

A Rare Case Report of Giant Isolated Pulmonary Arteriovenous Malformation

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ABSTRACT

Introduction: Pulmonary Arteriovenous Malformations are considered as very rare vascular anomaly of lung with abnormal communication between pulmonary artery and pulmonary vein leading to Right-to-Left shunt. Patients are usually asymptomatic and may present with orthopnea, haemoptysis.

Case report: We are presenting a case of 30-year old male presented with complaints of dyspnea, dysphagia and retrosternal burning pain since six months. On CECT chest with multiplanar reconstruction a well defined homogenous intensely enhancing mass seen in left lower lobe with a connecting vessel extending from the left pulmonary hilum suggesting pulmonary arteriovenous malformation. Imaging helps to evaluate the location, morphology, feeding and draining vessels of malformation.

Conclusion: Therapeutic options include embolotherapy with coils or amplatzer plugs, surgical ligation, excision and even lobectomy. Cross sectional imaging with multiplanar reconstruction plays an important role in diagnosis and timely intervention of PAVM.

Keywords: Pulmonary Arteriovenous Malformation (PAVM), Hereditary Hemorrhagic Telangiectasia (HHT), Right to Left Shunt (R – L shunt), Contrast Enhanced Computed Tomography (CECT).

CASE REPORT

A 30-year old male presented to the outpatient clinic with complaints of dyspnea, dysphagia and retrosternal burning pain since six months. No history of recent chest trauma or previous thoracic surgeries. No significant past medical illness

or family history. On clinical examination, the patient was afebrile and vitals are stable. No signs of skin involvement. He was referred to the department of Radiodiagnosis for further investigations. Incidentally, frontal chest radiograph showed a well-defined homogenous opacity in the left lower lung with a connecting vessel radiating from hilum (Fig.1).

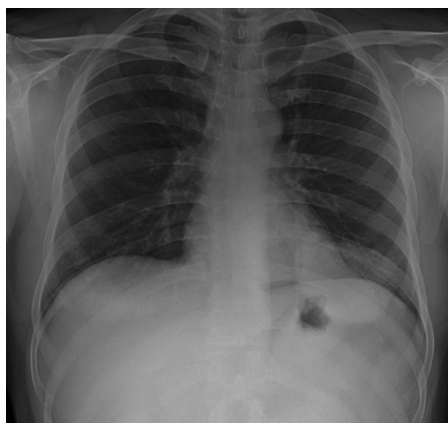


Figure-1: Chest X-ray showed a well-defined lobulated homogenous radio opacity in left lower zone with a connecting vessel radiating from hilum.

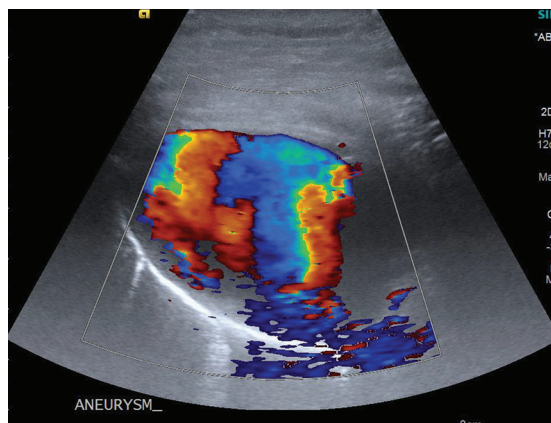


Figure-2: On ultrasound, an anechoic lesion is noted in left lower lung field showing a significant vascularity on color Doppler.

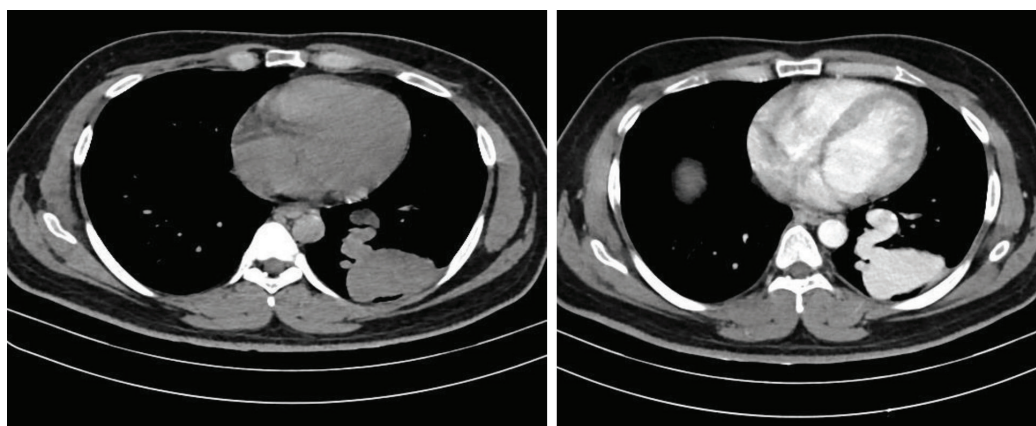


Figure-3: A&B. Axial plain and contrast enhanced CT chest, mediastinal window showing a well-defined intensely enhancing soft tissue density lesion in left lower lobe with a tortuous connecting vessel extending from left pulmonary hilum.

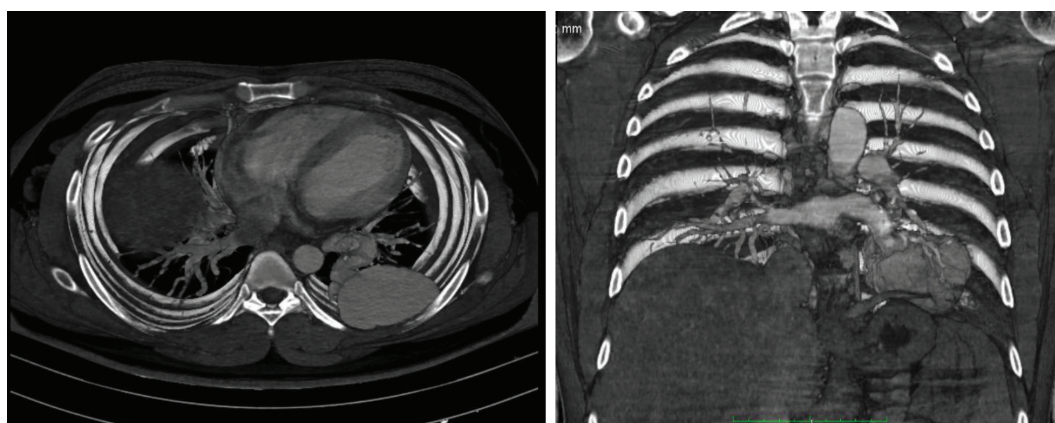


Figure-4: A&B. Axial and coronal MPR images showing pulmonary AVM with aneurysmal component. The feeding artery is seen arising from left pulmonary artery and draining into left inferior pulmonary vein.

On Ultrasonography, a well defined anechoic lesion noted in lower lobe of left lung with intense vascularity showing yin-yang sign suggesting arterial and venous flow on Doppler (Fig.2). Patient subsequently underwent Contrast-enhanced CT chest, axial and coronal images with multiplanar reconstruction showed a well defined homogenous intensely enhancing mass measuring 65 x 51 x 47 mm in posterior-basal segment of left lower lobe with a connecting vessel extending from the left pulmonary hilum. The aneurysmal component showed few peripheral non enhancing areas with wall calcifications, the feeding artery is seen arising from left pulmonary artery and draining into left inferior pulmonary vein (Fig.3,4).

Based on above imaging findings, diagnosis of Giant pulmonary arteriovenous malformation was made.

DISCUSSION

Pulmonary AVMs are considered as rare congenital malformation with abnormal communication between pulmonary artery and pulmonary vein leading to R-L shunt. These are considered as direct high flow, low resistance fistulous connections. The incidence of PAVM is 2–3 per 100 000 population¹. Most commonly associated with hereditary hemorrhagic telangiectasia, an autosomal dominant disease presenting with epistaxis, mucocutaneous and visceral telangiectasia involving lungs, brain and liver with a family

history seen in more than 60%. HHT related PAVMs usually present as multiple, bilateral predominantly involving basal areas. Septic thrombus or Paradoxical embolism can occur which can lead to cerebral embolic attacks².

The presence of HHT with a co-existing PAVM may have worse symptoms associated with multiple pulmonary arteriovenous malformations, rapid disease progression and higher complication rate and mortality⁴. Hence, clinician should be aware of these malformations.

PAVMs can show gradual increase in size when associated with conditions like puberty, pregnancy and pulmonary hypertension³. Pregnancy has been associated with an increased rate of PAVM growth and is associated with high risk of complications due to steroid hormone synthesis causing increased cardiac output are tend to rupture and leading to severe hemoptysis and death¹.

PAVMs are of two types

Congenital/ primary cause that occurs due to abnormal development of pulmonary arteries and veins.

Secondary causes include chest trauma, previous thoracic surgery like cardiac shunt procedures, long standing hepatic cirrhosis, metastatic carcinoma, mitral stenosis, hepatorenal syndrome, infections like Actinomycosis, Schistosomiasis, and Systemic amyloidosis⁵.

They may be either simple or complex type. Simple PAVM occurs in about 80%, comprises of a feeding artery arising

from a single segmental artery and draining vein with a large aneurysmal component. Complex PAVM accounts for 20% which is supplied by two or more feeders arising from different segmental arteries and draining veins connected to a septated aneurysmal sac⁸.

Embryologically PAVMs are classified into five groups, with Group I comprises multiple small AVMs without aneurysm, group II contain large AVMs with aneurysm, group III contain large or multiple small AVMs with anomalous venous drainage, group IV contain large venous aneurysm with systemic artery communication or without fistula and Group V contain anomalous venous drainage with fistulas⁶.

PAVMs are usually asymptomatic, most commonly present with haemoptysis. Symptoms are due to pressure related effects that occur during intrapulmonary shunt which include dyspnea, orthopnea, palpitations with cyanosis.

Complications include intrapulmonary shunt, intrabronchial rupture, hemothorax, hypoxemia, polycythemia, pulmonary hypertension, paradoxical embolism, stroke and brain abscess. Frontal and lateral chest radiographs are considered as screening tool in evaluation of PAVM and shows a homogenous radio-opacity with a connecting vessel radiating from hilum. Echocardiography shows R-L shunt. Contrast echocardiography helps in anatomical localization of PAVM and in follow up after embolotherapy.

Cross sectional imaging demonstrate abnormal communication of pulmonary artery and vein presenting as feeding artery and a draining vein. CECT chest shows intense enhancement with focal area of ground glass opacity. Pulmonary angiography is used as confirmatory investigation, useful in identifying the pulmonary vasculature and helps in preoperative assessment. CT pulmonary angiography with 3D reconstruction helps for optimal detection and treatment planning by evaluating the location, morphology, feeding and draining vessels.

MR angiography is considered as a noninvasive diagnostic modality and can demonstrate patency of PAVM, however it is not routinely used due to high cost and limited availability. Feeding artery can be visualized usually arises from pulmonary artery, rarely can arise from intercostal, internal mammary bronchial, and phrenic arteries. Draining vein is most commonly through pulmonary vein, rarely IVC and left atrium¹.

3D imaging is used to evaluate the location, morphology, feeding and draining vessels. Radionuclide scan is helpful in diagnosing and quantification of PAVM.

Treatment modalities include embolotherapy and surgery. Embolotherapy is done by balloons, metallic coils and amplatzer plugs. Coils are considered as the mainstay for endovascular occlusion. The amplatzer vascular plugs are foldable nitinol baskets forming shape of the vessel and is detached from the delivery device by a screw mechanism, allowing for precise localization as well as for repositioning of the plug. These are MRI compatible. Packing of the aneurysmal sac with coils is not typically necessary. The sac will gradually regress in size and involute if the feeding vessels are appropriately occluded. Some fistulas demonstrate dilated feeding arteries that cannot be coiled safely or treated with amplatzer¹⁰.

Embolotherapy is most suitable for elderly patients with

poor surgical compliance and in multiple lesions with feeding artery size more than 3mm and allows accurate location of PAVM with minimal recanalization rate and morbidity³.

Surgery is indicated in patients with serious bleeding complications and failed embolotherapy. Surgical techniques include excision, segmental resection, lobectomy, ligation and pneumonectomy. Recently, video assisted thoracoscopy (VATS) is employed in small PAVM resection¹.

Differential diagnosis of PAVM include Pulmonary artery aneurysm, pulmonary varix, bronchocele and tumors⁹.

CONCLUSION

Cross sectional imaging with multiplanar reconstruction plays an important role in diagnosis and timely intervention of PAVM. The presence of HHT with a co-existing PAVM may have worse symptoms associated with multiple pulmonary arteriovenous malformations, rapid disease progression, and higher complication rate and mortality. Hence, clinician should be aware of these conditions and imaging helps in prognosis of PAVM and long-term follow-up after treatment.

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