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Case Report

Dyspnoea and Cyanosis in Pregnancy: An Extremely Rare Cause

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ABSTRACT

Pulmonary arteriovenous malformation first presenting with platypnea and orthodeoxia in the 3rd trimester of pregnancy is an extremely rare entity and poses a great diagnostic challenge to the treating physician. We hereby report a patient with pulmonary arteriovenous malformation presenting for the first time in third trimester of pregnancy diagnosed by pulmonary angiogram with subsequent improvement in symptoms post-delivery.

Key words: Pulmonary Arteriovenous Malformation, Pregnancy, Platypnea, Orthodeoxia, Pulmonary Angiogram.

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INTRODUCTION

Although Pulmonary Arteriovenous Malformation (PAVM) is a relatively rare disorder but it is probably the most common anomaly of the pulmonary vascular tree.[1,2] 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.[2] PAVMs can be either congenital or acquired.[2,3] 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.[2,4] 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 (SpO_2) 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 SpO_2 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

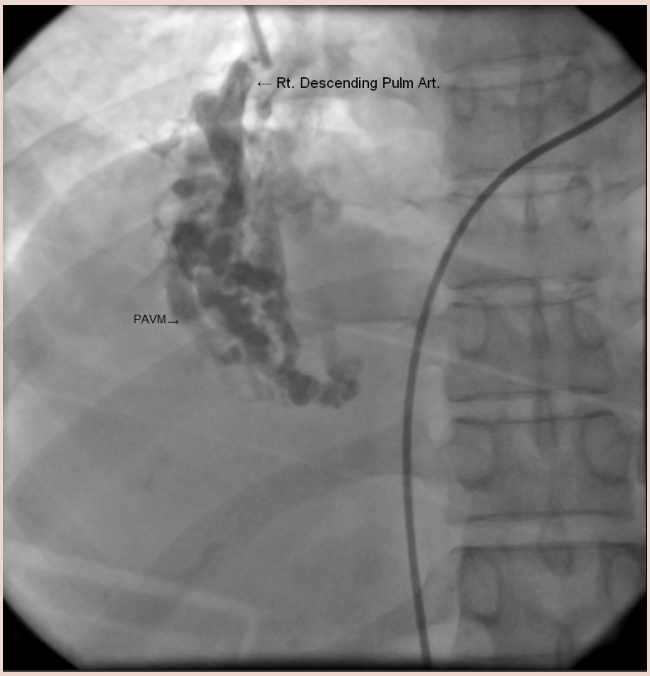

Figure 1: Judkins (Right) catheter being selectively engaged in descending branch of right pulmonary artery showing the feeding vessel and the arteriovenous malformation (AVM).

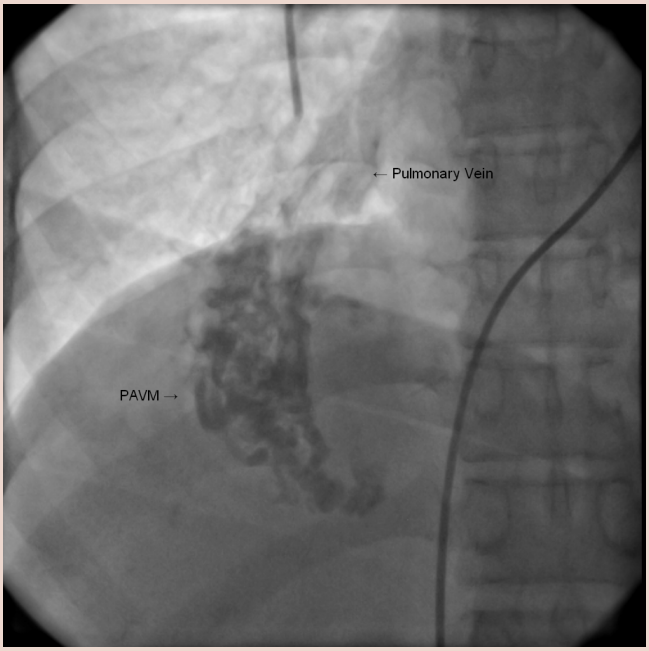

Figure 2: The Arteriovenous malformation in right lower lobe of lung draining into the Right lower pulmonary vein.

its appearance in the right sided chambers. The patient was managed conservatively with medications. She had an uneventful delivery by caesarean section under strict hemodynamic monitoring. Subsequent pulmonary angiogram confirmed the presence of large pulmonary arteriovenous communication in right lower lobe. Figure 1 shows a Judkins (Right) catheter being selectively engaged in descending branch of right pulmonary artery supplying the feeding vessel and the arteriovenous malformation (AVM). Figure 2 shows the AVM draining into the Right lower pulmonary vein. Following delivery the cyanosis and dyspnoea improved over time and the patient gradually became nearly asymptomatic 6 month after delivery. The patient refused any surgical or percutaneous intervention despite being repeatedly counselled for the same.

DISCUSSION

More than 80% of PAVMs are congenital, and of these 47%–80% are associated with Osler-Rendu-Weber disease or hereditary haemorrhagic telangiectasia (HHT).[2] Separate arterial and venous channels, interconnected by capillaries are created during the embryological development of the vascular system, which occurs between the 5th and 10th weeks of intrauterine life. Vascular malformations occur when there is any alteration in this process of differentiation of pulmonary vasculature.[3] PAVM in its acquired form usually occurs in cirrhosis (as a part of hepato-pulmonary syndrome), but has also been reported in patients with metastases of thyroid carcinoma and pulmonary schistosomiasis.[2,3]

Asymptomatic patients are common and account for between 13% to 55% of patients in different series.[2] PAVM, in the asymptomatic, non-pregnant patient, is diagnosed most commonly as an incidental finding on a routine chest x-ray. However, a classic triad of signs and symptoms of PAVM has been reported, consisting of dyspnoea, cyanosis, and clubbing. Other symptoms include epistaxis, haemoptysis, cough, palpitation and chest pain.[2] Chronic hypoxia associated with PAVM can result in cyanosis and clubbing. A shunt of at least 20% is required to produce hypoxia severe enough to result in these findings.[5]

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.[5]

Esplin *et al* summarized nine previous case reports of pulmonary arteriovenous malformations in pregnancy.[5] 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_2) or arterial oxygen saturation (SaO_2) that occurs when one assumes an upright position from the supine position. The fraction of cardiac output that shunts right-to-left circulation 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 PaO_2 and SaO_2 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%.[1]

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.[2]

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.[2,6] 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.[6] 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.[2,6]

Radionuclide perfusion lung scan is a useful adjunct in the diagnosis and quantification of PAVM.[2] 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.[2]

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.[2]

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.[2]

Since the first successful resection of PAVM in 1942, surgery was the only treatment available until in 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.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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