ORIGINAL ARTICLE

PULMONARY ARTERIOVENOUS MALFORMATION (PAVM): RARE CAUSE OF HEMOPTYSIS

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Pulmonary arterio-venous malformation is an abnormal communication between pulmonary arteries and pulmonary veins. These lesions are rare and are an important part of the differential diagnosis of common pulmonary problems such as hypoxemia and pulmonary nodules. Pulmonary arterio-venous malformation was diagnosed in a 33 year old man who presented with recurrent episodes of haemoptysis and also had taken anti-tuberculous treatment (ATT) before. Pulmonary angiography revealed fistulous communication between right bronchial artery and right pulmonary vein. He was treated by lobectomy.

Keywords: Arterio-venous malformations, Angiography, Lobectomy

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INTRODUCTION

Massive haemoptysis is a potentially life-threatening condition that requires prompt management. The most common cause is bronchiectasis, and pulmonary arterio-venous malformation (PAVM) remains the most unusual cause of this condition. First described in 1897, PAVM is an abnormal communication between the pulmonary artery and the pulmonary vein, which is most commonly congenital in nature.[1] PAVM arises as a result of communications between pulmonary arteries and veins in 95% of cases, causing right to left shunting. Whereas, communication between systemic arteries and pulmonary arteries/veins is extremely rare constituting only 5% of cases (left-to-right shunting).[6] Systemic vessels most commonly feeding these PAVM arise from bronchial arteries, less so from the internal mammary artery or even directly from the descending thoracic aorta. Here we report a case in which idiopathic fistulous communication existed between a branch of bronchial artery and pulmonary vein tributaries that was treated by right lower lobectomy as an emergency case. We report this case due to its rarity as only few cases have been reported to date posing diagnostic and therapeutic limitations.

CASE REPORT

A 33 years old previously healthy male with no history of hereditary hemorrhagic telengiectasis (HHT) presented with three years history of recurrent haemoptysis and massive haemoptysis of one day. Past history was significant for Anti Tuberculous Treatment (ATT) for nine months and one hospitalization during which all relevant baseline and radiological investigations were within normal limits. Last episode of haemoptysis six months ago brought him to radiology department where High resolution computer tomography Scan (HRCT) chest revealed vague areas of ground glass attenuation in right lung base postero-laterally (Figure-1).

Conventional pulmonary angiography revealed fistulous communication between inferior division of right bronchial artery and a tributary of pulmonary vein which was successfully embolized using polyvinyl alcohol (PVA) particles and push-able platinum coil (Figure 2). He remained asymptomatic for three months afterwards until he presented through emergency room with one day history of massive haemoptysis. He reported coughing up approximately 800 mL of fresh blood during a 24-hour period before arrival and suffered an episode of hypoxia immediately after admission. During the examination he continued to cough up small amounts of bright red blood. Vital signs were notable for a pulse rate of 110 bpm, blood pressure of 100/70 mmHg, respiratory rate of 25 breaths per minute and oxygen saturation of 96%. His extremities were slightly cold without clubbing and cyanosis. The skin was clear without a rash. The head and neck examination was notable for the presence of blood in the oropharynx and clear nares. He had reduced vocal fremitus and decreased breath sounds. Low pitched rhonchi were audible at right lower chest. However, rest of the physical examination was unremarkable. He was admitted to the intensive care unit, resuscitated and an urgent CT Pulmonary angiogram was arranged showing attenuated right posterior basal segment with collapsed right lower lobe. Urgent fibre-optic bronchoscopy was performed and showed blood clots obstructing the right lower lobar bronchus but no definite site of active bleeding was identified. Because of the patient's continued haemoptysis, haemodynamic instability, and a failed previous embolo-therapy, an emergency right postero-lateral thoracotomy and right lower lobectomy with pleural flap and right bronchial artery ligation was planned. During the induction of anaesthesia patient again started haemoptysis. Through a rigid bronchoscope, a 4 Fr Fogarty catheter was placed in

right lower lobe bronchus and balloon inflated. The right lower lobe parenchyma showed marked vascular congestion with no evidence of sub-pleural extension, so a standard right lower lobectomy was performed. The patient had an uneventful recovery. Follow-up 6 months after surgery showed the patient remained well and free of symptoms. Histopathology of resected specimen was consistent with pulmonary arterio-venous malformation right lower lobe (Figure-3).

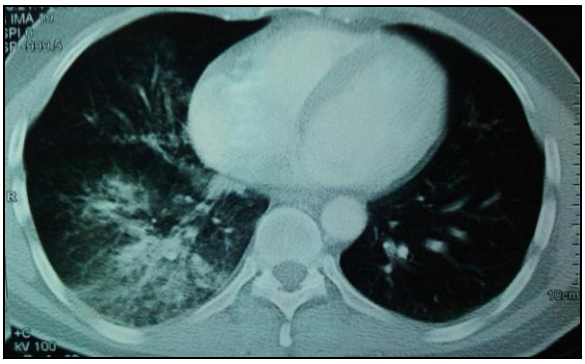

Figure-1: HRCT Scan chest showing vague areas of Ground Glass attenuation in right base postero-laterally and right middle lobe

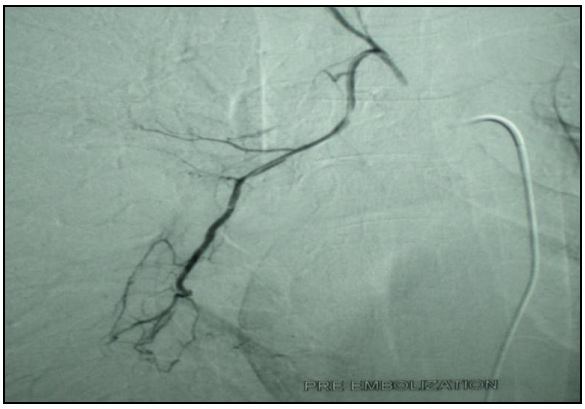

Figure-2: Conventional pulmonary angiography showing fistulous communication between inferior division of right bronchial artery and a tributary of pulmonary vein which was successfully embolized

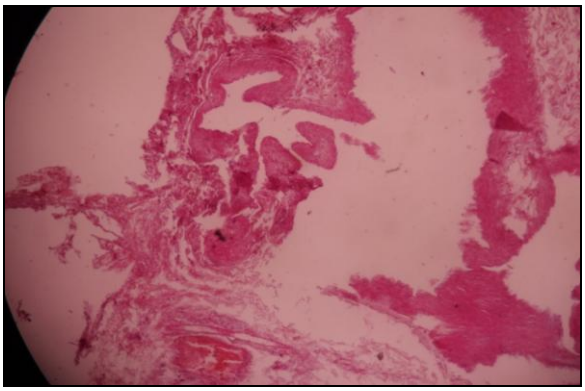

Figure-3: Histopathology showing PAVM

DISCUSSION

Although the PAVMs pathogenesis is not well delineated, they are considered to result from incomplete resorption of the vascular septa. These vascular septa separate the arterial plexus and the venous plexus, which normally anastomose during fatal development. With the rupture of intervening vascular walls, a single large, saccular PAVM develops.[2] Women are affected twice as often as men, but there is a male predominance in newborns.[3] Approximately 10% of PAVM cases are identified in infancy or childhood; however, the incidence gradually increases through the fifth and sixth decades of life.[4]

Pulmonary arterio-venous malformation may be simple (single pulmonary artery-to-pulmonary vein communication) or complex (multiple feeding arteries and draining veins)[5], and single (40%), multiple (40%), or bilateral (20%). The vast majority of PAVM are congenital; 36% of single lesions and 57% of multiple lesions are associated with HTT. Fifty-three to 70% of PAVMs are found in the lower lobes.

The work-up of patients with suspected PAVM should start with a detailed history of epistaxis, dyspnoea on exertion, or family history of telangiectasia. Many patients are asymptomatic, though 6% have brain abscesses[6] due to bacteria bypassing the natural pulmonary vascular filter and lodging in the cerebral circulation. Other frequent extra pulmonary symptoms and signs are headache in 43%, transient ischemic attacks in 57%, and cerebro-vascular accidents in 18%. Intra-cerebral abscess occurs in 33% of the cases. A detailed physical examination, with a specific search for chest bruits (30%), nasopharyngeal telangiectasia (33%), clubbing, or cyanosis is required. The classical triad of cyanosis, polycythaemia and clubbing has been noted in approximately 20% of the patients. Contrast echocardiography is an excellent tool for evaluation of cardiac and intrapulmonary shunts.[7] Radionuclide perfusion scan is also useful in the diagnosis of PAVMs, and may be used as an adjunct to the contrast echocardiography or alone. The presence of a PAVM and its vascular anatomy can also be evaluated by contrast enhanced chest tomography. Despite advances in the different techniques, contrast pulmonary angiography remains the gold standard in the diagnosis of PAVM, and is usually necessary if resectional or obliterative therapy is being considered.

Treatment of PAVM should be based on the size, number, location of the lesions and the specific complications, as well as the patient's

general condition. Traditional indications for treatment have been progressive PAVM enlargement, paradoxic embolization, and symptomatic hypoxemia. As PAVM has a tendency to progression and complications, the most radical and least invasive method of treatment should be used. It is recommended that all symptomatic PAVM and PAVM >2cm in diameter be treated with either surgery or percutaneuos transcatheter embolization, and nowadays embolotherapy (balloon and coil embolization) is considered first-line therapy. White *et al*[4] reported 276 cases of PAVMs successfully treated by balloon embolo-therapy with an overall complication ratio of 1%. The complications of embolo-therapy, such as pleuritic chest pain, pulmonary infarction, and deep venous thrombosis, have in general been infrequent and self-limited. Such complications, as well as paradoxical embolization of coils and balloons may occur especially in patients with PAVMs with large feeding arteries. Because most fistulas are located sub-pleurally, they can be removed with conservative local resection. Before 1978, surgical resection was the only method available; ligation, local excision, segmentectomy, lobectomy, or pneumonectomy was performed in most cases, though some clinicians opted for conservative management and observation in some asymptomatic patients. For large and centrally localized lesions, or for lesions with large feeding vessels and failed previous embolization, as in this case, surgery is still a proposed treatment modality. It carries little risk and when properly performed in well-selected patients, it is associated with minimal morbidity and mortality, with only rare postoperative recurrences.[8]

CONCLUSION

Pulmonary arterio-venous malformation is amongst rare causes of massive haemoptysis and can present both a diagnostic as well as therapeutic challenge and should be considered in differential diagnosis of recurrent haemoptysis refractory to conventional treatments.

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