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Hypoxaemia in a pregnant lady

Case report

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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?

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A pregnant lady at gestational age of 32 weeks presents with refractory hypoxaemia in this case report http://ow.ly/MM9C301zeVx

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.

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].

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

Answer 2
Normally, agitated normal saline micro-bubbles 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.

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 3
What do the CTPA images show?

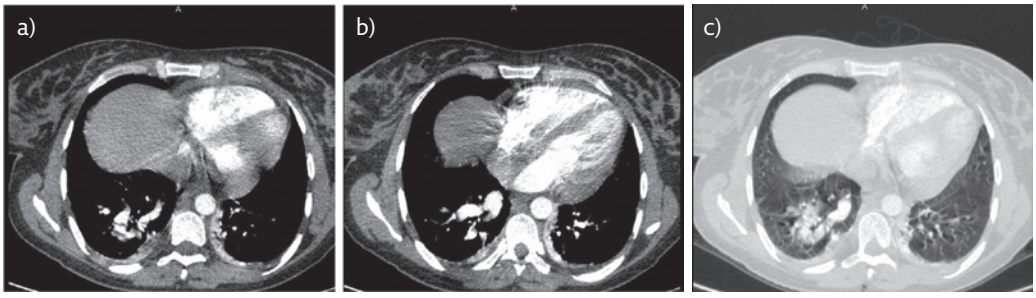

Figure 1 *a–c) CTPA images.*

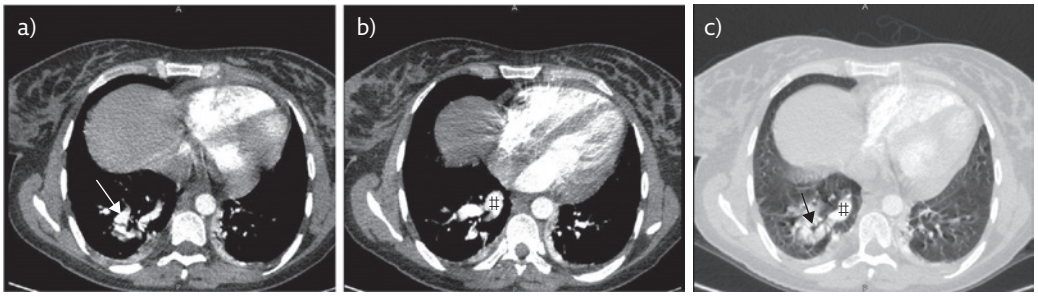

Figure 2 *a–c) An abnormal tuft of vessels in the right lung lower lobe (arrows), and enlarged right lower pulmonary vein (#) are shown in the CTPA images.*

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).

For further delineating the nature of this tuft of vessels, a pulmonary angiogram was performed (video S3).

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?

Answer 5

A diagnosis of solitary pulmonary arteriovenous malformation (PAVM) was made, and the patient underwent right lower lobe resection (figure 3). Following surgery, her saturation improved to 97% on room air. Other therapeutic options include angiographic occlusion of the feeding arteries to a PAVM under fluoroscopic guidance.

Repeat bubble echo and CTPA were negative for shunts (video S4 and figure 4) (arrow indicates the chest tube with residual post-operative pneumothorax). Histopathological examination of the resected lobe showed only the abnormal connection seen on the imaging, with no evidence of other pathologies.

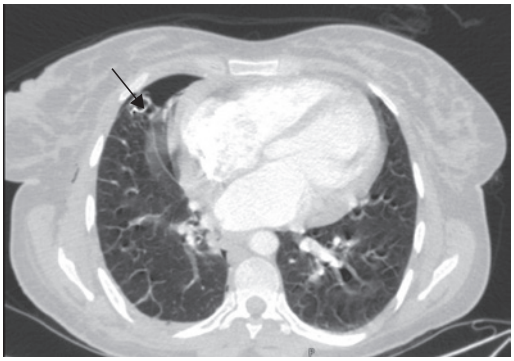

Figure 4 *Post-operative chest CT. Arrow indicates chest tube with residual post-operative pneumothorax.*

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].

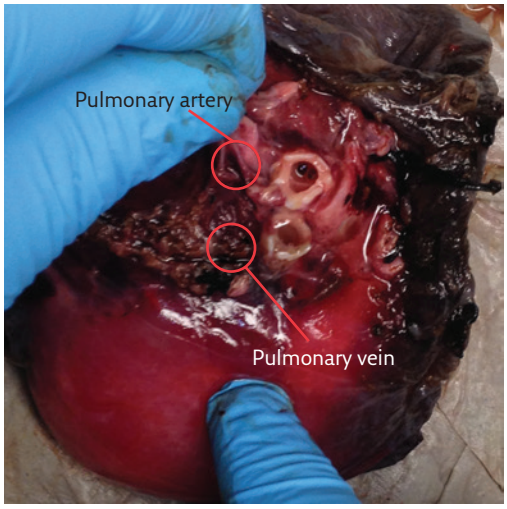

Figure 3 *Gross pathological specimen with pulmonary artery and pulmonary vein highlighted.*

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.

References

1. Gossage JR. The role of echocardiography in screening for pulmonary arteriovenous malformations. *Chest* 2003; 123: 320–322.
2. Cartin-Ceba R, Swanson KL, Krowka MJ. Pulmonary arteriovenous malformations. *Chest* 2013; 144: 1033–1044.
3. Swanson KL, Prakash UB, Stanson AW. Pulmonary arteriovenous fistulas: Mayo Clinic, 1982-1997. *Mayo Clin Proc* 1999; 74: 671.
4. Khurshid I, Downie GH. Pulmonary arteriovenous malformation. *Postgrad Med J* 2002; 78: 191–197.
5. Esplin MS, Varner MW. Progression of pulmonary arteriovenous during pregnancy: case report and review of the literature. *CME Rev Obstet Gynecol Surv* 1997; 52.