

Surgical Management of Medium to Large Size Pulmonary AV Malformations- A Case Series

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Abstract

The communication between pulmonary artery and pulmonary vein or between a bronchial artery and the pulmonary vein is called as pulmonary arterio-venous malformation. Dyspnea on exertion, fatigue, nasal bleeding, bluish discoloration of lips and nail beds and hemoptysis are common presenting complaints. In childhood only 10% of cases are identified. Cerebral vascular accidents, brain abscess, hemothorax, life threatening hemoptysis are the complications of large pulmonary arteriovenous malformation. We present you a series of three cases that were diagnosed to have pulmonary AV malformations and presented to us with various clinical pictures. All three patients were surgically managed and discharged uneventfully.

Keywords: Arteriovenous malformation; Hereditary hemorrhagic telangiectasia

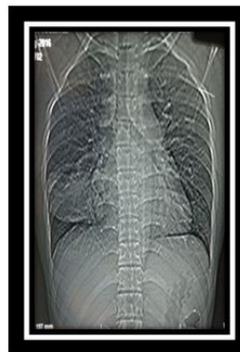
Introduction

The abnormal communications between the pulmonary arteries and the pulmonary veins are called pulmonary arteriovenous malformations (PAVMs) or fistulae first described in 1897. Pulmonary arteriovenous malformations commonly have autosomal dominant trait with associated hereditary hemorrhagic telangiectasia (HHT). Pulmonary arteriovenous malformations also found in patients with liver disease like liver cirrhosis and absence of hepatic factors may be the cause. Cyanotic congenital heart disease patients also develop pulmonary arteriovenous malformations. Chronic infections like schistosomiasis, actinomycosis, tuberculosis and metastatic thyroid cancer can also lead to pulmonary arteriovenous malformations. It is the communication between pulmonary artery and pulmonary vein or between a bronchial artery and the pulmonary vein.

Case 1

A 21 year old boy presented to us with complaints of breathlessness on exertion since 3 months. He was apparently asymptomatic before three months when he first became aware of his symptoms after a short run. Patient had no h/o cough, chest pain, palpitations, fever and loss of weight or appetite. Patient had no history of similar complaints anytime in the past. He was a non-alcoholic, non-smoker and no history of tobacco chewing or any other addictions in the past. Patient is not a diabetic or hypertensive and no history of asthma or tuberculosis in the past. Patient was evaluated for the same and his blood investigations revealed hemoglobin of 19.2g% and hematocrit was 64.5, platelet count was 1.7 lakh and his arterial blood gas showed a PO₂ of 87%. His chest X-ray revealed a nodular shadow in the right lower lobe (Figure 1). Patient was evaluated for the same using HRCT thorax which revealed a hyper dense vascular lesion in the right lower lobe suggestive of pulmonary AVM (Figure 2).

Case 1

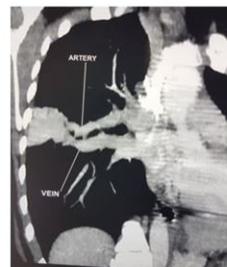


Case 2



Figure 1: Chest X-rays showing haziness in right middle and lower zone of case 1 and 2.

Case 1



Case 2

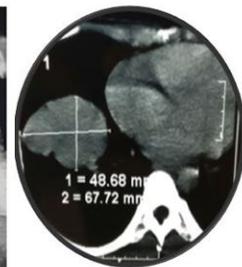
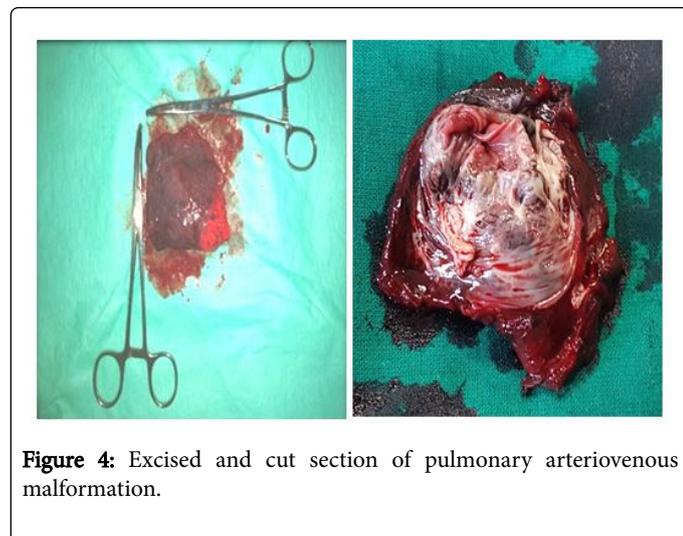


Figure 2: HRCT chest arteriovenous malformation of right lower lobe apical segment.

On pulmonary angiogram the lower lobe apical segment showed pulmonary AVF fed by the right inferior pulmonary artery and drained by right inferior pulmonary vein. The bronchial segmental collaterals were well developed. Patient underwent right posterolateral thoracotomy and underwent ligation of the base of the AVM and lobectomy of the right lower lobe (Figures 3 and 4). Patient was discharged on day 9 with uneventful postoperative period.



On histopathological report hematoxylin-eosin stained sections revealed small and medium sized thickened arteries anastomosing with dilated veins. Based on these findings, a diagnosis of pulmonary arteriovenous fistula was confirmed.

Case 2

A 26 year old female patient presented to us with complaints of severe breathlessness on exertion since 1 year and occasional episodes of hemoptysis once in 3-4 months and persistent cough since 6 months. There was no significant past or family history. She was cyanotic and had significant breathlessness at the time of presentation. She was extensively evaluated for the same. On examination patient was thin built there is central cyanosis, no pallor, clubbing, icterus or pedal edema. Her respiratory system examination revealed significant

congestion on the right middle and basal regions. Other systems examinations were however normal. Her baseline investigations revealed hemoglobin of 18 g% and hematocrit of 54. Her arterial blood gas revealed significant decrease in PO₂ upto 81%. Right lower lobe nodular opacity was seen on chest X-ray (Figure 1), however she was further evaluated using a contrast enhanced CT scan which revealed a lobular high density lesion in the right lower lobe measuring about 1-1.5 cm approximately (Figure 2). She underwent pulmonary angiography which revealed a large pulmonary AV malformation with feeding artery and draining vein and aneurismal dilatation of the feeding vessel. Patient underwent attempted embolization of the feeding vessel but it was not complete successful, hence she was planned for segmentectomy with resection of the AVM. Patient underwent thoracotomy and ligation of the feeding vessel and segmentectomy of apical segment of the lower lobe (Figures 3 and 4). Patient post-operative period was uneventful and she was discharged home on the 7th Post-operative day.

Case 3

A 42 year old male patient presented to us with complaints of sudden onset breathlessness and severe right sided chest pain. Patient had no h/o cough, hemoptysis, fever, bluish discoloration of the skin. Patient was apparently asymptomatic before this episode of sudden onset breathlessness which he started developing after his previous night meal. Patient has no history of any such similar complaints anytime in the past. He was not a diabetic or hypertensive and no history of bronchial asthma or no history of tuberculosis in the past. On examination patient was well built and nourished, dyspnoeic at rest and his respiratory system examination revealed significantly diminished breath sounds on the right side. He was evaluated for the same using a chest radiograph which showed a massive effusion on the right side. Patient immediately underwent a HRCT thorax which showed a massive hemothorax with possibility of a AVM on the right middle lobe. Patient's blood investigations revealed hemoglobin of 8 grams and total WBC count of 11200 and a platelet count of 1.4 lakhs. An intercostal drainage tube was inserted on the right side and drained 600 ml frank blood. Patient was stabilized after the same and later underwent a pulmonary angiography which revealed a medium size pulmonary AVM in the right middle lobe measuring about 5 cm. The feeding artery was right inferior pulmonary artery and vein. Thoracotomy and excision of the pulmonary AVM after ligation of the feeding artery and vein was done and patient was discharged without any complication on 10th day.

Discussion

Pulmonary arteriovenous malformations are of two types simple and complex based on morphology. Simple having single feeding segmental artery leading to single draining pulmonary vein whereas complex having two or more feeding arteries and draining veins. Around 79% of pulmonary arteriovenous malformations are of the simple type commonly affecting lower lobes and 21% of pulmonary are complex type occurring in the lingula and right middle lobe [1].

T Churton in 1897 first described pulmonary arteriovenous malformation (PAVM) with female to male ratios ranging from 1.5 to 1.8:1 [2]. The estimated incidence is around 2-3 per 100,000. Anatomically, they are classified into focal, complex and diffuse. Focal type is commonest has a single feeding segmental artery whereas complex types have multiple segmental feeding arteries (20%). The diffuse type is rare (5%) have combination of simple and complex

AVMs within a diffuse lesion. Based on etiology, PAVM is of four types of which most common is HHT (85%) [3]. Type 1-HHT is an autosomal dominant disorder in which vascular malformations is commonly seen in the skin, nasopharynx, GI tract, lungs, and brain. HHT is a triad of cutaneous telangiectasia, recurrent epistaxis and a family history of this disorder. Type 2-Idiopathic congenital pulmonary arteriovenous malformations (15%) are likely to be single [4]. They are less likely to become enlarged, and successfully treated with embolotherapy. Type 3- Pulmonary arteriovenous malformations may also be acquired rarely secondary to chronic infections (<1%) such as schistosomiasis, actinomycosis, tuberculosis; liver cirrhosis and metastatic thyroid cancer. Type 4-PAVM may be iatrogenic (<1%) due to cardiovascular intervention like Glenn or modified Fontan procedures for congenital cyanotic heart disease. *Endoglin* and *AVRL-1* genes are implicated in angiogenesis in HHT responsible for pulmonary arteriovenous malformation. In a PAVM, blood bypasses the normal oxygen-exchanging pulmonary capillary bed returning desaturated blood to the pulmonary veins [5]. When the shunting becomes significant cyanosis result which leads to a compensatory rise in hematocrit and hemoglobin concentration. PAVM are more common in lower lobes which leads to orthodeoxia (accentuation of arterial hypoxemia in erect position, improved by assumption of recumbent position) and platypnea in advanced cases. Bleeding from nose and GI telangiectasias may reduce the hemoglobin in HHT leading to anemia and stroke from paradoxical embolization. Mortality is higher in bilateral, diffuse pulmonary arteriovenous malformations (upto 55%) than the focal variety due to availability of effective therapeutic interventions [6]. The age of presentation of PAVM depends on size of lesion, diffuse or isolated, unilateral or bilateral. The patient presents earlier if size of PAVM is large, diffused or bilaterally located. Most common presentations were dyspnea with or without exertion and cyanosis. Symptoms in early life may vary from being totally absent to severe with cyanosis, congestive heart failure and even fulminant respiratory failure [7]. The most common complaint in symptomatic patient is epistaxis mostly associated with HHT which was absent in this patient. The second most common complaint is dyspnea. Superficial telangiectases attributable to HHT are the most common and frequently the only physical finding in patients with PAVM which was absent in this patient. Murmurs or bruits over the site of the PAVM are heard in about 50% of patients. The murmurs are most audible during inspiration, and usually increase on assuming positions which put the PAVM in dependent position. Digital clubbing and cyanosis were seen in only 40% of patients. In one study the classic triad of PAVM dyspnea, cyanosis and clubbing was present in only 10% of patients with PAVM [8]. Hemoptysis may occur and less commonly seen symptoms are chest pain, cough, headache, tinnitus, dizziness, dysarthria, syncope, vertigo and diplopia which are all due to hypoxemia, polycythemia or paradoxical embolization through malformations (PAVM).

Conclusion

Pulmonary AVM can be managed by transcatheter embolization but it is costly and not possible in all cases. Another option to manage is open surgical excision. Our experience with these cases shows that if patient is not affording for transcatheter embolization or if pulmonary AVM not manageable by intervention then open surgical excision of AVM was the best option. Surgery for pulmonary AVM can be done safely and without any complication with adequate knowledge of lung segmental anatomy.

Consent

Informed consent has been obtained

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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