



Case Report

Pulmonary Arterio-Venous Malformation (PVAM) Presenting as Recurrent Hemoptysis

Authors

Mohd Yousoof Dar*¹, B K Menon², Sarfaraz Jamal³

¹Senior Resident [MD,DM], ²Professor [MD] ³Junior Resident [MBBS]

Department of Pulmonary Medicine, Vallabhbhai Patel Chest Institute, University of Delhi

Corresponding Author

Mohd Yousoof Dar

Email: yousufhumi@gmail.com

Abstract

17 years old non-smoker male farmer presented with complaints of recurrent non massive hemoptysis for last 1 year with past history revealing history of un-noticed blunt trauma to right lower chest two years back. Family evaluation was non-revealing and treatment history was intake of ATT (anti-tubercular treatment) with no response. Systemic examination revealed no significant abnormality. Investigations showed-normal renal profile, liver function, Coagulation profile, ANA and also bronchoscopy were non-revealing. Radiograph chest showed a radio-opaque shadow in right lower zone, which on CT(computerized tomography) pulmonary angiography revealed to a large PAVM(pulmonaryarterio-venous malformation) in Lower lobe right of lung. This case illustrates that how a common pulmonary symptom of hemoptysis can be presenting manifestation of a rare pulmonary condition known as PVAM and possibly blunt trauma to chest was the cause of this rare condition in our case.

Key words: PVAM-pulmonary arterio-venous malformation.

Introduction

Pulmonary arteriovenous malformations (PAVM) are rare pulmonary vascular anomalies and are probably the most common anomalies of the pulmonary vascular tree. These abnormal vascular structures provide a direct communication between the pulmonary arterial and pulmonary venous circulations with bypassing of capillary network.^[1] PVAM have large variation in size and complexity. The true anatomic shunts of PAVMs are usually distinct and are distinguished from the diffusion-perfusion defects that arise in patients

with intrapulmonary vascular dilations secondary to the hepato-pulmonary syndrome.

Cases

17 years male farmer presented with chief complaints of coughing of blood (hemoptysis) 1 year.

It was insidious in onset, paroxysmal, bright red in color, around half a tea spoon (aprox.2.5 ml) in amount in each episode with frequency of around 1-2 episodes per day and repeated episodes with normal intervals of weeks to months. There was

no history of cough otherwise, wheezing/ breathlessness, any nasal discharge/ epistaxis, maleana, hematuria or easy brusibility. There was no significant drug exposure, no past significant medical history and no such illness in any family member. Patient was a non-smoker and had no high risk behavior. Patient had history of ATT(anti-tubercular treatment) intake prescribed by a physician on radiological basis. Patient stopped ATT on his own after 2 months because of no relief in his symptoms. He had occasional use of cough syrups during episodes.

On examination patient was conscious, oriented and afebrile. Vitals were stable with pulse of 78 beats / min, regular, no special character, and

blood pressure of 110 / 72 mm Hg, temperature 98.6⁰ F, respiratory rate of 16/minute and Spo₂ 97% room air. There was no pallor, icterus, cyanosis, lymphadenopathy, clubbing or pedal edema. There was no evidence of any mucocutaneous telenjectasias. Chest examination revealed bilateral vesicular breath sounds with no adventitious sounds and rest of systemic examination was also normal. Investigations revealed normal CBC, renal profile, liver function and normal coagulation profile. X ray chest P/A view showed a heterogeneous radio-opaque shadow in right lower zone with fluffy margins suggestive of alveolar nature.

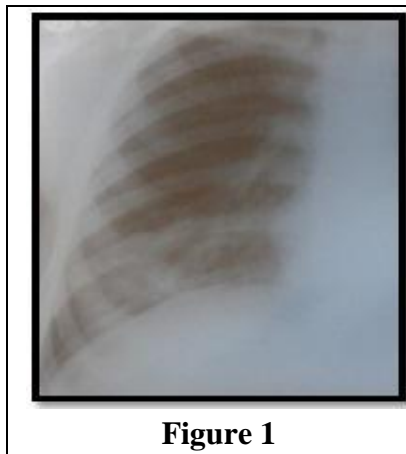


Figure 1

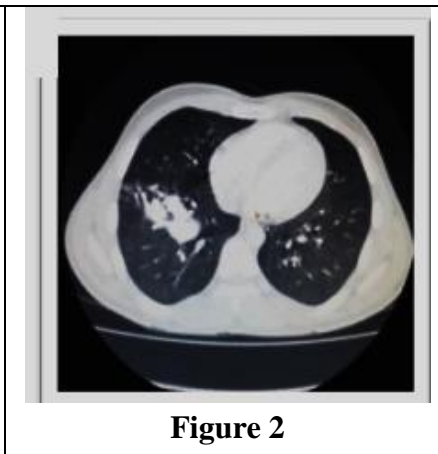


Figure 2

Patient was carrying with him an old chest x-ray (figure 1) and CT (computerized tomography) thorax(figure 2) performed 3 months back which was reported as having a soft tissue shadow with areas of tree in bud suggestive of infective pathology. Patient workup for ANA, P- ANCA and C- ANCA was negative and HIV 1 and HIV-2 was non- reactive. As patient was not producing any sputum or blood at presentation, so he was planned for bronchoscopy. Bronchoscopy was done under conscious sedation and it showed normal tracheobronchial tree, no endobronchial lesion and BAL (broncho-alveolar lavage) was grossly non hemorrhagic. BAL cytology showed no hemosiderin laden macrophages and was negative for malignant cells seen. BAL culture revealed no growth after 48 hours, BAL staining for AFB (acid fast bacilli) and staining for fungus

was negative. TBLB (transbronchial lung biopsy) from multiple segments showed no granuloma, no necrosis, no inflammatory changes and no signs of malignancy. Old CT chest scan again reviewed and possibility of a lobulated lesion was made. Patient history was again reviewed which revealed history of blunt trauma in right lower chest by a cat wheel 2 years back but which needed nothing other than over the counter pain killers and patient did not get a formal medical checkup. So diagnostic possibility of? Vascular malformation was made because of lobulated nature of lesion on CT thorax. So patient underwent a CT Pulmonary angiography (figure 2-6) which revealed a large AV malformation in right lower lobe. Family evaluation (by clinical examination and x-ray chest) showed no evidence of HHT in any family member.

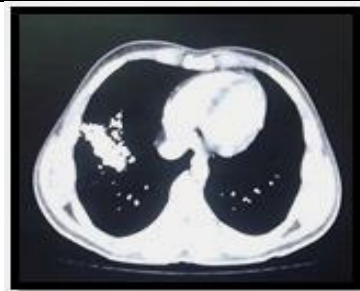


Figure 3

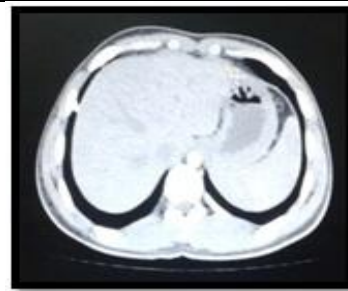


Figure 4



Figure 5



Figure 6

Final diagnosis was Pulmonary A-V malformation possibility traumatic. Echocardiography showed normal chambers, normal LVEF (left ventricular ejection fraction) of 67% and no signs of pulmonary hypertension. So patient referred to thoracic surgeon and patient operated with wedge resection as malformation was large and coiling was not available. Post-surgery patient has no fresh episodes of hemoptysis and he is doing well.

Discussion

Pulmonary A-V malformations: AVMs represent abnormal communications between the pulmonary arterial and venous systems that bypass the capillary bed and hence lead to a right-to-left (R-L) shunt. Their frequency varies, occurring in roughly 10 to 20 persons per 100,000.^[2] In a recent study by contrast chest computed tomography (CT) scanning, PAVMs were estimated to affect 38 per 100,000 individuals (95% confidence interval [CI] = 18-76).^[3] Pulmonary AVMs may be classified as simple with a single feeding and draining vessel (80% of cases), or complex, with 2 or more feeding or draining vessels (20% of cases). Up to 65% of pulmonary AVMs are found in the lower lobes of the lung, which is believed to be due to

gravitational reasons and hence the shunt fraction tends to increase when patients stand up.^[4]

Etiology: Most common cause of PAVMs is inherited vascular disorder called hereditary hemorrhagic telangiectasia (HHT, or Osler-Weber-Rendu syndrome). Hereditary hemorrhagic telangiectasia (HHT) has been reported to account for 50% to 90% of all pulmonary AVMs. This condition affects the arteries of the nose, skin, brain, lung, and gastrointestinal tract.^[2] In the absence of HHT, PAVMs may develop sporadically, as a result of surgical treatments for several forms of complex cyanotic congenital heart disease, chronic lung infections like tuberculosis (resmeuns aneurysm), actinomycosis and trauma.^[5,6] Sporadic PAVMs are usually single, so multiple PAVMs always raisesuspicion of underlying HHT.^[7]

Clinical features: Pulmonary AVMs frequently go unrecognized until the late teens. Most individuals with HHT usually develop recurrent non-traumatic nosebleeds, but are otherwise often minimally symptomatic. In HHT telangiectasia's are common and are seen around face and oral cavity. In advanced cases symptoms include dyspnea, fatigue, cyanosis, and orthodeoxia (decreased arterial oxygen content while upright),

all due to right-to-left shunting of blood through the pulmonary AVM.^[8] The most serious complications of pulmonary AVMs are potentially fatal hemoptysis or hemothorax (in up to 10% of patients).^[4] This risk is reportedly greater in pregnant women during childbirth.^[9] The other complications are neurologic sequelae which develop as a result of the unfiltered passage of paradoxical emboli through the pulmonary system to the brain resulting in stroke and brain abscess.^[10] Neurologic symptoms may be the presenting symptoms in up to 40% of patients.^[10] Screening Patients: Pulmonary AVM screening is recommended for all families with HHT as importance of screening programs for HHT is highlighted by the high proportions of patients with HHT/PAVM who are undiagnosed at the time of their PAVM-induced ischemic stroke or cerebral abscess (in one series, 66.7% and 64.3% respectively).^[10]

Pulmonary function tests: With large R-L shunts (>20%), the carbon monoxide diffusing capacity (DLCO) is often moderately reduced (71% to 78%), but in the majority of cases with smaller R-L fraction, DLCO is equal to or greater than 90% of predicted (interquartile range, 76% to 100%). Also vital capacity is generally normal in majority of cases. Even the work capacity is well preserved in patients with PAVM even when arterial oxygen saturation drops to less than 80% on exercise.^[11]

Complications: Pulmonary hypertension is believed to be common in PVAM because of high blood flow in pulmonary circulation but actually overall prevalence of pulmonary hypertension is relatively low. In one echocardiographic study of 68 HHT patients, estimated systolic PPA values (40 to 58 mm Hg) were above the normal range in 9 (20.5%).^[12] Catheter-based studies in a group of 143 PAVM/HHT patients undergoing PAVM embolization, identified values for mean PPA as 13 (11 to 16) mm Hg, compared to normal values of 7 to 19 mm Hg.^[13]

Diagnosis: Classically PAVM are suspected in persons with mucocutaneous telangiectasia's who

present with cyanosis, clubbing, a vascular bruit, and characteristic chest radiographs features. The classic appearance of a PAVM on chest radiography and chest CT pulmonary angiography consists of a circumscribed, rounded, soft tissue mass or nodule of any size associated with enlarged feeding and draining vessels.^[3]

Contrast Echocardiography: This is method of choice which demonstrates intrapulmonary shunts by tracing the circulatory transit of microbubbles generated by intravenously injected echo-contrast material. CE consistently detects more intrapulmonary R-L shunting in HHT patients than any other PAVM screening modality.^[14] Radionuclide perfusion lung scan is a another useful adjunct in the diagnosis and quantification of PAVM. However, as with contrast echocardiography, a positive result is not specific for PAVM, whereas a negative result essentially excludes the diagnosis.^[15]

Management: Percutaneous transcatheter embolization, which was introduced in 1978, is now the treatment of choice for the vast majority of patients. Pulmonary AVM treatment is recommended for symptomatic patients or those AVMs with a feeding artery diameter 2-3 mm.^[16] Transcatheter embolotherapy with stainless steel coils or detachable balloons is most commonly performed.^[16] Furthermore, it has been suggested that smaller malformations or smaller feeding vessels may become enlarged after successful embolotherapy of larger AVMs. Therefore, long-term follow up, including chest CT examinations every 1 to 2 years, is recommended.^[17]

Surgery: Surgery remained the treatment of choice until the 1980s but was never the ideal solution for the multiple PAVMs of HHT; more recently, however, surgery has been a useful adjunctive therapy for selected cases. It is used in cases where PAVMs are single or sufficiently localized for thoracoscopic resection and when embolization is not feasible.^[18] In emergency situations, particularly associated with massive hemoptysis, lobectomy or pneumonectomy may be appropriate.^[18] Lung transplantation has been

undertaken in a few patients with severe hypoxemia secondary to diffuse disease.^[19]

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