

Short Report

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A rare presentation of pulmonary arteriovenous malformation in a 65-year-old patient

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Abstract

Pulmonary arteriovenous malformation is a rare clinical condition with incidence of 2-3 patients per 100,000 population generally present with symptom like dyspnoea, orthodeoxia. Our case a 65-year-old female presented with symptom of gradually progressive shortness of breath with hypoxemia and CTPA revealed AV malformation with aneurysmal dilatation in poster-basal segment of left lower lobe 4.8 x 4.5 cm in measurement. Pulmonary AV malformation is a rare pulmonary condition, HHT is the most common underlying cause. CTPA is investigation of choice to identify such PAVMs and Management options are surgical, like embolization of feeding artery by coil or inflatable balloon, ligation of feeding artery, lobectomy, segmentectomy and Pneumonectomy.

Keywords: Pulmonary arteriovenous malformation (PAM); Hereditary haemorrhagic telangiectasia (HHT); Right to left shunt.

Abbreviations: PAVM: Pulmonary Arteriovenous Malformation; HHT: Hereditary Haemorrhagic Telangiectasia; MMRC: Modified Medical Research Council; CTPA: CT Pulmonary Angiogram; CTVS: Cardio-Thoracic and Vascular Surgery.

Introduction

A pulmonary arteriovenous malformation (PAVM) is a rare pulmonary disorder marked by an abnormal right-to-left shunt, which impairs normal gas exchange and the filtration of venous blood within the pulmonary circulation. Commonly linked with hereditary haemorrhagic telangiectasia (HHT), PAVMs can give rise to serious complications, including ischemic stroke, myocardial infarction, cerebral abscess, massive haemoptysis, and hemithorax. Incidence of PAVMS is around 2-3% per 100,000 population most of the cases are associated HHT (Hereditary Haemorrhagic Telangiectasia) [1,2].

Case description

A 65-year-old female was admitted in our hospital with complain of shortness of breath for 3 years, gradually progressive (initially march grade 1 and progressed to grade 4 since last 3 month) with associated history of dry cough for same period. On examination her oxygen saturation was 70 percent on room air and she also have grade 2 clubbing. In respiratory system ex-

amination there was equal air entry on both side with no added sound. ABG (arterial blood gas) shows type 1 respiratory failure. Chest x ray was done and findings were normal. Possibility of pulmonary embolism or any vascular anomaly was suspected. In view of that CTPA was done and it shows tuft of vessels with aneurysmal dilatation in postero basal segment of left lower lobe suggestive of arterio-venous malformation. It also reveals feeder is arising from left pulmonary artery and draining in to left pulmonary vein causing significant right to left shunt. Largest aneurysmal dilated segment measuring 4.8 x 4.5 cm (Figures 1,2).

Discussion

Pulmonary AV malformations are very rarely seen in clinical practice. Incidence is 1:50000 and more commonly seen in female. Most of the PAVMs cases are congenital and most common cause is HHT. It can also be due to acquired due to congenital heart surgery, chest trauma, cirrhosis of liver, schistosomiasis, tuberculosis, metastatic lung disease and bronchi-



Figure 1: Aneurysmal dilatation at left postero basal segment.

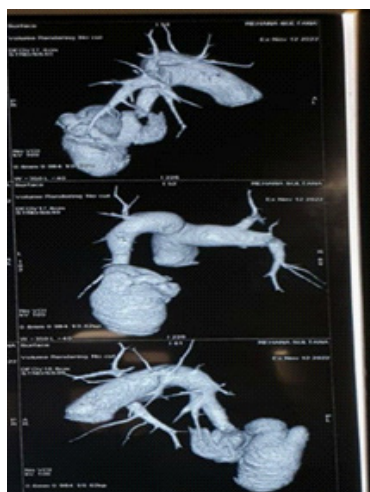


Figure 2: CTPA showing feeder vessel arising from left pulmonary artery and draining in left pulmonary vein.

ectasis etc. Abundant formation of TGF beta binding protein, endoglin has been currently identified as mechanism of pulmonary AV malformation in HHT 1 [3]. Generally, PAVMs classically present with dyspnoea, cyanosis, clubbing but most of the patients remain asymptomatic. Complications include cerebrovascular accident, brain abscess, paradoxical embolism, hemoptysis due to intrabronchial rupture and hemothorax due to intrapleural rupture [4]. CTPA is investigation of choice to identify such PAVMs and Management options are surgical, like embolization of feeding artery by coil or inflatable balloon, ligation of feeding artery, lobectomy, segmentectomy. Pneumonectomy [5]. In our case the PAVM was quite large and it was causing significant right to left shunt leading to significant restriction of daily routine activities. Patients with PAVM usually present early in life, either shortly after birth, during adolescence, or in middle age, typically between 30-40 years. In our case, however, the PAVM was quite large, and the patient developed the syndrome much later, making it a rare presentation of PAVM. After initial stabilisation and conservative management patient was then transferred to CTVS department for definite surgical management.

Conclusion

PAVM is rare disease may be asymptomatic or present late. Patient having significant hypoxemia with normal chest X ray, after ruling out pulmonary embolism we should think of PAVM and CTPA is the investigation of choice. Patient with significant should undergo surgical management.

Conflict of interest: There is no conflict of interest in this report.

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