and exercise tolerance continues to improve at follow-up. Screening colonoscopy did not show any AVM and she is awaiting screening CT angiogram of the brain and esophagoscopy for the detection of occult AVM.

Learning points

- ▶ Dyspnoea is the most common lung symptom in patients with pulmonary arteriovenous malformation (PAVM).³ Hypoxaemia at presentation along with incomplete correction with supplemental oxygen and elevated Alveolar-arterial gradient should alert the clinician towards a right to left shunt and a possible PAVM.
- ▶ Transthoracic contrast echocardiography with agitated saline is the best screening test to detect PAVM whereas chest X-ray is more specific in detecting PAVM.² All positive finding should be confirmed with multidetector thoracic CT (MDCT) with thin cut (1–2 mm) reconstructions.² In the present era, the utility of pulmonary angiography is restricted to treatment only.^{1 3}
- ▶ Treatment must be offered to all symptomatic patients whereas asymptomatic patients, especially children should be evaluated on case to case basis.² Embolisation is the intervention of choice whereas surgery is reserved only for patients with fatal haemorrhages.² Follow-up with MDCT at 6–12 months must be performed to detect reperfusion of PAVM and opening up of occult PAVM.²

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Competing interests None.

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