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RESEARCH ARTICLE

PULMONARY ARTERIOVENOUS MALFORMATIONS AT TERM PREGNANCY – A RARE CASE REPORT

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ABSTRACT

Pulmonary arteriovenous malformations (PAVMs) are rare congenital anomalies resulting from the direct communication between pulmonary arteries and pulmonary veins without the interposition of a capillary bed with an increased risk of severe complications such as rupture, haemothorax, and hypovolemic shock. We report a case of Complex type PAVMs with 34 week pregnancy presented with dyspnoea, cyanosis and underwent intrauterine death.

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INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are rare congenital anomalies resulting from the direct communication between pulmonary arteries and pulmonary veins without the interposition of a capillary bed. Approximately 10 to 20% of patients with symptoms of PAVMs are isolated sporadic cases, whereas the remaining consistent percentage is related to Hereditary Hemorrhagic Teleangiectasia (HHT), an autosomal dominant vascular disorder also known as Osler–Weber–Rendu Syndrome [1, 2]. Other rare causes are represented by trauma, malignancy, hepatopulmonary syndrome, and cardiac surgery [3]. Pregnancy has been considered as a precipitant factor for PAVMs; the reason is related to the increase of cardiac work and blood volume as well as the effect of progesterone on vessels during the gestation. In most of the cases, patients and pregnant women affected by PAVMs are asymptomatic, but when the clinical manifestations occur, they are often related to the right-to-left shunting and may include dyspnoea, hypoxia, and pulmonary hypertension [4]. Moreover, it has been reported that the presence of one or multiple PAVMs during pregnancy is associated with an increased risk of severe complications such as rupture, haemothorax, and hypovolemic shock [5].

Our study discusses a case of a symptomatic pregnant woman affected by a PAVM that caused hypoxia to our patient.

Case Report

A 25 yr old female with amenorrhoea of 34 weeks pregnancy (primigravida) referred to medicine department from gynaecology and obstetrics department with complain of insidious onset dyspnea on exertion and chest pain on right side from 5 days. She also complained of lower abdominal pain from 2 days. Per abdominal examination is suggestive of pregnancy of 34 weeks with cephalic presentation. Fetal heart sounds was not heard, hence ultrasonography abdomen for fetal wellbeing was done that suggestive of intrauterine death (IUD). Vaginal delivery was conducted with opinion of gynaecologist. Patient was dyspnoeic and cyanosed, chest pain on right side continued, so we planned further evaluation of our patient on the line of suspected pulmonary embolism. Her past history suggestive of recurrent epistaxis since childhood which spontaneously resolved by applying local pressure. Past history is not suggestive of dyspnea, cyanosis, seizures and haemoptysis. On physical examination, central cyanosis present without clubbing, JVP- Normal, nasal mucosa hyperemic (right side), nasal septum was healthy. No Peripheral edema. Vital signs – BP-100/70mmHg, Pulse rate-120/min Regular, Body Temperature- 98.6F , Respiratory Rate- 28/min, Pulse oximetric examination suggestive of Spo2 of 80 % on room air and 88% on Oxygen via 5litre facemask.

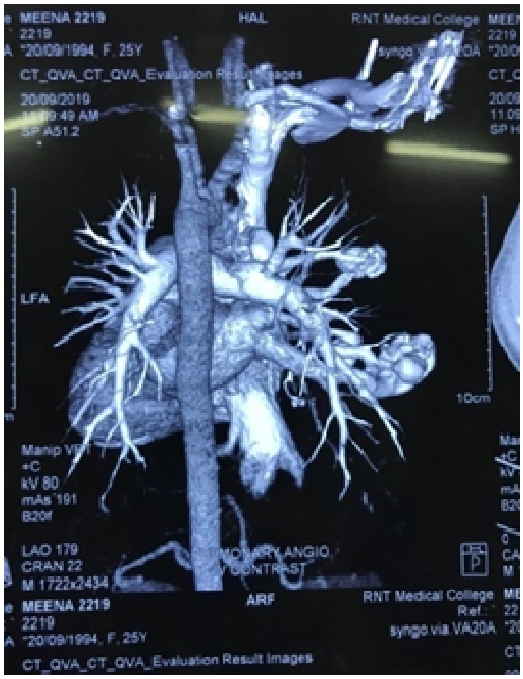

Fig.1. CT pulmonary Angiography showing multiple AV malformations (marked by arrows)

On systemic examination- On Chest auscultation , a continuos murmur heard in the axillary area on 4th and 5th intercostal space on right side of chest. No rhonchi and crepts audible. Cardiac sounds were normal. No organomegaly and no ascites detected on abdominal examination. On laboratory investigations , routine haemogram was within normal range. Liver function tests, renal function tests ,thyroid profile and D-Dimer was within normal limits. ECG suggestive of Sinus tachycardia, Chest X Ray PA view showed peripheral wedge shaped opacity in right lower zone.

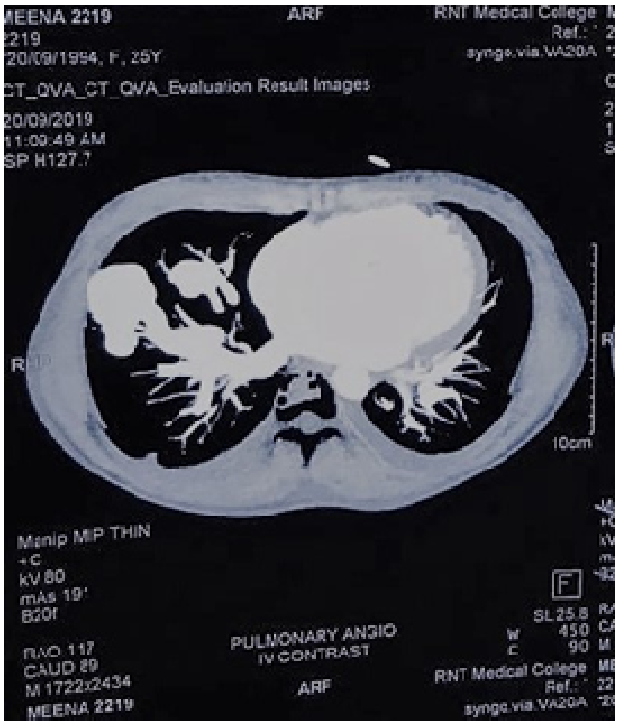

Fig .2. Showing AV malformations over right side (marked by arrow)

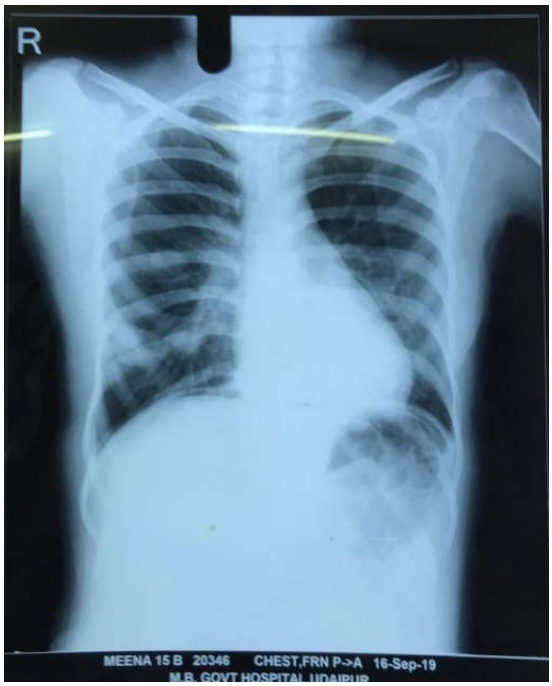

Fig.3:- Chest X ray showing Right Lower zone opacity(marked as arrow);

2D-Echocardiography suggestive of Global Hypokinesia of left ventricle, with severe LV systolic dysfunction, LVEF- 25-30%, moderate MR, No PAH, suggestive of Peripartum Cardiomyopathy. We planned CT Pulmonary Angiography to rule out pulmonary embolism that suggestive of "Multiple intensely opacified lobulated lesions in right lung , largest measuring (17 x 28mm) in right upper lobe and (24x29mm) in right lower lobe,supplied by branches of right pulmonary artery and drained by right superior and inferior pulmonary vein suggestive of Complex type Pulmonary Arteriovenous Malformations (PAVMs)".

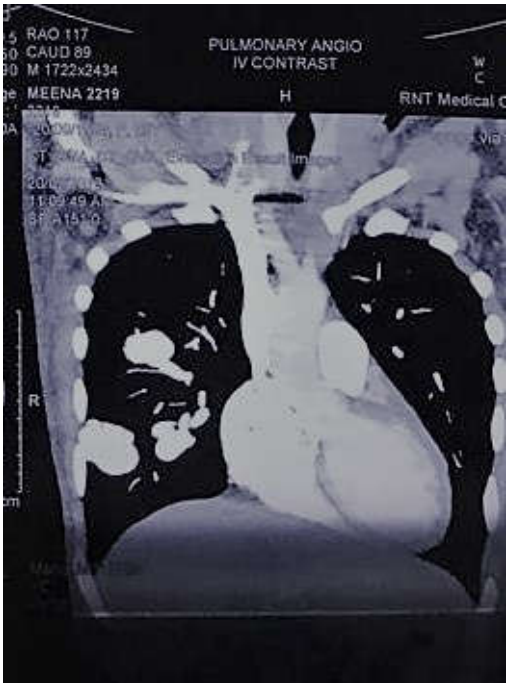

Fig.4. CT pulmonary Angiography showing AV malformation (marked as arrows)

TYPES OF PULMONARY AV MALFORMATIONS[7]

TYPES	INCIDENCE	CHARACTERISTICS
SIMPLE	75%	the commonest;has single segmental artery feeding the malformation
COMPLEX	20%	Multiple segmental feeding arteries
DIFFUSE	5%	it is characterized by hundreds of malformations.

DISCUSSION

Pulmonary arteriovenous malformations are result of anomalous connections between pulmonary arteries and pulmonary veins through a thin walled aneurysm. There is a recognized female predilection with F:M ratio ranging from 1.5 to 1.8:1. The estimated incidence is thought to be around 2-3 per 100,000[(6)] They cause right to left shunting of blood, and produce signs and symptoms when the shunt is large, (i.e. greater than 20 percent)[(7).] Churton first described pulmonary arteriovenous malformations in 1897[(15)] , which histologically vary from diffuse telangiectasiae to complex, large structures, consisting of a bulbous aneurysmal sac with feeding arteries and draining veins.. The remainders are supplied by the bronchial artery, internal mammary artery[(8)] and descending aorta, in decreasing order of incidence. PAVM in pregnancy is exceedingly rare. But when present they increase in size rapidly due to several physiological reasons. The increase in blood volume during a normal single pregnancy is 1570 ml[(9)] . This increase is most rapid in the second trimester. Furthermore, the cardiac output increases by as much as 30-50 percent during pregnancy[(10)] . The combined effect of increased blood volume and cardiac output result in heightened pulmonary blood flow and preferential shunting across the low-resistance PAVM, which in turn leads to its dilatation. The dispensability of the venous system increases by as much as 150 percent during pregnancy [(11)] . This is induced by the effect of progesterone, which causes smooth muscle relaxation. The resultant dilation of the vasculature further reduces the resistance across the AVM and leads to progressive increase in shunt fraction and size.

There is an increased incidence of AVM rupture associated with pregnancy, leading to hemothorax or hemoptysis. This is also attributed to the high level of steroid hormone circulation[(8)] . Myocardial infarctions also occur as a result of paradoxical emboli [(18)] . Pregnancy is a hazardous period for women with PAVMs. Absolute risks were quantified in two separate series of 487 [(19)] and 244 [(20)] pregnancies. In one series, 1.0% of pregnancies (95% confidence interval, 0.1–1.9) resulted in a major PAVM bleed, and the maternal death rate was 1.00% (95% confidence interval, 0.13–1.92%). There are also enhanced risks of pulmonary emboli and myocardial infarction with normal coronary arteries [(19)] . The gold standard method of investigation is angiography. This allows the localization of the pathology along with the identification and picturization of the feeding vessels prior to embolotherapy or surgical management. But it should be avoided in pregnancy due to significant radiation exposure nevertheless; angiography may be indicated in patients with pregnancy in whom non-invasive tests do not suggest the presence of pulmonary arteriovenous malformations. Pulmonary arteriovenous malformations with haemoptysis need urgent management either by TCE or surgical resection. There is scarce literature on the management and procedure of choice for PAVM in pregnancy. Though success rate of TCE is encouraging but recurrence of the lesion is not uncommon[(12)] and recurrence may be troublesome with advancing pregnancy. Therefore surgical resection, being more curable[(13)] , should be preferred in pregnancy and it also avoids dangerous exposure of radiation to fetus. Trans catheter embolization may be reserved for poor risk surgical candidates or for patients having multiple lesions, where curative resection is not possible[(14)]. In our case, surgical resection was done by Cardiothoracic and vascular surgery department and patient is stable on regular followups.

Glossary of Abbreviations

- **PAVM –** Pulmonary arteriovenous malformation
- **AVM –** Arteriovenous malformation
- **HHT-** Hereditary Haemorrhagic telengiectasia
- **IUD –** Intrauterine death
- **F:M-** Female : Male Ratio
- **TCE:** Transcathter embolization

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