INTRODUCTION

Although Pulmonary Arteriovenous Malformation (PAVM) is a relatively rare disorder but it is probably the most common anomaly of the pulmonary vascular tree. Since the first reported case in 1897, more than 500 cases have been reported in the literature. The natural history of this rare entity is not completely understood. PAVMs can be either congenital or acquired. A sizable number of patients are asymptomatic. The commonest complaint of this entity is dyspnoea. Other complaints include platypnea and orthodeoxia but the patients are not usually symptomatic from hypoxemia unless lesions are ≥2 cm. Other clinical features include clubbing, cyanosis, haemoptysis or haemothorax. Pregnancy may be a risk factor for haemoptysis in patients with PAVMs by causing growth of PAVMs (particularly in the last trimester) due to increased cardiac output, blood volume, and hormone-related changes of the vasculature. We hereby report a 28 year old lady presenting for the first time in her 3rd trimester of pregnancy with cyanosis and clubbing and was subsequently diagnosed as having Pulmonary Arteriovenous Malformation.

CASE REPORT

A 28 year old pregnant lady was admitted to the antenatal ward with progressive dyspnoea in supine position. The patients’ relatives also noticed bluish discolouration of her lips and nail. On examination, she had minimal cyanosis and clubbing of fingers. There was no icterus, oedema, JVP was not raised and there was no lymphadenopathy. The patient preferred to lie in the supine position. The percentage of her peripheral oxygen saturation (SpO₂) in supine posture was 96% which fell to 85% as she assumed a standing position and was associated with dyspnoea. Her dyspnoea improved markedly and her SpO₂ again reached 96% a few minutes after she lied down. Cardiovascular system examination revealed normal heart sounds and a soft pulmonary ejection murmur. There was bilateral vesicular breath sounds with a continuous murmur heard in the right infrascapular region. On per abdominal examination, there was no organomegaly, uterus was gravid with a single foetus at 36 weeks, floating foetal head, positive foetal heart sounds and normal foetal movements. A complete blood count was ordered which was unremarkable as was her liver function test, renal parameters, electrolytes and routine urine analysis. The patient’s symptoms worsened progressively as she approached term necessitating her to be shifted to Critical Care Unit of our hospital a few days before delivery. An echocardiographic screening was done which revealed mild dilatation of left atrium and ventricle with no intracardiac shunt. Suspecting a pulmonary arteriovenous malformation, an agitated saline bubble contrast echocardiographic study was done which showed appearance of the bubbles in the left side 5-6 cycles after...
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PAVMs expand during pregnancy because of increase in blood volume, cardiac output, and venous distensibility and may be a potentially life-threatening cause of haemoptysis during pregnancy. Progesterone causes smooth muscle relaxation leading to arterial and venous distension and subsequent decrease in resistance across the AVM leading to progression in size and shunt fraction. PAVMs may initially present in pregnancy with fatigue, dyspnoea and musculoskeletal symptoms seen in late pregnancy. The most common presenting symptoms in pregnancy are dyspnoea and chest pain sometimes resulting in a mistaken diagnosis of pulmonary embolism. Other common symptoms include haemoptysis and haemothorax. Pregnancy is associated with increase in steroid hormone synthesis resulting in an increased incidence of spontaneous haemothorax secondary to intrapleural rupture of PAVM. Esplin et al summarized nine previous case reports of pulmonary arteriovenous malformations in pregnancy. In our patient, the presenting symptoms were progressive dyspnoea, cyanosis, platypnea and orthodeoxia and she was symptomatic for the first time in pregnancy and her symptoms gradually waned off after delivery by caesarean section.

Orthodeoxia is a decrease in partial pressure of oxygen in arterial blood (PaO) or arterial oxygen saturation (SaO) that occurs when one assumes an upright position from the supine position. The fraction of cardiac output that shunts right-to-left is elevated in patients with PAVM; normal values are less than 5%. The shunt fraction is most accurately assessed by using the 100% oxygen method, which involves the measurement of PaO2 and SaO2 after the patient breathes 100% oxygen for 15 to 20 minutes. A shunt fraction of more than 5% has a sensitivity of 87.5% and a specificity of 71.4%. The chest X-Ray in PAVM show round or oval sharply defined mass of uniform density, frequently lobulated, and ranging in size from 1–5 cm in diameter; two thirds are located in the lower lobes. A plain chest radiograph may show a connecting vessel radiating from the hilum.

Contrast echocardiography involves injection of agitated saline or dye into a peripheral vein; it is extremely sensitive in detecting left-to-right shunt but it does not provide quantitative or anatomic detail of the shunt. In patients without right-to-left shunt, an air bubble or dye may rapidly appear in the right atrium and then gradually dissipate as the bubbles become trapped in the pulmonary circulation. In the case of intracardiac shunt, bubbles will be visualised in the left atrium within three to four cardiac cycle after their appearance in the right atrium. In contrast, a PAVM will demonstrate a delay of more than four cardiac cycles before the bubbles will be visualised in the left atrium as was seen in our patient. On occasion, if bubbles or contrast can be seen entering the left atrium through a single pulmonary vein, it confirms the ipsilateral anatomical localisation of the PAVM.

Radionuclide perfusion lung scan is a useful adjunct in the diagnosis and quantification of PAVM. Contrast enhanced computed tomography is a valuable tool in diagnosis and defining the vascular anatomy of PAVM. 3-D helical computed tomography allowed full analysis of 76% of PAVM, compared with only 32% by conventional unilateral pulmonary angiography. The use of magnetic resonance imaging to diagnose PAVM has been limited compared with that of computed tomography. Most lesions within the lung have a relatively long relaxation time and produce medium to high intensity signals. In contrast, PAVMs and aneurysms with rapid blood flow in the lesion result in a signal void and produce low intensity signal. Pulmonary angiography remains the gold standard in the diagnosis of PAVM. Angiography should be performed on all portions of the lung to look for any unsuspected PAVM and the source of intrathoracic or extrathoracic vascular communications. In our patient PAVMs were seen in the lower lobe of the right lung. Computed tomography and magnetic resonance angiography should be reserved for those patients who cannot undergo angiography or for the follow up of patients with a proved PAVM.

Figure 2: The Arteriovenous malformation in right lower lobe of lung draining into the Right lower pulmonary vein.
Since the first successful resection of PAVM in 1942, surgery was the only treatment available until 1978, when Taylor et al reported the first successful percutaneous embolisation. Currently, the preferred treatment for the majority of patients with a PAVM is percutaneous embolization using coils or balloons largely replacing surgical intervention with minimal morbidity and no mortality thus rendering radiological intervention as the first line of treatment for PAVM. Complications of embolization include pleuritic chest pain, pulmonary infarction, air embolism, device migration, myocardial rupture, cerebrovascular accident, vascular injury, early deflation of balloon, deep vein thrombosis, and pulmonary hypertension.\(^2\)

The appropriate mode of delivery in this group of patients is unclear but worsening hypoxemia, haemoptysis or haemothorax warrants caesarean section.\(^5\)

**CONCLUSION**

The unique feature in our case is that the patient presented for the first time in third trimester of pregnancy without any previous cardiovascular symptoms which is extremely rare. PAVM should be considered in the differential diagnosis of all pregnant women presenting with dyspnoea with cyanosis and clubbing on physical examination. Careful history taking and physical examination and echocardiography including bubble contrast study may help in arriving at a diagnosis in this rare disorder. 