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Research Article

TRANSCATHETER TREATMENT OF LARGE COMPLEX PULMONARY ARTERIOVENOUS MALFORMATION

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A B S T R A C T

Pulmonary arteriovenous malformation (PAVM) is a rare clinical condition with abnormal direct communication between pulmonary arteries and pulmonary veins causing intrapulmonary right to left shunt. The incidence of PAVM is 2.5/100000 population. Female to male ratio is 2:1. Clinical manifestations may range from asymptomatic to exertional dyspnoea, cyanosis, clubbing, hemoptysis or cerebral abscess. CT pulmonary angiography is the gold standard for diagnosis of PAVMs. Treatment of choice in majority of cases of PAVMs is transcatheter embolotherapy with balloons, stainless steel coils or vascular plugs.

We present a 26-year old male who developed exertional dyspnea during an acute febrile illness in COVID-19 pandemic. During investigation for COVID-19, PAVM was diagnosed. Infection was not considered as the cause of PAVMs as the fever was of very short duration and PAVMs were of large size with large feeder arteries. We speculated that PAVMs were already present and were asymptomatic until the acute febrile episode precipitated the exertional dyspnea. We, therefore, considered the PAVMs as the cause of exertional dyspnea in the absence of lung parenchymal disease and treated the PAVMs percutaneously with vascular plugs.

Our case is a good example where even large PAVMs may be asymptomatic until some acute illness precipitates symptoms, and also where large PAVMs can be successfully treated percutaneously.

INTRODUCTION

Pulmonary arteriovenous malformation (PAVM) is a rare clinical condition with abnormal direct communication between pulmonary arteries and veins through a thin walled aneurysmal sac. This results in an intrapulmonary right to left shunt. Whereas, single or small PAVM is largely asymptomatic, larger and multiple PAVMs may present with hemoptysis, dyspnea , cyanosis, cerebral infarction or cerebral abscess.[1]

The incidence of PAVM is around 2.5/100000 population. About 80% of these are congenital. Most of the congenital PAVMs are associated with Osler-Weber-Rendu Syndrome or hereditary hemorrhagic telangiectasia (HHT). Majority of PAVMs are present in left lower lobe.[2]

Male to female ratio is 1:2. There is a male predominance in newborns.[3]

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PAVM is classified as simple (80%) with single feeder vessel and complex(20%) with two or more feeder vessel.[4]

Endovascular embolization is the current mode of treatment in the majority of PAVMs.[5]

We present a case with large complex PAVMs which was treated by occluding the large feeding arteries with vascular plugs. This underscores the fact that complex PAVMs can also be treated percutaneously.

Case Presentation

A 26 year old male patient was admitted with fever and dyspnea on exertion for two days. He did not admit of having dyspnea in the past on repeated questioning. His pulse rate was 90/min. Blood pressure was 140/76 mm of Hg. SpO_2 was 81%. Physical examination showed mild cyanosis and grade 1 clubbing. Laboratory investigation revealed Hb 22.4gm/dl, WBC 4000 /mm ^{3}, hematocrit 65% and paO_2 44 mm of Hg. RT-PCR for COVID-19 virus was negative. Chest X-ray (CXR) showed a lobulated oval opacity in right lower lobe. 2D

echocardiography did not show any significant abnormality. In view of cyanosis, clubbing, low SpO_2 (81%), polycythemia , raised hematocrit (65%), CXR findings and normal echocardiography, PAVM was highly suspected. To rule out parenchymal lung disease, HRCT thorax was done. HRCT thorax showed a well defined opacity in lower lobe of right lung. Lung parenchyma was normal. CT pulmonary angiography was done which showed two lobulated well marginated enhancing lesions measuring 27 mm × 26 mm and 23 mm × 22 mm with drainage into pulmonary veins. Pulmonary angiography revealed two large fistulas in right lower lobe of lung with large feeder vessels. Because of larger size of feeder vessels, coil embolization was not done. Instead Amplatzer vascular plugs were used to occlude the feeder vessels. Post procedure pulmonary angiography showed one small patent feeder vessel for which staged coil embolization was planned due to prolonged procedural time and dye overload. Post procedure patient`s oxygen saturation improved to 97.5% and paO_2 improved to 96.6 mm of Hg. Patient was discharged uneventfully.

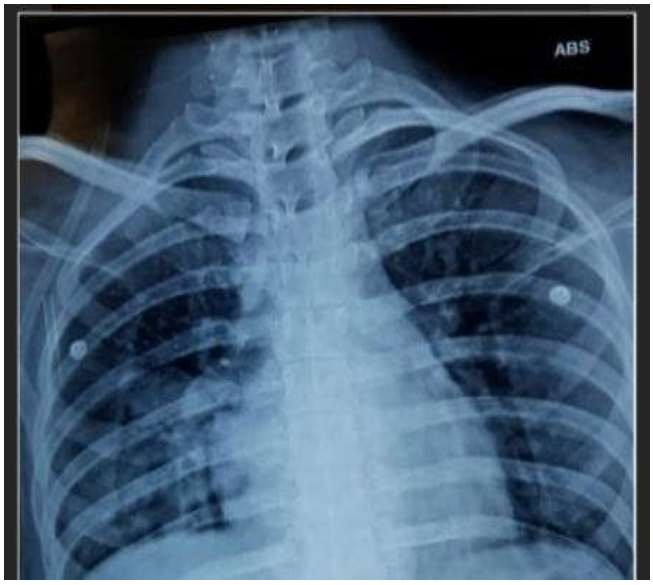

Figure 1 CXR of the patient showing lobulated oval opacity in right lower lobe

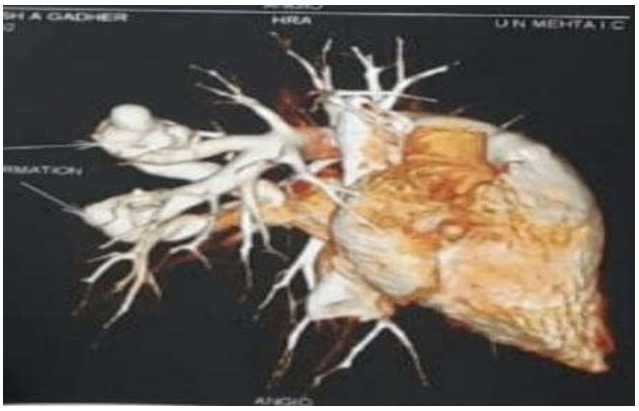

Figure 2 CT pulmonary angiography showing two lobulated well marginated enhancing lesions measuring 27mm x 26mm and 23mm x 22mm with large feeding arteries.

DISCUSSION

PAVM is characterized by direct communication between pulmonary artery and pulmonary vein through a thin walled aneurysmal sac.[1]

Incidence of PAVM is 2.5/100000 population.[2] Male to female ratio is 1:2 with male predominance in newborns. Lower lobes are involved in 53-70% of cases. In 75%, it is unilateral. Thirty six percent have multiple lesions and 50% of these have bilateral localization. More than 80% are present subpleurally.[3] Left lower lobe is the most common location of PAVMs.[4]

PAVM can be classified as either simple or complex. About 80% are simple PAVMs defined by one feeder artery and one draining vein. Rest are complex PAVMs defined by two or more feeder arteries and/or two or more draining veins.[6]

PAVMs may also be classified as primary or congenital and secondary or acquired.[3] Eighty percent are primary. Majority of those are associated with hereditary hemorrhagic telangiectasia.[2] In patients with HHT the PAVMs are multiple (35% to 65%) and usually associated with severe complications and infection.[7] Secondary or acquired PAVMs are caused by infections (tuberculosis, actinomycosis, schistosomiasis), metastatic thyroid carcinoma, thoracic trauma, hepatic cirrhosis, mitral stenosis, modified Fontan operation or iatrogenic.[3]

PAVMs are divided as small (less than 5 cm) or big (more than 5 cm).[3]

The fundamental defect is a right to left shunt. Signs and symptoms of right to left shunt may include exertional dyspnea ,cyanosis, clubbing or hemoptysis.[2]

Majority of the patients develop symptoms between fourth and sixth decade of life.[6] In patients with HHT symptoms like epistaxis occur before the age of 20.[7]

The most common complaint in patients with PAVM is dyspnea on exertion and is present in 50% of the patients.Other symptoms include finger clubbing (20%), cyanosis(18%), hemoptysis (10%), chest pain (6%) and thoracic murmurs (3%).[7] Of the symptomatic patients one-third may develop neurological complications like cerebral infarction or cerebral abscess.[2]

Orthodeoxia is decrease in oxygen saturation upon standing and is one of the hallmarks of PAVM. This is caused by blood pooling at lung bases where most PAVMs are present resulting in more shunting of deoxygenated blood into systemic circulation.[3]

Chest X-ray is usually the initial investigation. Its sensitivity is 28 % and negative predictive value is 85%.[2] The most common CXR finding is that of a round or oval opacity of uniform density, frequently lobulated but sharply defined, mostly in lower lobes.[6]

Transthoracic contrast echocardiography is the best screening test to detect PAVM.[5]

CT angiography usually confirms the diagnosis and is the gold standard for diagnosis of PAVMs.[7] Pulmonary angiography is currently restricted to treatment only.[5]

Treatment of PAVM include surgery or embolotherapy. All symptomatic PAVMs must be treated. Asymptomatic PAVMs larger than 2 cm, or if feeding arteries are larger than 2 mm should be treated.[7] The treatment of choice for PAVMs is transcatheter embolotherapy with balloons, stainless steel coils or vascular plugs.[7]

Our patient developed exertional dyspnea during an acute febrile illness in COVID-19 pandemic. During investigation for COVID-19, PAVM was diagnosed. Infection was not considered as the cause of PAVMs because the fever was of very short duration and PAVMs were of large size with large feeder arteries. We speculated that PAVMs were already present and were asymptomatic, and that the exertional dyspnea was precipitated by the acute febrile illness. We, therefore, considered the PAVMs as the cause of exertional dyspnea in the absence of lung parenchymal disease and treated the PAVMs percutaneously with vascular plugs. Fever was successfully treated symptomatically.

Our case is a good example where the patient was asymptomatic despite large PAVMs, until fever precipitated dyspnea, and also where large PAVMs can be successfully treated percutaneously.

We recommend that physicians should have high index of suspicion for PAVM while evaluating patients with cyanosis, clubbing, dyspnoea on exertion or hemoptysis and no obvious diagnosis.

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