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PULMONARY ARTERIO-VENOUS MALFORMATION ASSOCIATED WITH PULMONARY ALVEOLAR HAEMORRHAGE: A RARE CASE REPORT

Pulmonary Medicine

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ABSTRACT

PAVM are rare pulmonary vascular anomalies. Patient may present with hemoptysis, shortness of breath and hypoxemia. Direct communication between the branches of pulmonary artery and pulmonary veins,without an intervening pulmonary bed are probably the most common anomalies of pulmonary vascular tree.Although most patients are asymptomatic because of paradoxical emboli, various central venous system complications have been described including stroke and brain abcess.

There is a strong association between PAVM and hereditary haemorrhagic telangiectasia. Chest radiography and contrast enhanced computed tomography are essential initial diagnostic tools but pulmonary angiography is the gold standard. Contrast echocardiography is useful for diagnosis and monitoring after treatment. Therapeutic options include angiographic embolisation with metal coil or baloon occlusion and surgical excision.

More than 80% of PAVMs are congenital and of those 47-80% are associated with osler weber render disease or HHT. Secondary or acquired PAVM are very rare.

We present a case of PAVM,which was picked on a chest x-ray done in a patient with hemoptysis.

KEYWORDS

INTRODUCTION:

PAVMs refer to an abnormal fistulous connection between arterial and venous branches, without a Customary intervening capillary network that is vital for gas exchange.

CASE-REPORT:

A 26 year old young male patient, had a traumatic history of fall down from bike 2 months back, presented to EMENGENCY DEPARTMENT with a spurious hemoptysis of about 500 ml.

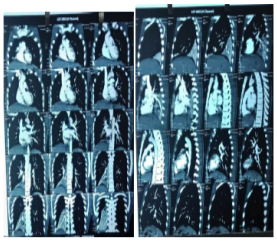
He complained of massive hemoptysis about 600-700 ml daily since 1 month which was occasionally controlled by hemocoagulants but re-started on stopping of medications.CXR was done and all other causes of hemoptysis were ruled out.

CECT Thorax was done, which suggested vascular tangle noted in sub-mucosal region in bronchus intermedius supplied by Right Bronchial Artery arising from anterior aspect of Descending Thoracic Aorta, possibility of vascular malformation with subsegmental collapse of medial segment of right middle lobe and complete collapse of right lower lobe.

Angiography was advised to co-relate.

CECT Pulmonary Angiography suggestive of- 95x10x5 mm (APxTRxCC) and 5x4x6 mm (APxTRxCC) sized tangle of dilated tortuous vessel noted in subpleural location of lateral basal segment of right lower lobe which is supplied by segmental branch of Right Pulmonary Artery and drained by segmental tributary of right pulmonary vein,s/o AV malformation.similar sized characteristic multiple lesions in other regions of right lower lobe.

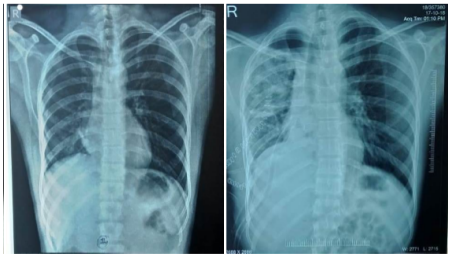
Associated ground glass opacity with septal thickening in right middle and lower lobe with crazy pavement pattern s/o pulmonary hemorrhage.



2D ECHO was normal. Patient was first tried with embolisation but on exploration it was found that PAVM was associated with intra lobar and parenchymal involvement, so middle and lower lobe lobectomy was done.Post operative lung expanded completely.Patient was kept in ICU for 2 days under observation and was successfully discharged after 5 days.

At present,he is doing all his routine activities very well and has not yet developed a single episode of hemoptysis post operative period day 28.

He has not developed any post operative complications.



Before **After**

***DISCUSSION*:**

Approximatlly 80% of PAVMs have a single feeding and a single draining vessel.

Upto 65% of PAVMs are located in corner lobes.[7]

Age of onset is usually in the third or fourth decade.[8]

The estimates of frequency with which PAVMs are due to HHT vained substantially from 36-95%.[3,4,5,8,9]

Pulmonary symptoms include dyspnea on exertion,frequency ranging from 27-71%[1,4,7,11]

Hemoptysis ranges in frequency from 4-18%.[1,6,7] which may be massive and life threatening.

Hypoxia,cyanosis and clubbing focuses secondary to right to left shunt,which may resolve after the PAVM is resolved.[12]

Hemothorax may also occur presumably caused by rupture of large sub pleural PAVMs into the pleural space.

37% [7] of patients may develop symptomatic cerebro vascular accidents.(TIA) 3-10% of patients develop brain abcess. [2,10]

Discovery of a nodule on chest x-ray is typically Followed by ct scan and angiography. Evidence of intra pulmonary right to left shunt confirms the diagnosis.

In Patients with diffuse small PAVMs or telengectasia, transesophageal contrast echocardiography may,provide the definitive evidence which delays the appearance of the contrast material in the left side of chest.[14]

Earlier,surgical removal of PAVM was done.Thoracoscopic Resection has more recently been described.

With recent advances,Embolisation of PAVMs has proved to be an excellent alternative.Success rate is greater than 93% [16] and embolisation therapy is now the procedure of choice.

Recently,many interventional radiologists have prepared using vascular plugs for large feeding vessels and reserving coils for small or more tortuous vessels.

Reperforation of the embolised vessel may occur in upto15%[11,17,18]

Some patients may have persistent intra pulmonary shunt and should receive prudential antibiotic prophylaxis.[15]

The most common post embolisation symptom is pleurisy and has been reported at rates ranging from 10% to 31% [7,13,19]

Diffuse PAVMs resulting in hypoxemia,difficult to treat with embolisation therapy have been successfully treated with lung transplantation.[20,21]

CONCLUSION:

In summary, Patients with PAVM can be successfully treated with resolution of essentially all symptoms and substantial reduction in risk of complications. Embolotherapy is the treatment of choice for most patients.Education materials for patients with HHT and the location of specialized centers for management of HHT and PAVM are available from the HHT foundation international at http:// hht.ors

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