*IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*
*e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 20, Issue 10 Ser.7 (October. 2021), PP 28-30*
*www.iosrjournals.org*

# Pregnancy Complicated by Pulmonary Arteriovenous Malformation and its Management

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***Abstract:***
*Pulmonary arteriovenous malformations (PAVM) are anatomical abnormalities of direct connection between pulmonary arteries and veins. Determinants like puberty, pulmonary arterial hypertension and pregnancy causes the malformation to increase in size. Right-to-left shunting manifests as pulmonary hypertension, dyspnoea and hypoxia. When pregnancy and PAVMs coexist, complications like haemothorax, rupture and hypovolemic shock are more prone to occur. Increased progesterone and estrogen levels in pregnancy results in venous distensibility which in turn increases the growth of PAVM. While the rise in blood volume and cardiac output could increase the pulmonary blood flow and could result in dilatation and rupture of the thin-walled vessels. This is the reason behind increased hemorrhage during the 2nd and 3rd trimesters. The management was the surgical resection of the lung lobe with the malformation. Upon acceptance of endovascular technique, TCE (transcatheter embolization) becomes the mainstay treatment for pulmonary arteriovenous malformation. This case report describes a pregnant woman diagnosed with PAVM that caused a massive hemothorax. Patient was taken up for emergency lower segment cesarean section following which she required excision of PAVM. A multidisciplinary approach was needed in the successful management of this patient.*

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Date of Submission: 06-10-2021 Date of Acceptance: 21-10-2021

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## I. Introduction:

Pulmonary arteriovenous malformation is an abnormal communication between arteries and veins of the pulmonary vasculature without interposition of capillary bed. Its incidence is rare. It can be asymptomatic in most cases but when clinical manifestations occur, it is due to profound cardiovascular compromise and adverse neurological sequelae, as a result of right to left shunting of deoxygenated blood.[1] Patients can present with dyspnoea, hypoxia and pulmonary hypertension. Pregnancy and its physiological demands can unmask and exacerbate pulmonary arteriovenous malformations due to increase in cardiac work, blood volume and the effect of progesterone on blood vessels.[2] This case report describes a pregnant woman diagnosed with PAVM that caused a massive hemothorax and required excision of PAVM. A multidisciplinary approach to management is highly essential considering the challenges involved in deciding the appropriate therapeutic management in pregnancy which has to be weighed against potential maternal and fetal risks.

## II. Case Report:

A 27 year old, G3P1L1A1, 34 weeks 3days, previous normal vaginal delivery came with complaints of severe dyspnoea and chest pain. She had a history of fever with cough and was tested positive for Scrub typhus at 33weeks of gestation and was on conservative management with Azithromycin. Growth scan was satisfactory. Her previous pregnancy was uneventful.

Ultrasound thorax done showed minimal left sided pleural effusion. Diagnostic tapping was done and 600ml of exudative hemorrhagic fluid drained. CT Pulmonary Angiogram showed solitary intra-luminary simple arteriovenous malformation in lingular segment of left upper lobe with feeding artery and draining vein, massive hemothorax causing massive collapse of the entire left lung sparing the anterior segment of left upper lobe. No evidence of aneurysmal dilatation/ contrast extravasation from the AVM/ primary or secondary signs of acute or chronic pulmonary thromboembolism. 2D ECHO done showed normal left ventricular systolic function with right sided pleural effusion.

Two doses of steroids were covered. She was then taken up for emergency lower segment cesarean section in view of maternal pulmonary compromise. The cesarean section was performed under general anaesthesia. Intraoperative period was uneventful. The Fetal outcome showed an apgar index of 8 and 9 at 1 and 5 minutes respectively. Baby weighed 2.21kg at birth and was shifted to NICU for observation. Macroscopic examination of placenta showed umbilical cord with 2 vessels and microscopic examination showed placental parenchyma with no pathological changes.

Postoperatively patient was shifted to ICU due to the underlying lung pathology. Patient was then taken up for Excision of PAVM. Through Left posterolateral thoracotomy and chest X Ray, via 5th intercostal space - clots were evacuated. The intrapulmonary lesion (2x1.9x2.5cm) in inferior lingular segment of left upper lobe was dissected and clipping and ligation of feeder vessel was done. Postoperatively patient was stable and improved well. Histopathology report showed features consistent with AV malformation.

Abdominal dressing was removed and wound found to be healthy. Baby was by mother's side and feeding well. Thoracotomy site dressing was removed and wound found to be healthy. Abdominal site sutures were removed on postoperative day 7 and patient was asked to review after 1 week.

## III. Review Of Literature:

PAVMs with pregnancy is known to cause higher mortality. Physiological changes that occur during pregnancy lead to rise in blood volume and cardiac output, predominantly in the 2nd and 3rd trimester, which increases the pressure in the PAVM. In addition, increased progesterone levels increase venous distensibility. This cascade results in rupture. All these factors lead to rupture of PAVMs during pregnancy. Literature have shown that PAVM-related morbidity during pregnancy is mostly because of hemothorax.[3]

A study done by Shovlin CL et al [4] showed that 5 (1%) among 484 pregnancies died. Accordingly, the British Thoracic Society guidelines stated that only 1% have a chance of maternal death. Nevertheless, a few literatures have suggested that this might be an underestimation. A review conducted in 1995 among 161 pregnancies in HHT with and without PAVMs.[5] Among those without PAVM, 1 patient had ischemic stroke of unknown etiology, otherwise there was no mortality. Among those with PAVM, 23% had nonfatal complications. The non-fatal complications include rise in pulmonary shunt and ischaemic stroke. 2 participants had lethal pulmonary hemorrhages. 8.7% of the study participants with PAVM died during the study period. Similarly, de Gussem EM et al[6] conducted a study in which 38 mothers with known history of PAVM was included. 8 participants who had past history of PAVMs treatment did not have complications. Among those without treatment, 11 had complication like hemoptysis, Transient ischemic attack and post-partum hemothorax and post-partum MI.

Likewise, the British Thoracic Society defines pregnancy as a relative contraindication to elective embolization because of radiation exposure and hazard of preterm labor[7]. Nevertheless, literature have stated that complications due to PAVMs embolization is rare. Pollak JS et al[8] examined 205 non pregnant participants who underwent PAVM embolization and found <1% complication rate. Gershon et al[9] did a case series examining the effect of embolization among 13 PAVMs patients in 7 pregnant mothers. The study was conducted among the gestational age of 16 to 36 weeks and found no complications. Studies on association between PAVM embolization and preterm labor is scarce.

Another main concern is the radiation exposure, which may have detrimental effect in fetus. However, it has been reported that such detrimental effect occurs only with higher radiation dose.[10] It is also to be noted that the risks of radiation exposure reduce with increasing gestational age. Dauer LT et al[10] has stated that during 8–15 weeks gestation, at 100mG there occurs decreased intelligence quotient (IQ) and at radiation above 250 mGy, growth retardation is seen. Still, from 16 weeks to term, at 100 mGy decreased IQ is characteristically observed but growth retardation is observed only at >1500 mGy.[10] From PAVM embolization, the fetal radiation dose is predicted to be < 1–2 mGy, which is far below the adverse thresholds. It has also shown that PAVM embolization has projected 1 in 5000 risks of cancer, and no detrimental effects, when compared to a larger than 1 in 50 risks of fetal or maternal death from untreated PAVM. Thus, benefits outweigh the risks.

Next fear is the safety of contrast agents during pregnancy. Generally, iodinated contrast media are considered to be safe for pregnant and lactating mothers.[11] Even though there is a transplacental transfer of iodinated contrast, evidence of teratogenic effects in humans is less.[11] Puac P et al[11] has stated that usage of nonionic iodinated contrast agents are favored in pregnant women, as they have minimal effect on the neonatal thyroid function.

Thus, evidence showed that the risk of untreated PAVM in pregnancy is high, when compared to the risk from PAVM treatment. The patients with PAVMs should be treated prior to pregnancy whensoever possible. Suspected Pregnant patients should be screened for PAVM with TTCE(Transthoracic contrast echocardiogram) or low-dose non contrast chest CT. Upon diagnosis, multidisciplinary approach should be recommended for treatment.

## IV. Discussion

PAVM in pregnancy can cause life-threatening complications like haemothorax and heart failure. Recent reviews of case reports provide mortality rates of 0–15% for untreated PAVM.[12] It can occur primarily or in association with HHT (Hereditary hemorrhagic telangiectasia), an autosomal dominant genetic disorder characterised by the presence of multiple visceral arteriovenous malformations. The majority of morbidity and mortality from PAVM occurs during second and third trimesters of pregnancy, furthermore in the intrapartum

period. Previously stable patients can dramatically deteriorate in this relatively short space of time.[12]

In PAVM, Chest X ray appearances are largely non-specific and as such if there is strong clinical suspicion of malformation, more definitive imaging is required to make the diagnosis. There is a place for CT and Magnetic Resonance Imaging (MRI), useful in visualising the location and size of such lesions, but Pulmonary Angiography remains the gold standard and in stable women.

Treatment is by transcatheter embolisation (TCE). Surgical resection is still recommended in the setting of haemodynamic instability or the severely compromised. TCE in the non-pregnant patient is well described in the literature, although its use in pregnancy is still surrounded by controversy regarding exposure of the fetus to radiation.[13]

In the event of worsening or recurrence of PAVM after treatment, management needs to be focused on maintaining cardiac and respiratory function, particularly as labour is a time of exertion and possible worsening of hypoxia. The haemodynamic effect on the vasculature is also more pronounced during this period. An intrapartum anaesthetist plan is to be sought to assess the safety of general anaesthesia versus regional anaesthesia should this become necessary. The latter because there may be coexisting epidural AVM in women with PAVM and so to determine this, CT/MRI of the lumbar spine is recommended in the early third trimester. This, and any other plans, needs explicit documentation in the notes so that all attendants at the birth, midwives and doctors alike, are aware of what is safe and more importantly what is contraindicated.

PAVM is not a contraindication to vaginal delivery; however, the consideration in the event of worsening or recurrence is to minimise haemodynamic stresses and systemic hypertension. The advice is to shorten the second stage where possible. Saturations need close monitoring during the intrapartum period.[13]
Long-term follow-up has identified subsequent complications such as persistence of PAVM, recanalisation, growth of accessory vessels and ischaemic strokes. [14]

## V. Conclusion:

A multidisciplinary approach is needed in the care of women with PAVM in pregnancy. The considerations include the risks of further deterioration with advancing gestation with potential lethal consequences in the event of PAVM rupture. Furthermore, the choice of therapeutic intervention is crucial as the option of surgical resection under general anaesthesia is not without attendant maternal morbidity risks in a haemodynamically stable patient.

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