A misdiagnosed case of Pulmonary Arteriovenous malformation

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Abstract: Pulmonary arteriovenous malformations (PAVMs) are rare pulmonary vascular anomalies, in which there is an abnormal connection between the pulmonary artery and pulmonary vein. Although they are rare, they form the important differential diagnosis for patients with dyspnoea, clubbing and cyanosis. We describe this condition in a 17 year-old male patient with cough, breathlessness, cyanosis, and clubbing for 3 months who was admitted in our hospital for the evaluation for Persistent pneumonia. A chest X-ray showed a homogeneous opacity in the left lower zone. Computed tomography angiography of the thorax confirmed the presence of pulmonary arteriovenous malformation. The patient was treated with embolotherapy and he is doing well.

Keywords: Pulmonary arteriovenous malformations, tomography, angiography.

INTRODUCTION:
Pulmonary arteriovenous malformations (PAVMS) are rare abnormal communication between the pulmonary artery and pulmonary vein. Although it is rare, it can cause hypoxemia, haemoptysis, hemotorax, paradoxical embolization, polycythemia, pulmonary hypertension, endocarditis, transient ischemic attack, migraine headache, brain abscess and congestive heart failure [1]. Chest X-ray and contrast enhanced computed tomography may be used as an initial tool for diagnosis, but pulmonary angiography is the gold standard for diagnosis. Treatment options include surgical excision, radiotherapy, and angiographic embolisation with metal coil or balloon occlusion.

Persistent pneumonia is defined as the one in which the pneumonia symptoms do not clear within 14 days and radiographic picture that do not revert to normal within 4 to 6 weeks. Here, we report a case of PAVM which had been misdiagnosed as persistent pneumonia.

CASE REPORT:
A 17 year-old boy was admitted in our hospital for the evaluation of persistent pneumonia. He complained of productive cough without haemoptysis, and breathlessness on exertion for 3 months. There was a history of bluish discoloration of fingers, toes and lips for 1 year. He is neither a smoker nor a drug addict. For these complaints, he had been treated with multiple antibiotics, but his symptoms did not subside. The past and family histories were insignificant.

On examination, he was normally nourished with normal build. His pulse rate was 88 per minute with blood pressure of 116/70 mm of Hg. He had grade III clubbing, and central cyanosis without pallor, icterus, lymphadenopathy, pedal oedema and telangiectasia. On chest examination, there was a bruit heard over the left infra axillary area. Other system examinations were within normal limits.

His hematological reports showed hemoglobin - 22.5g/dl, total leucocyte count -9,500/mm³, haematocrit - 61.3%, platelets - 2.2 lakhs/mm³, and normal renal and liver function tests. Arterial blood gas analysis showed pH - 7.28, PO₂ - 58.6%, PCO₂ - 39.6%, SPO₂ - 89%. Chest X-ray showed a homogenous opacity in the left lower lobe (Figure 1). Electrocardiography and echocardiography were normal. A computed tomography pulmonary angiography showed a 40 mm arteriovenous malformation (AVM) within the patient’s left lower lobe (Figure 2).
The patient was treated with immobilization which immediately resulted in an increase in oxygen saturation up to 98%. Before embolotherapy, the patient was also treated for polycythemia. At the time of discharge patient’s oxygen saturation was 99% without oxygen supplementation.

**DISCUSSION:**

A 17 year-old boy with a history of cough, breathlessness on exertion for 3 months was misdiagnosed and treated for persistent pneumonia. But, after careful history taking, thorough clinical examination, and appropriate investigations, PAVM were diagnosed in this boy.

PAVMs are rare vascular anomalies in which there is a direct communication between the pulmonary artery and the pulmonary vein, without any intervening capillary bed. It is a rare disorder with an incidence of 2- 3 per 1000,000 population [1]. During the embryological development of the vascular system, which occurs between the 5th and 10th weeks of intrauterine life, a continuous differentiation of the vascular bed occurs, resulting in the creation of separate arterial and venous channels, interconnected by capillaries [2]. When a mistake or halt occurs in this process of vascular differentiation, vascular malformations will appear at different anatomical sites and with variable morphology, depending on the stage of differentiation. PAVM may be acquired or congenital and approximately 70% of the cases of PAVM are associated with Hereditary Hemorrhagic telangiectasia (HHT). PAVM occurs twice as often in the female gender compared with the male gender[3]. PAVM can be either simple or complex. The simple type (80% of cases) consists of a single feeding segmental artery and a single draining vein, and the complex type (20% of cases) has two or more feeding arteries or draining veins. More than half of the lesions are in the lower lung fields, followed by the middle lobes and then the upper lobes. 25% of total blood volume. Symptoms of HHT commonly become noticeable before the age of 20 (for example, epistaxis, and skin telangiectasias). Dyspnoea on exertion is the most common complaint in patients with PAVM and it is seen in about half of patients. Other symptoms attributable to PAVM include the following: haemoptysis (10%), chest pain (6%), finger clubbing (20%), cyanosis (18%) and thoracic murmurs (3%). Our patient had dyspnea on exertion with chronic hypoxia. Thoracic bruit was revealed on physical examination.

Orthodoxy is a decrease in the partial pressure of oxygen in arterial blood (PaO2) or SaO2 that occurs
when one assumes an upright position from the supine position. The fraction of cardiac output that shunts right-to-left circulation is elevated in patients with PAVM; normal values are less than 5%[4]. The classic roentgenographic appearance of a PAVM is that of a round or oval mass of uniform density, frequently lobulated but sharply defined, more commonly in the lower lobes, and ranging from 1 to 5 cm in diameter. In our case, PAVM was in the lower lobe. The sensitivity of chest radiograph alone is 70% in diagnosing PAVM. CT angiography is considered the ‘gold standard test’ for the diagnosis of PAVM with sensitivity over 97%. Our case also confirms this observation [5].

The natural course of PAVM is not benign. These lesions, without treatment, can be associated with a variety of life-threatening complications, such as stroke, brain abscess, hemothorax, and haemoptysis, especially in women. Rupture of a PAVM can occur at any age, independent of lesion size. Without appropriate treatment, mortality exceeds 11% [6].

All symptomatic PAVMs and asymptomatic PAVMs larger than two cm, or if feeding arteries are larger than two mm, should be treated with surgery or embolotherapy because of the risk of paradoxical embolism. The treatment of choice in patients with multiple or bilateral PAVM is transcatheter embolotherapy with balloons or stainless steel coils, and vascular plugs [5].

CONCLUSION:

PAVMs are rare disorders with varying presentations. A proper history taking, thorough clinical examination, and a high degree of suspicion are important for the diagnosis. Though the cause of PAVM in our case is not found, he needs close follow-up to find out the cause.

REFERENCES: