

# Lung Arteriovenous Malformation Surgery Better Cost Effective Option

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#### Abstract

Pulmonary arteriovenous malformations (PAVM) are direct communications between the smaller pulmonary arteries and veins. These are lesions in the vasculature that allow blood to bypass the capillary system, flowing from arteries directly into veins. These lesions are congenital in nature and are caused by failure of differentiation of the embryonic vascular plexus. Patients may be asymptomatic in the milder forms, or have respiratory distress and haemoptysis in the severe types. The right to left shunt may be complicated by polycythaemia, cyanosis and cerebral abscesses. Treatment options for localised form are vascular intervention like embolization using coil but are costly. Peripherally located lung arteriovenous malformation better managed surgically either segmentectomy or lobectomy. Here we discussed a case of peripherally located right lung arteriovenous malformation. In addition to high cost of vascular intervention it was also difficult in this case. So, managed it surgically by ligation of feeding vessel with excision of arteriovenous malformation successfully and cost effectively by right thoracotomy.

Keywords: Arteriovenous Malformation; Pulmonary Function Test

## Abbreviations

AVM: Arteriovenous Malformation; PFT: Pulmonary Function Test

## Introduction

Pulmonary arteriovenous malformations (PAVM) are direct communications between the smaller pulmonary arteries and veins. These are lesions in the vasculature that allow blood to bypass the capillary system, flowing from arteries directly into veins. These lesions are congenital in nature and are caused by failure of differentiation of the embryonic vascular plexus. These vessels expose the low resistance venous system to systemic pressures leading to abnormal vascular formations. Such lesions may be solitary and discrete or generally diffuse [1].

It has been reported that anywhere from 50% to 90% of pulmonary AVMs are associated with hereditary hemorrhagic telangiectasia(HHT), an unusual autosomal dominant disorder also referred to as Rendu-Osler-Weber syndrome. This condition affects the arteries of the nose, skin, brain, lung, and gastrointestinal tract. Pulmonary AVM screening is recommended for families with HHT, and screening includes chest radiography and arterial blood gas measurements taken in the upright and supine positions.

Patients may be asymptomatic in the milder forms, or have respiratory distress and haemoptysis in the severe types. The right to left shunt may be complicated by polycythaemia, cyanosis and cerebral abscesses [2].

The gold standard in diagnosis is pulmonary angiogram showing abnormal peripheral vascular formations that may be either localized or diffuse. Contrast echocardiography has been proven useful and accurate in screening for PAVM. Due to the rapid transit of blood flow through the pulmonary veins, the left atrium is seen to be filled early with contrast (within 2 - 5 seconds) following contrast passage through the right atrium. Multi-slice computed tomography is also useful [3].

Pulmonary AVM treatment is recommended for symptomatic patients or those AVMs with a feeding artery diameter  $\geq$ 3 mm. Transcatheter embolotherapy with stainless steel coils or detachable balloons is most commonly performed. Following embolotherapy, a considerable decrease in pulmonary AVM size is expected; persistent size may indicate persistent perfusion. But the cost of such intervention is very high so sometime localised AVM can be managed by open surgical excision. Here we have discussed such similar case of localised pulmonary AVM. Patient was not affording and AVM is difficult to be embolise so removed by open surgery [4].

#### **Case Report**

23-Year-old female was admitted with exertional breathlessness and palpitations since 3 months. Symptoms gradual in onset, gradually progressive in nature to grade three breathlessness on admission. No h/o chest pain, orthopnoea or PND. No h/o comorbid illnesses or previous surgeries.

On examination, she had central cyanosis with no heart murmurs and normal vesicular breath sounds in lung fields. Spo2 of 84% in room air and 92% with oxygen supplementation.

Hemoglobin of 18.9gm/dl, hematocrit- 54, arterial blood PAO<sub>2</sub>- 78

Chest x-ray showed right middle zone haziness. PFT: normal spirometer values with impaired diffusion (Figure 1). HRCT Chest shown pulmonary arteriovenous malformation (3 x 3.6 x 4.4 cms) in subpleural parenchyma of right lower lobe apical segment (Figure 2). Dilated afferent vessel from right pulmonary artery and efferent vessel draining to left atrium. Pulmonary angiography shown right middle zone pulmonary AVF (Figure 3) supplied by branch of right pulmonary artery and draining into right pulmonary vein. Diameter of the feeding artery is 1.2cms (Figure 4).



Figure 1: PFT [ Pulmonary function test] Normal spirometer values with impaired diffusion.

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Figure 2: HRCT Chest shows right lower lobe apical segment arteriovenous malformation of size 3cm by 3.6 cm by 4.4 cm.



Figure 3: Right lower lobe apical segment arteriovenous malformation 3 cm by 3.6 cm by 4.4cm.



Figure 4: Feeding artery of AVM is arising from right pulmonary artery.

2d echocardiogram was normal with LVEF 60% and no pulmonary hypertension.

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Treatment options for localised form are vascular intervention like embolization using coil but are costly. Peripherally located lung arteriovenous malformation better managed surgically either segmentectomy or lobectomy. Decision taken for open surgical excision of AVM. Patient underwent thoracotomy and excision of right pulmonary AV malformation under general anaesthesia. Intra operative findings are subpleural AV malformation of approximate size 3x 3.6 x 4.4 cms in apical segment of right lower lobe. Dilated azygous vein draining into SVC. Rest of the lung parenchyma appeared normal. Ligation of the feeding vessels was done and the arteriovenous malformation was excised in total (Figure 5) resulting in immediate improvement in spo2 to 100%. Single right pleural apical ICD put and thoracotomy closure done. Patient extubated on operation table after skin closure. Postoperative period uneventful with patient maintaining sp02 98 to 100% in room air.



Figure 5: Excised and cut open pulmonary arteriovenous malformation.

### Discussion

Pulmonary arteriovenous malformation is abnormal communications between the pulmonary arterial and venous systems that bypass the capillary bed. Their frequency varies, occurring in roughly 10 to 20 persons per 100,000. Pulmonary AVMs may be classified as simple with a single feeding and draining vessel (80% of cases), or complex, with 2 or more feeding or draining vessels (20% of cases). Up to 65% of pulmonary AVMs are found in the lower lobes of the lung [5].

Pulmonary AVMs frequently go unrecognized until the late teens, but they may remain asymptomatic throughout life. Pulmonary symptoms include dyspnea, fatigue, cyanosis all due to right-to-left shunting of blood through the pulmonary AVM. The most serious complications of pulmonary AVMs are potentially fatal hemoptysis or hemothorax (in up to 10% of patients). Neurologic sequelae may develop as a result of the unfiltered passage of paradoxical emboli through the pulmonary system to the brain. Brain abscess may also result following dental procedures, for instance. On chest radiography, pulmonary AVMs may be seen as rounded, circumscribed pulmonary nodules. Feeding vessels may or may not be visualized. On chest CT, a homogeneous, circumscribed, noncalcified nodule may be seen [6].

Studies have shown that CT is the best non-invasive modality for evaluation of pulmonary AVMs. However, pulmonary angiography is necessary before embolization.

Treatment of localized PAVMs includes surgical lobectomy or transcatheter embolization using coils. There is no definitive treatment for the diffuse type

#### Conclusion

Management of pulmonary AVM may vary depending upon type like for diffuse no treatment followed by local type for which two options one is transcatheter embolization which is costly and not possible in all cases and another one is open surgical excision. Our ex-

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perience with this case shows that open surgical excision was better option in two scenero one in which pulmonary AVM is difficult to be managed by intervention and another one is patient is not affording for intervention treatment.

## Consent

Informed consent has been obtained.

# Funding

No funding was required for this study.

## **Conflict of Interest**

No potential conflict of interest exist.

## **Ethical Approval**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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