

Pulmonary Arterio-Venous Malformation (PAVM): A diagnostic dilemma

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ABSTRACT

Pulmonary arterio-venous malformation (PAVM) is an abnormal communication between the pulmonary artery and vein which is most commonly congenital in nature. Sometimes, the clinicians are in a dilemma to reach the diagnosis of this rare disease. Clinical findings are often misleading. ECGs are generally normal. Echocardiographies are suggestive, But CT scan proves to be the diagnostic modality of choice. Here, we report two cases of PAVM that were diagnosed using CT scan at Ibrahim Cardiac Hospital & Research Institute (ICHRI).

Key Words: Pulmonary arterio-venous malformation (PAVM), reticulo-nodular densities, tortuous vessels.

INTRODUCTION

Pulmonary arterio-venous malformation (PAVM) is an abnormal communication between the pulmonary artery and vein. These are most commonly congenital in nature. However, there may be acquired forms that usually occur following juvenile cirrhosis, pulmonary schistosomiasis or also in patients with metastasis of thyroid carcinoma.¹ Morphologically, pulmonary AVMs are divided into two types: 1. Diffuse lesions and 2. Isolated pulmonary AVM. Pulmonary AVMs range from small pinpoint lesions (1mm) to huge tubular or saccular multilobulated structures.² These lesions are quite uncommon but they are an important part of the differential diagnosis of common pulmonary problems such as persistent pulmonary shadow. CT scan can play a very important role in diagnosing this rare anomaly. Here we report a couple of cases of both diffuse and focal varieties of PAVMs in the lung which were diagnosed using CT scans in the Radiology & Imaging Department of Ibrahim Cardiac Hospital and Research Institute (ICHRI).

Case Reports:

Case 1

A six year old boy presented with exertional dyspnoea for 1 year, generalized weakness and fatigue for 5 months and bluish discoloration of lips, tongue, fingers and toes for the last 2 months. The discoloration worsened when he cried and improved during rest and sleep. He didn't have any feeding difficulties or history of cyanotic spell.

Cyanosis of lips, tongue, fingers and toes were clearly visible. This is confirmed by a transcutaneous oxygen saturation of 78 % in room air. There were no peripheral signs of chronic hepatic disease. Biochemical investigations revealed a haemoglobin level of 13.9 gm/dl and a packed cell volume (PCV) of 44%. The serum creatinine level was 0.34 mg/dl. The indurations following Montoux skin test was 5 mm.

Chest radiograph showed reticular opacities and increased vascular markings in both lower zones in para-cardiac locations (Fig-1). Echocardiogram

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revealed normal cardiac anatomy and mild right ventricular hypertrophy. When so called 'bubble test' was performed using agitated saline, LA and LV were filled with bubbles after 5 cardiac cycles, which signify pulmonary arterio-venous malformation. Contrast CT scan of chest showed many centrilobular and reticulo-nodular densities as well as small dilated tortuous vessels distributed in both lungs. These findings were predominant in the posterior basal segments of both lower lobes. Small dilated tortuous vessels represent the capillary telangiectasia in lung parenchyma and are highly suggestive of pulmonary AVM (Fig-2-a,b).



FIGURE 1 : Chest radiograph showing reticular opacities and increased vascular markings in both lower zones in para-cardiac locations.



Figure 2(a)



Figure 2(b)

FIGURE 2a-b : Contrast enhanced axial CT scan of chest showed many centrilobular and reticulo-nodular densities as well as small dilated tortuous vessels distributed in both lungs.

Case- 2

A 13 year boy presented with haemoptysis for two months. On physical examination he was found completely healthy. All his biochemical parameters were normal. But his chest X-ray showed an opacity in the left lower lobe, and on the basis of which he was treated as a case of pulmonary inflammation. But the patient did not respond to treatment; rather his condition was worsening day by day. Echocardiogram of the boy showed normal cardiac anatomy and so called 'bubble test' was positive. So, he was referred to our department for a contrast CT scan of chest. CT scan showed a



Figure 3 (a)



Figure 3 (b)

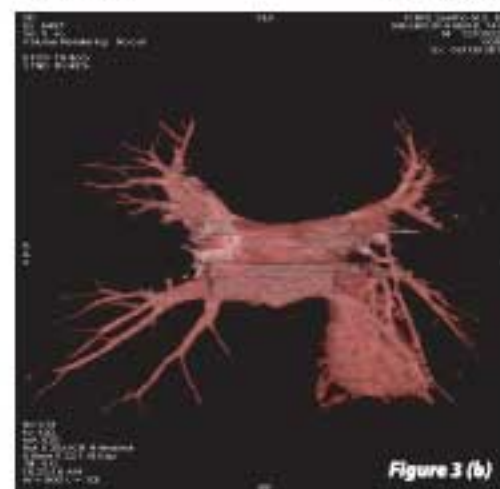


Figure 3 (c)

FIGURE 3a-b-c : Contrast enhanced axial, sagittal & 3D reconstruction CT scan showing a fairly large inhomogeneous soft tissue mass, involving anterior and lateral basal segment of left lower lobe having multiple tortuous vessels within.

fairly large inhomogeneous soft tissue mass, involving anterior and lateral basal segment of left lower lobe having multiple tortuous vessels within. These tortuous vessels appear extending from the left lower hilum possibly from lower segment of pulmonary artery and drains into left atrium through left lower pulmonary vein (Fig: 3-a,b,c). These features of the lesion are highly suggestive of PAVM in left lower lobe.

DISCUSSION

PAVM are vascular lesions that involve abnormal communication between pulmonary arteries & pulmonary veins. Although the pathogenesis of PAVMs is not well delineated, they are considered to result from the venous plexus, leading to formation of tortuous loops and multiloculated sacs. With rupture of intervening vascular walls, a single large saccular PAVMs develops.¹ Most of these are congenital in origin. Majority of the patients with PAMVs has AV malformations located elsewhere, including skin, mucous membranes, and other visceral organs and associated with the autosomal dominant disorder Osler-Weber-Rendu or hereditary hemorrhagic telangiectasia (HHT).³ Approximately 70% of the cases of PAVMs are associated with HHT. Conversely 15 to 35 % of the patients with HHT have PAVM.⁴

Very few cases of PAVMs are diagnosed in neonates, with less than 30 neonatal cases reported out of 500 cases in the literature. However, the clinical manifestations of hypoxia, tachypnoea, and audible bruit over the lung fields in a newborn should suggest the possibility of a PAVM.^{3,5}

Most cases of PAVMs remain asymptomatic and do not present until the fourth decade of life.⁵ The presentation of PAVM may be dyspnoea with associated hypoxia as a result of right to left shunting. The more severe presentations may include massive haemoptysis and hemothorax. In the first case, the young boy presented with generalized weakness, hypoxia and clubbing which is a sign of chronic disease. The hypoxia did not improve with administration of oxygen with non breathing mask. This was a strong pointer towards right to left shunt. Since echocardiography showed normal cardiac anatomy, a strong suspicion of extra cardiac right to left shunting lesion arose.

In the second case the boy presented with only haemoptysis, otherwise he was healthy. But this haemoptysis was not improving after medical treatment, so suspicion of PAVM was kept in mind. In addition to above symptoms, the right to left shunt may facilitate passage of emboli into

the cerebral circulation resulting in transient ischemic attacks, strokes, and cerebral abscess and may be responsible for two-thirds of the CNS symptoms seen with HHT.⁶

The gold standard for diagnosis of PAVM is a CT scan of the chest, which has been shown to be as sensitive & specific as pulmonary angiography and has the advantage of being less invasive.⁷ More recently, the combination of contrast echocardiography and an antero-posterior chest radiograph has been shown to have 100% sensitivity and negative predictive value, with the advantage of lower cost, minimal invasiveness, wide availability, and less radiation exposure.⁸

Patients with microvascular telangiectasis or diffuse AVM may have normal chest radiograph, or only vague increased pulmonary vascular markings at the base and radiographic shadows of PAMV may come and go. The classic radiographic appearance of a focal PAVM is that of a round or oval mass of uniform density, frequently lobulated, more commonly in the lower lobes. In our first case, the chest X-ray showed increased vascular markings and fine reticulonodular opacity in both lower lobes. In the second case, there is persistent rounded opacity in left lower lobe. In CT scan of chest, our first case showed centrilobular and reticulonodular opacities in both lungs, predominantly at the posterior aspect of both lower lobes. In the second case, there was a fairly large mass having multiple tortuous vessels extending from posterior left lower hilum and draining into left atrium through left lower pulmonary vein.

If initial screening is positive, cardiac catheterization should be performed in all patients to define the vascular anatomy and to rule out multiple or bilateral lesions.

PAVMs can be treated either by transcatheter embolization at the time of cardiac catheterization or by surgical resection. Embolization is done using metal coils or detachable silicon balloons.⁹ The procedure of choice for treating a PAVM is therapeutic embolization rather than lung resection. Surgical treatment is indicated in patients with unstable hemodynamics, large fistula occupying major

portion of lung and failed or contraindicated embolization.^{10,11} Finally lung transplant has also been performed in rare cases with severe and diffuse PAVMs. The decision to perform surgery or embolization should be made according to available resources and experience.

Pulmonary arteriovenous fistula should be included in the differential diagnosis of children with severe persistent cyanosis when congenital cardiomyopathy, pulmonary hypertension and pulmonary parenchymal disease are ruled out. The treatment of the patients with PAVM should be initiated early as they tend to increase with age and may increase the risk of respiratory and neurological complications.

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