Case Report

An unusual case of pulmonary arterio-venous fistula (PAVF)

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Abstract

Pulmonary Arteriovenous Fistula (PAVF) is an abnormal communication between the pulmonary veins and pulmonary arteries. Most individuals may have the condition since birth (congenital occurrence), but it can also be an acquired condition. Most individuals with Pulmonary Arteriovenous Fistula also have a hereditary haemorrhagic telangiectasia. The common signs and symptoms of Pulmonary Arteriovenous Fistula are shortness of breath, haemoptysis, chest pain, dizziness, and syncope. Pulmonary Arteriovenous Fistula is treated with embolization and surgery. PAVF are more common in females than males. Complete evaluation of medical history along with a thorough physical examination required to diagnose PAVF. The CT scan is more sensitive than a chest x-ray in establishing a diagnosis. However pulmonary angiography is the gold standard. Many clinical conditions may have similar signs and symptoms. PAVF is associated with variety of complications, some of which may be life-threatening. These may include: Stroke, bleeding in the lung and brain abscess. Typically, individuals with mild conditions presenting no symptoms may not require any treatment. The treatment of Pulmonary Arteriovenous Fistula may include: Embolization and surgery (the later may be required in a cases not responding to embolization). During surgery, the abnormal vessels are removed along with surrounding lung tissue. We report a rare case of large right pulmonary arterio-venous fistula (PAVF) which was misdiagnosed as mass lung in a 30-year-old lady who presented with generalized seizures due to secondary polycythemia, right sided hemiparesis, central cyanosis and clubbing. She had hypoxemia while breathing ambient air and little improvement with 100% oxygen. Diagnosis was clinched by pulmonary CT angiography which revealed a large PAVF.

Keywords: Computerised tomography, hereditary haemorrhagic telangiectasia, pulmonary arterio-venous fistula

Case Report

A 30- year-old lady presented 6 hours after she had convulsions. There was no documented past history of any serious illness and her family members were reported healthy. Physical examination revealed a confused state, clubbing of all fingers and toes [Figure 1], cyanosis, right sided hemiparesis, right parasternal bruit and dyspnoea. General physical examination showed no evidence of telangiectasia. Blood investigations revealed haemoglobin level of 20 g/decilitre and haematocrit of 64%. Chest radiography showed a smooth bilobed opacity in right lung [Figure 2, arrows]. Transthoracic echocardiography [Figures 3 and 4] and ECG [Figure 5] revealed no abnormality. A non- enhanced computed tomography (CT) of brain showed a small infarct in left fronto-parietal cortex [Figure 6]. Pulmonary CT angiography revealed a large pulmonary arterio-venous (AV) fistula [Figure 7], red, yellow and white arrows showing pulmonary artery, draining vein and fistula respectively). Right pulmonary artery coursing into the fistula and draining vein was also seen in 3Dimensional volume rendering tomographic still image [Figure 8]. She was diagnosed to have pulmonary AV fistula causing hypoxemia and cyanosis leading to secondary polycythaemia, hypercoagulable state, cerebral infarct and seizure. She was treated with oxygen, oral levetiracetam, enoxaparin and underwent phlebotomy thrice. She refused surgical intervention. She recovered from hemiparesis within 72 hours and was discharged on the sixth day.

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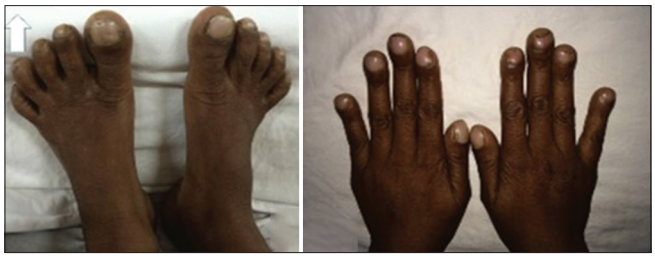

Figure 1: Clubbing of nails

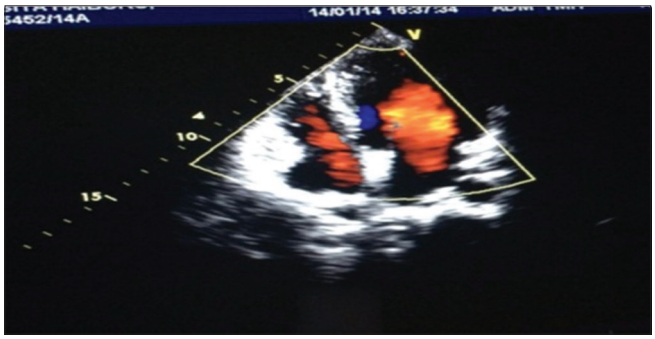

Figure 3: 2D Trans thoracic echocardiography. (Normal study)

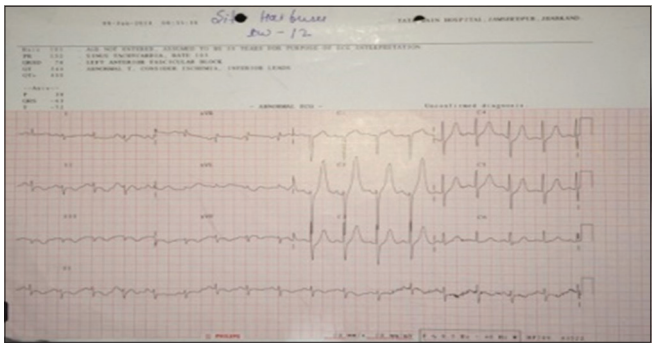

Figure 5: Electrocardiogram-Non specific ST-T changes

Discussion

Pulmonary arteriovenous malformations (PAVM) were first described in 1897 by Churton. They consist of abnormal communications between the pulmonary arteries and the pulmonary vein without intervening vascular bed. Although most patients are asymptomatic, PAVMs can cause dyspnoea from right-to-left shunt. Because of paradoxical emboli, various central nervous system complications occur. 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. Therapeutic options include angiographic embolization with metal coil or balloon occlusion and surgical excision. Since the first reported case in 1897, more than 500 cases have been reported in the literature. It can be congenital (80% cases) and acquired (20%). Congenital PAVMs is mainly associated with the Osler–Weber render disease or hereditary haemorrhagic telangiectasia (HHT).[1] 5-15% of the population with HHT have a PAVM.[2] In patients with HHT, telangiectasia's of the skin and

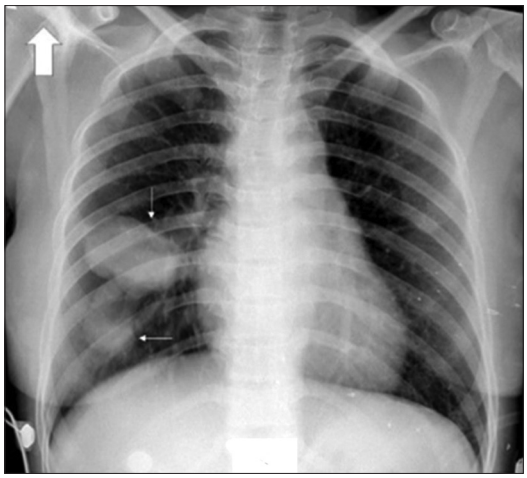

Figure 2: X-ray chest PA view- Bilobed opacity right mid & lower zone

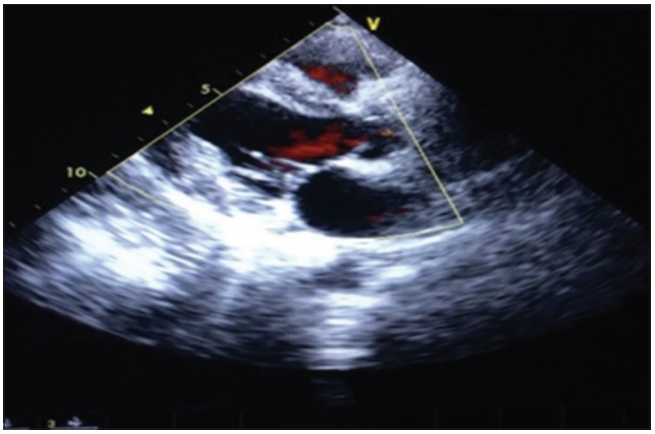

Figure 4: 2D Trans thoracic echocardiography. (Normal study)

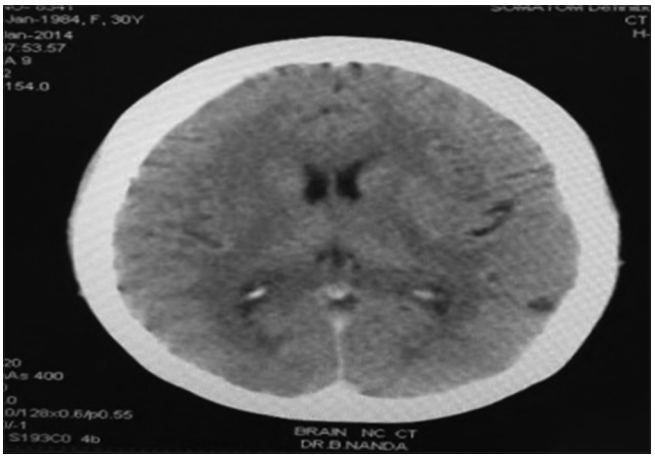

Figure 6: Non contrast CT Brain showed lacunar infarct left fronto-parietal cortex

oral, nasal, and conjunctival mucosa become apparent in the second and third decades of life. The presence of HHT in a patient with a PAVM may be of prognostic value since the patient with coexisting HHT tends to have worse symptomatology, multiple arteriovenous malformations, rapid disease progression, and a higher complication rate. While most patients (70%) with

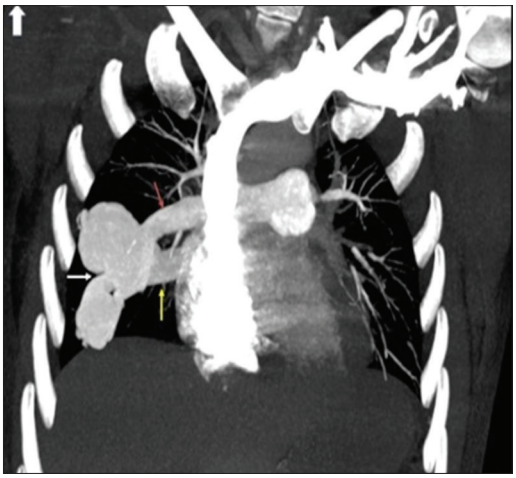

Figure 7: Pulmonary CT angiography showing a large AV fistula with calcification

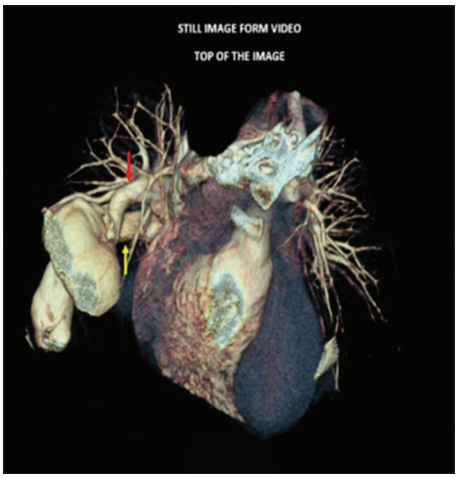

Figure 8: Volume Rendering CT- AV fistula with feeding vessels

PAVMs have the autosomal dominant disease of hereditary haemorrhagic telangiectasia (HHT), they are also found in patients with Secondary or acquired PAVM, although very rare, has been reported in the literature. Causes of secondary PAVM include chest trauma, thoracic surgery, long standing hepatic cirrhosis, metastatic carcinoma, mitral stenosis, infections (actinomycosis, schistosomiasis), and systemic amyloidosis. The incidence of PAVM is 2-3 per 100,000 population. The male to female ratio varies from 1:1.5 to 1.8 in several studies.[3] Majority of cases are diagnosed in the first three decades of life.[4,5] PAVMs may be single or multiple in occurrence and the incidence of single PAVMs ranges from 42% to 74%.[5-7] Most solitary PAVMs are seen in bilateral lower lobes, the left lower lobe being the most common location, followed by right lower lobe, left upper lobe, right middle lobe, and right upper lobe.[4] The majority of multiple PAVMs are also confined to bilateral lower lobes; the incidence of bilateral PAVMs ranges from 8-20%.[8] In a PAVF, blood bypasses the normal oxygen-exchanging pulmonary capillary bed, returning desaturated to the pulmonary veins, which results in arterial oxygen desaturation and cyanosis. This leads to secondary polycythemia roughly proportional to the degree of chronic desaturation. Clinical features correlate with the number and size of the shunt. Since the first successful resection of PAVM in 1942, surgery was the only treatment available until 1978, when Taylor *et al.* reported the first successful percutaneous embolization. The current preferred treatment for the majority of patients with a PAVM is percutaneous embolotherapy using coils or balloons; this method has largely replaced surgical intervention.[9] Embolotherapy, being less invasive, has definite advantages over surgery. Two methods have been used for embolization, that is, balloons and metallic coils. Each method has its advantages, disadvantages and complications.[9] PAVM radiologically may mimic infiltrations, nodules, cavity or mass lesions as also in our case. However, in view of the clinical scenario, oxygenation status and high clinical suspicion, multidetecter CT (MDCT) with 3D construction of pulmonary vasculature was done. The volume rendering technique provides angiographic like images of the pulmonary vasculature, thus giving a complete picture of the malformation, including information on the feeding and draining vessels, that is required before a therapy is planned. The treatment options for a pulmonary vascular malformation are either a surgical excision or embolization. Our patient refused both. Thus, our patient probably had single, large, idiopathic or congenital PAVM in the right lower lobe presenting as seizure which brought the patient to the hospital and breathlessness which the patient ignored and attributed to her household chores. This condition could be misinterpreted as a mass lesion by an untrained eye. Hence, detailed history taking, examination and a strong index of suspicion are of immense value in the diagnosis of this rare disorder, else we will miss this potentially treatable condition. On the X-Ray chest PA view, the lesion looked very much like a tumour. Any attempt to biopsy the lesion would have been catastrophic (and there have been anecdotal incidences in the past), but in this case, the presence of cyanosis and clubbing was a pointer towards something more sinister i.e. Pulmonary AV fistula, which was later proven on CT angiography.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflict of interest

There is no conflict of interest.

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