Hypoxaemia in a pregnant lady

Case report

A 35-year-old pregnant lady (gravidity: 8; parity: 7) was referred to our hospital at gestational age of 32 weeks for evaluation of refractory hypoxaemia; she received the diagnosis of possible pulmonary embolism and was given intravenous heparin with no improvement.

She had a 2-month history of progressive shortness of breath on both exertion and rest, associated with cough productive of frothy red sputum and sometimes frank haemoptysis.

On presentation to our hospital, the patient was found to be tachypnoeic (respiratory rate of 22 breaths·min$^{-1}$) and hypoxaemic (76% on room air), heart rate and blood pressure were within normal limits. On physical examination, she was found to have clubbing of both hands and feet, normal heart and lung examinations and gravid uterus appropriate for her gestational age of 32 weeks, she had no skin or mucosal telangiectases.

She was immediately admitted to the medical intensive care unit and connected to jet nebuliser at inspiratory oxygen fraction of 100%. Oxygen saturation was 88% in sitting position and rose to 92% upon lying down (a phenomenon called orthodeoxia).

Task 1
What is the significance of the rising saturation upon lying flat and what are the possible aetiologies? What is the difference between orthodeoxia and platypnoea?
Electrocardiography and chest radiography were unremarkable; blood tests were unremarkable apart from a D-dimer of 800 ng·mL⁻¹ (normal up to 500 ng·mL⁻¹). Because the patient was pregnant, we opted to perform an echo bubble study shown in videos S1 and S2 [1].

In light of these findings a diagnosis of intrapulmonary arteriovenous malformation was suspected. Two days later the patient underwent Caesarean section under spinal anaesthesia, and a rather healthy but premature baby girl was born. Following delivery, chest computed tomography pulmonary angiography (CTPA) was performed (figure 1).

**Task 2**
What does the echo bubble study in videos S1 and S2 show?

**Answer 2**
Normally, agitated normal saline microbubbles get filtered-out in the pulmonary capillaries, in this echo bubble study (long parasternal axis view), microbubbles escaped the pulmonary capillary network and appeared in the left atrium within three cardiac cycles, otherwise normal right ventricle, left ventricle, valves and no visible ventricular septal defect or atrial septal defect. This is clearly a positive study.

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**Task 3**
What do the CTPA images show?

**Answer 1**
This indicates the presence of an anatomical, functionally significant cardiovascular or intrapulmonary defect that increases the positional right-to-left shunting. When the subject sits up, gravity augments blood flow through the shunt causing more desaturation. Orthodeoxia is defined as a decrease in the arterial oxygen tension (by ≥4 mmHg) or arterial oxyhaemoglobin desaturation (by ≥4%) when the patient moves from a supine to an upright position, which is improved by returning to the recumbent position. Platypnoea is shortness of breath that accompanies the desaturation in the upright position.

**Figure 1** a–c) CTPA images.
For further delineating the nature of this tuft of vessels, a pulmonary angiogram was performed (video S3).

**Answer 3**
The CTPA images show an abnormal tuft of vessels in the right lung lower lobe (arrow), and enlarged right lower pulmonary vein (♯) (figure 2).

**Task 4**
What does the pulmonary angiogram (video S3) show?

**Answer 4**
The pulmonary angiogram shows an abnormal connection between the right lower lobar pulmonary artery and large tortuous right lower pulmonary vein. No other abnormal connections were found on the same side or on the left side.

**Task 5**
What is the final diagnosis? What are the therapeutic options?
Discussion

PAVMs are low-resistance, high-flow abnormal vascular structures that most often connect a pulmonary artery to a pulmonary vein, bypassing the normal pulmonary capillary bed and resulting in an intrapulmonary right-to-left shunt. They are uncommon in the general population and more common among women compared with men. They can be inherited or sporadic, vary in their number, distribution, and size, but are still an important entity in the differential diagnosis of common pulmonary symptoms such as cough and shortness of breath [2]. They can carry significant morbidity and mortality, and thus should be treated [3].

It is probable that recurrent pregnancies stimulated growth of the PAVM in this patient. [4]; several factors encourage growth of PAVM during pregnancy [5].

First, a myriad of haemodynamic changes occur, plasma volume expands as early as the fourth week of gestation and peaks at 28–34 weeks. Cardiac output also rises 30–50% above baseline during normal pregnancy, this leads to increased pulmonary blood flow, preferentially across the low resistance PAVM. The increased blood flow across the PAVM and causes its dilatation.

Second, progesterone causes smooth muscle relaxation, and leads to arterial and venous dilatation which further decreases resistance and augments blood flow across the PAVM, thus increasing its size.

This article has supplementary material available from breathe.ersjournals.com

Conflict of interest

None declared.

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