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Case Report | Medical Sciences

A RARE CAUSE OF HYPOXEMIA IN ELDERLY

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ABSTRACT Pulmonary arteriovenous malformations (PAVM) are rare pulmonary vascular anomaly in which there is 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. Although most patients are asymptomatic, PAVMs can cause dyspnoea from right-to-left shunt. Because of paradoxical emboli, various central nervous system complications have been described including stroke and brain abscess. 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 embolisation with metal coil or balloon occlusion and surgical excision. We report a rare case of pulmonary arterio-venous malformation in a 65 year old elderly male who was suspected to have lung mass in chest x-ray. He presented with breathlessness, anemia, clubbing and hypoxemia which was not improving with oxygen supplementation. Diagnosis was clinched by contrast enhanced computed tomography which revealed PAVMs.

KEYWORDS : pulmonary arteriovenous malformations (PAVM), hypoxemia, clubbing

Case report

A 65 year old male, presented with complaints of exertional breathlessness, intermittent chest pain, palpitations and swelling of the feet since a year and half. He also complained of intermittent epistaxis and hemoptysis since 1 year. He was admitted in the Government Medical College, Aurangabad one year before. At that time, he was found to have atrial fibrillation with congestive cardiac failure and was put on beta blocker & diuretics. He also had anemia, for which he received two packed cells blood transfusions. As his condition improved, he was discharged after a week on beta blockers, diuretics, warfarin and haematinics.

Current admission

He presented again with complaints of exertional breathlessness (NYHA grade 3), intermittent chest pain, palpitation and lower limb swelling since a month. He had hemoptysis and epistaxis four times in the last month. He did not had cough, orthopnea, paroxysmal nocturnal dyspnoea, syncope, abdominal pain, decreased urine output or weight loss.

Past history-

He gave a history of left thoracotomy for cyst removal 45 years back-- operation details are not available.

Atrial fibrillation with anemia since last 1 year on beta blockers, diuretics, warfarin and hemetinics. No addiction history.

On examination- Patient was conscious and oriented. Pulse was 120/min, high volume, irregularly irregular, he had an apex -pulse deficit of 20, BP- 120/70 mmHg right arm supine, RR- 24/min (abdomino thoracic), He was afebrile. He had pallor, clubbing (grade 3), and bilateral pitting pedal edema. JVP was raised (6cm). His saturation on room air was 78% and did not increase with oxygen supplementation.

On cardiovascular examination- transverse surgical scar was present below the 5th Intercostal space. Apex beat was felt at 5th intercostal space in midclavicular line. On auscultation, heart sounds were irregular, a pansystolic murmur was present over the mitral area, which radiated to the axilla and increased on expiration. A pansystolic murmur was present in the tricuspid area which increased on inspiration and a loud pulmonary systolic bruit was present. There was bilateral crackles in the infrascapular area.

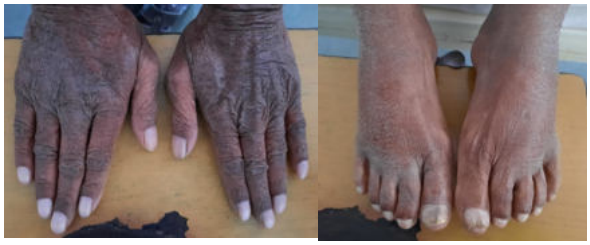

Figure 1- clubbing of nails

Tab Diltiazem for atrial fibrillation and diuretic was started. His haemoglobin at admission was 3.5 and he was given four packed red cell transfusion. Carbimazole 10 mg thrice a day was added as he was found to have hyperthyroidism (TSH- 0.112, T3- 146 , T4- 16.5). In view of persisting hypoxemia (which was not improving with oxygen supplementation) and chest x-ray showing well defined radiopacity in the left hilar border with silhouetting, CECT chest was done and it was suggestive of pulmonary arterio venous malformations(4 in number) in left hemithorax in left upper lobe adjacent to left pulmonary artery and vein. As the patient was symptomatically better, he was discharged on beta blocker, diuretics, tablet dabigatran , hematinics and was referred to higher center for embolization and further management.

Investigations

ABG- pH- 7.36, pCO2-42.7, pO2- 59.6, HCO3- 22.7. Rest of the complete hemogram, renal function test, liver function test, serum electrolytes and blood sugar levels was with in normal limits.

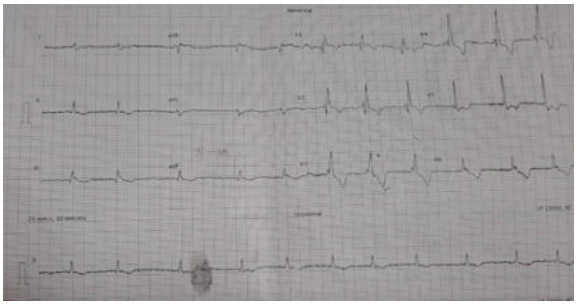

Figure 2- ECG showing atrial fibrillation with complete RBBB

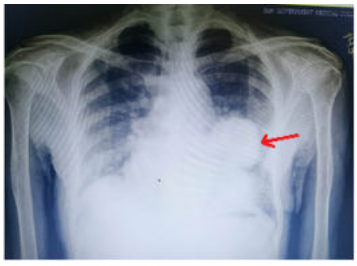

Figure 3- Chest X-ray- well defined radioopacity is noted in left lower midzone silhouetting left cardiac border with cardiomegaly

Ultra sonography abdomen and pelvis- liver span- 13.4 cm
Evidence of 8 x 11.6 x 11cm anechoic cystic lesion is noted in 7th segment of right lobe of liver- simple hepatic cyst.
Spleen- 12.7cm, borderline splenomegaly

CECT chest – few (4) pulmonary arterio- venous malformation largest measuring 9.5 x 5.6 x 6.6 cm in left hemithorax in left upper lobe adjacent to left pulmonary artery and vein.

- **Cardiomegaly**

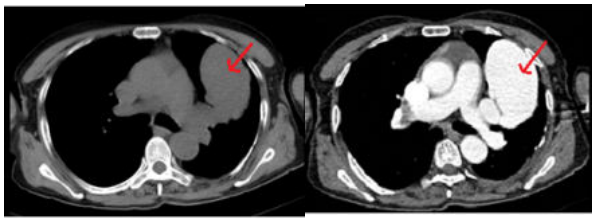

Figure 4- Computed tomography chest- (a) non-contrast study- well defined soft tissue density lesion is noted in left hemithorax in left upper lobe. (b) contrast study- lesion shows enhancement pattern similar to that of great vessels- suggestive of PAVM

2D ECHO- Dilated LA and RA; No RWMA; good LV / RV function (EF-60%)

Grade 2 tricuspid regurgitation, Grade 1 mitral regurgitation
Moderate pulmonary hypertension

DISCUSSION

A pulmonary arteriovenous malformation (PAVM) or a pulmonary arteriovenous fistula is the direct communications between the branches of pulmonary artery and pulmonary veins, without an intervening pulmonary bed[1, 2]. After the first reported case in 1897, more than 500 cases have been reported in the literature[3]. PAVMs can be either congenital or acquired. More than 80% of PAVMs are congenital, of which 47%-80% are related to Osler-Weber-Render disease or hereditary hemorrhagic telangiectasia (HHT). Conversely, it is estimated that about 5%–15% of the people with HHT have a PAVM[1]. The HHT genes in some patients are located on chromosome 9 (9q 33-34 or OWR-1) and in others on chromosome 12 (12q or OWR-2)[4]. Causes of secondary PAVM include chest trauma, thoracic surgery, long standing hepatic cirrhosis, metastatic carcinoma, mitral stenosis, infections (actinomycosis, schistosomiasis), and systemic amyloidosis[1,5]. Pregnancy has been allied with an increased rate of PAVM growth and its associated complications. The pregnancy associated increase in steroid hormone synthesis results in an increased incidence of spontaneous hemothorax secondary to intrapleural rupture of PAVM[6].

The incidence of PAVM is 2–3 per 100 000 population[7]. The male to female ratio varies from 1:1.5 to 1.8[8]. The age at the first presentation ranges from newborn to 70, but most cases are diagnosed in the first three decades of life. PAVMs may be single or multiple in occurrence and the incidence of single PAVMs ranges from 42% to 74%[9]. 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. The majority of multiple PAVMs are also confined to bilateral lower lobes; the incidence of bilateral PAVMs ranges from 8% to 20%[10].

In contrast to systemic arteriovenous malformation, PAVMs do not affect cardiac hemodynamics. Cardiac output, cardiac index, pulmonary capillary wedge pressure, heart rate, blood pressure, and the electrocardiogram are usually within normal limits[3]. The fundamental defect is right-to-left shunt from the pulmonary artery to the pulmonary vein; the degree of shunt is what determines the clinical effects on the patient[1]. Symptoms are usually subacute or even absent if shunting is minimal. If the right-to-left shunt is greater than 20% of the systemic cardiac output or there is reduction of hemoglobin more than 50 g/l, the patient will have obvious cyanosis, clubbing, and polycythemia. In some cases of HHT, cyanosis may be hidden by anemia caused by epistaxis or gastrointestinal blood[1,3].

Asymptomatic patients are common, accounting for between 13% to 55%. So absence of symptoms does not exclude the diagnosis of PAVM. The most common presenting symptom is dyspnea on exertion, which is seen in 31% to 67% of patients[8]. The severity of dyspnea is related to the degree of hypoxemia and the magnitude of the shunt. The majority of the patients with PAVMs tolerate hypoxemia very well and are relatively or completely asymptomatic unless the arterial oxygen pressure is less than 8.0 kPa (60 mm Hg)[1,8].

CLINICAL FEATURES OF PAVMs [1]
Symptoms • Dyspnoea • Palpitations • Chest pain • Epistaxis • Malaena

- Haemoptysis

Signs
- Pallor
- Clubbing
- B/L pedal oedema
- Raised JVP
- Systolic Murmur
- Telengectasia

Complications
- Hypoxemia
- Intrabronchial rupture, haemoptysis
- Intrabronchial rupture ,haemothorax
- Paradoxical embolization
- Polycythaemia
- Pulmonary hypertension
- Endocarditis
- Transient Ischaemic attack
- Cerebrovascular accident
- Migraine headache
- Brain abscess
- Congestive heart failure

Chest radiography is a key diagnostic tool not only in diagnosis but also in the follow up of patients with a PAVM. In about 98 percent of patients, a simple chest radiograph reveals anomalies. The classic radiographic features of PAVM are a round or oval sharply defined mass of uniform density, frequently lobulated, and ranging in size from 1–5 cm in diameter[8].

Contrast enhanced computed tomography is a valuable tool in diagnosis and describing the vascular anatomy of PAVM. The superiority of computed tomography scanning in detecting PAVM is attributed to the absence of superimposition of lesions in transaxial computed tomography views. However, angiography was better able to determine the angio architecture of individual PAVMs than computed tomography[3]. The use of magnetic resonance imaging to diagnose PAVM has been limited compared with that of computed tomography[11].

Contrast echocardiography involves administration of agitated saline or dye into a peripheral vein; it is extremely sensitive in detecting left-to-right shunt but it does not provide quantitative or anatomic detail of the shunt. In patients without right to left shunt, an air bubble or dye may rapidly appear in the right atrium and then gradually disperse as the bubbles become trapped in the pulmonary circulation[12].

In 88 percent to 100 percent of selected patients with a PAVM, the shunt fraction, the fraction of cardiac output that shunts from right to left through a PAVM, is raised. The shunt fraction is most precisely determined by the 100% oxygen method, which requires oxygen saturation and arterial oxygen pressure measurements after breathing 100% oxygen for 15 to 20 minutes. A shunt fraction of equal to or greater than 5% by this method is considered abnormal[13].

A useful addition for the diagnosis and quantification of PAVM is the radionuclide perfusion lung scan. However, as with contrasting echocardiography, the positive outcome is not unique to PAVM, while the diagnosis is essentially excluded from the negative result. Radionuclide scanning also allows the quantification of shunt magnitude, the results of which are comparable to the 100% oxygen shunt calculation method[14].

In diagnosing PAVM, pulmonary angiography is the gold standard. Pulmonary angiography is warranted to confirm the diagnosis in virtually all cases. A pulmonary angiogram not only identifies the PAVM but also further outlines the angioarchitecture of pulmonary vasculature, which is necessary before therapeutic embolisation or surgical resection[9].

Treatment should be offered to all symptomatic patients and asymptomatic patients with lesions less than 2 cm in diameter on chest radiography[9]. Prevention of neurological complications, progressive hypoxia and its related consequences, and high-output heart failure are the main goals of treatment. The morbidity associated with PAVM was up to 50% in untreated patients compared with 3% in patients who received treatment[8].

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 embolisation[15]. For most PAVM patients, percutaneous embolotherapy using coils or balloons is the current preferred treatment; this procedure has largely substituted surgical intervention. Embolotherapy, being less invasive and easy to repeat, has definite advantages over surgery. Embolotherapy is a suitable alternative to surgical intervention in the elderly who are poor surgical candidates, in patients with multiple lesions, and patients who decline surgery[13].

Surgical resection of PAVMs is indicated in patients who fail embolotherapy, develop serious bleeding complication despite embolotherapy, have intrapleural rupture of the PAVM, or have untreatable contrast allergy and lesions not amenable to embolotherapy. Various surgical procedures, including local excision, segmental resection, lobectomy, ligation and even pneumonectomy, have been used. Lung conserving resection, local resection, or segmentectomy is the procedure of choice whenever possible[16].

CONCLUSION

PAVMs are an uncommon clinical problem. The classic triad of dyspnoea on exertion, cyanosis(will be absent if anemia is present-as in this case), and clubbing should alert the clinician to the possibility of a PAVM. This condition could be misinterpreted as a mass lesion 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. Contrast enhanced computed tomography or pulmonary angiography is usually diagnostic. In our case, anemia and hyperthyroidism may have added to his symptoms. PAVMs are known to occur after thoracic surgeries, this patient does give a history of thoracic surgery 45 years ago, but we cannot comment that the pulmonary A-V malformation in this case was congenital or acquired.

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