Case report

Hypoxia and fractions: an example of how to uncover this uncommon connection

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Learning Point for Clinicians

Anatomic shunts such as those associated with congenital heart disease and pulmonary arteriovenous malformations complete the wide differential of hypoxia. When the common causes of hypoxia are ruled unlikely by investigations these serious and treatable connections need to enter the differential by, as this case highlights, calculating a shunt fraction.

Pulmonary arteriovenous malformations (PAVM) are rare abnormal communications between arterial and venous vessels. They affect 1 in 15,000–24,000 within a large tertiary centre (The Mayo Clinic, New York) recording 4.5 cases per year.1,2 Despite their rarity they can result in life-threatening complications including cerebral abscess and stroke.3 Diagnosis requires a high clinical suspicion. It is estimated that 70% are associated with hereditary haemorrhagic telangiectasia, which can present with epistaxis, mucocutaneous telangiectasia and gastrointestinal bleeding.4 Idiopathic PAVMs are harder to diagnose, with reviews reporting that half of patients were asymptomatic.2–4 Dyspnoea, haemoptysis and chest pain can be presenting symptoms. We present a case of a PAVM presenting to the general medical on call to help highlight useful steps in the approach to unexplained hypoxia.

A 72-year-old lady was referred to the medical assessment unit from her general practitioner regarding episodes of acute onset, left-sided chest pain. This came on when she was lifting some shopping bags and was described as pleuritic in nature. She did not report dyspnoea but, since returning from a holiday in Cyprus, noted a 3-month period of lethargy and tiredness. There were no other particular symptoms. She was a lifelong non-smoker and had a background history of hypertension, mild asthma, carpal tunnel and knee surgery. Her drug history includes nifedipine, fluoxetine, cetirizine, simvastatin and inhaled salbutamol and inhaled fluticasone/salmeterol. Heart sounds were normal and lung fields were clear to auscultation. Observations showed that she was hypoxic with oxygen saturations of 90% on air. Her D-Dimer test was negative and her Haemoglobin was 132 g/l. Renal, liver and biochemical profile were normal. Lung fields on chest X-ray (CXR) were unremarkable and electrocardiogram showed sinus rhythm at 72 beats per minute.

An abnormal finding on CXR is a common presentation of PAVMs, manifesting usually as well defined round opacities.2 As a result of the CXR being a common presenting feature there has been a potential overestimation of its sensitivity in diagnosis.2 Consensus from major reviews suggests the best initial investigations to rule out a PAVM is therefore the 100% oxygen rebreathing method.2–4 This involves a patient breathing 100% oxygen for 15–20 min to allow the deduction of a shunt fraction. The shunting of venous blood to the arterial circulation is normally <2% of cardiac output. This is accounted for by bronchial, mediastinal and thesbian veins.4 Our patient’s shunt fraction, calculated using a shunt fraction nomogram, was 25% (at room air, pH 7.41, pO2 56 mm Hg, pCO2 40 mm Hg, alveolar-arterial
gradient 43.7 mm Hg, SaO2 90.1%; at FiO2 100%, pH 7.40, pO2 92 mm Hg, pCO2 43 mm Hg, alveolar-arterial gradient 567 mmHg, SaO2 97.0%).5

Finding a large shunt fraction prompted a contrast-enhanced computed tomography of the thorax, which revealed a large left-sided PAVM. Our patient subsequently underwent bubble contrast echocardiography, which showed a large number of bubbles in the left atrium after five beats. The agitated saline in control cases should disappear in the pulmonary circulation. Recent studies exploring the grading of PAVM via echocardiogram class the shunt as very severe with complete opacification of the left ventricle with saline bubbles (Figure 1).6 For the acute physician, when hypoxia remains unexplained, the correct use of the 100% oxygen rebreathing method will help rule in or out a rare but important differential.2–4

Conflict of interest: None declared.

References