Life-threatening Haemothorax: A Rare Presentation of Pulmonary Arteriovenous Malformation

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Abstract
Arteriovenous malformations of the lung are rare pulmonary vascular disorders which can suddenly lead to life threatening complications. Haemothorax due to rupture of a pulmonary arteriovenous malformation (PAVM) is very rare. We report here a case of a 39 year-old lady who presented with an acute onset of shortness of breath due to right-sided massive haemothorax and was subsequently detected to have pulmonary as well as cerebral arteriovenous malformation (CAVM).

Introduction
Pulmonary arteriovenous malformations (PAVMs) are abnormal direct communications between pulmonary arteries and pulmonary veins, resulting in a low resistance right to left shunt. Since the first reported case in 1897, more than 500 cases have been reported in the literature.

Approximately 70% of patients with PAVMs are associated with Hereditary Haemorrhagic Telangiectasia (HHT); likewise 15-35% of patients with HHT have PAVMs. So a diagnosis of PAVM requires screening for HHT and vice versa.

Though up to 50% of PAVMs may be asymptomatic, they are not benign lesions as neurologic complications like embolic strokes or sudden death may occur. Approximately 5% develop symptoms due to right-to-left shunting of blood. Less frequently, the dilated thin walls of the aneurysmal centre of a PAVM may spontaneously rupture leading to life threatening haemoptysis (~11%) or haemothorax (<1%). We report a case of life threatening haemothorax in a 39 year-old woman detected to have multiple PAVMs and cerebral AVM (CAVM), a “suspected HHT”, treated with surgical resection of the bleeding PAVM.

Case Report
A 39 year-old housewife presenting with shortness of breath and a right-sided haemorrhagic pleural effusion was admitted for evaluation.

She was apparently well till about 20 days back when she developed a sudden onset of shortness of breath and a right-sided pleuritic chest pain necessitating admission in a local nursing home. She had no wheeze, orthopnoea, paroxysmal nocturnal dyspnoea, pedal oedema, cough, haemoptysis or fever. Right sided pleural effusion was diagnosed and 800 ml of haemorrhagic pleural fluid was aspirated.

Two days after discharge she had a ‘blackout’ and a fall in her home and was admitted to the emergency of our institute. She was found to be very dyspnoeic, pale and was hypotensive – requiring resuscitation with 3 units of blood transfusion.

Intercostal drain was put in the right pleural space draining about 1500ml of bloody fluid. Subsequently she was transferred to our unit.

The patient had several episodes of dizziness with brief loss of consciousness without any tongue bite, incontinence, weakness, dysphasia, vertigo, palpitations or headaches over the last 5 years.

She also admitted to many episodes of epistaxis since her twenties, which however did not require any local treatment or blood transfusion.

The patient had no history of diabetes, hypertension, ischaemic heart disease, deep vein thrombosis, arthritis, tuberculosis, previous chest trauma, malignancy, epilepsy, addictions, intake of oral contraceptives or any risk factor for HIV infection.

Her brother (50), sister (32) and daughter (17) were well.

On examination, the patient had pallor but no cyanosis, peripheral edema, clubbing, lymphadenopathy, jaundice or rash. The JvP was not raised, the pulse 110/min., bP 100/70 mmHg, respiration 20/min. The oxygen saturation (SpO2) was 85% with supplemental O2 @ 6L/min via a face mask.

The upper respiratory tract was normal. There was right-sided pleural effusion without a mediastinal shift; a continuous murmur (II/VI) was present in the right parasternal 2nd / 3rd intercostal space. Other system examinations were normal.

Her initial chest X-ray (Fig. 1) showed a right sided moderate pleural effusion with a 3cm by 3cm lobulated mass shadow in the right mid zone. Her subsequent chest X-rays (Fig. 2) showed an opaque right hemithorax.

The haemoglobin was 8.5gm/dl, TC 8500/mm3 (N65, L30); platelet count, coagulation profile and blood biochemistry were normal. Pleural fluid was grossly haemorrhagic (haematocrit 18%), with RBCs> 1,600,000 /ml, and negative for malignant cell. Bronchoscopy did not reveal any intra bronchial pathology. Echocardiography revealed a mild TR but was otherwise normal. ABG on room air showed pH: 7.432, PaCO2: 35.8 mmHg, PaO2: 39.7 mmHg, SaO2: 79.6%.

CT pulmonary angiogram (Fig. 3) show dilated upper lobar branch of the right main pulmonary artery associated with a dilated nidasus in the right upper lobe and a dilated draining vein draining into the left atrium favouring a pulmonary arterio venous malformation. Another smaller AVM is seen in right lower lobe, with feeder from lower lobare branch of main pulmonary artery.
CT brain showed a calcified nodule in the left frontal lobe. MR angiogram of brain (Fig. 4) shows an AVM in the left anterior frontal cortex with feeder from A-3 segment of left anterior cerebral artery.

Right sided thoracotomy under GA showed gross adhesion between lung parenchyma and parietal pleura; decortication was done. There was aneurysmal dilatation of the AVM which was full of clots. There was massive bleeding from the malformed vessels which were directly connected to the left atrium. The aneurysmal sac was excised, the malformed vessels closed. The patient received 8 units of blood transfusion in the perioperative period. In the postoperative period her Spo2 increased to 93-94% with 2 lit/min. of supplemental O2.

After discharge she is under follow-up and has been advised embolotherapy for her remaining PAVM in right lower lobe and CAVM. She is advised regarding antibiotic prophylaxis before any dental and surgical procedures.

**Discussion**

Pulmonary AVMs are thin-walled abnormal vessels that replace normal capillaries between the pulmonary arteries and veins, often resulting in bulbous aneurysmal sac-like structures. Since 70% of the cases of PAVMs are associated with HHT, finding of PAVM should prompt a thorough review of the patient and her/his family. Current consensus diagnostic criteria for HHT require the presence of 3 of the 4 key features for a “definite” diagnosis – namely spontaneous recurrent epistaxis, mucocutaneous telangiectasia, visceral involvement and an affected first-degree relative. The term “suspected HHT” is used if two features are present – as in our patient (epistaxis and visceral involvement but no telangiectasia or family history). Multiple PAVMs (2 in our patient) favour an association with HHT, in which case AVMs tend to increase in size with age and have more frequent complications.
Although most cases of PAVM are congenital, symptoms often develop between the fourth and sixth decades. The most common presenting symptom is dyspnoea on exertion seen in 31% to 67% of patients,\(^2\) the severity of which is related to the degree of hypoxaemia and magnitude of right to left shunt. However, the majority of patients with PAVMs tolerate hypoxaemia quite well like our patient who only suddenly became very dyspnoeic after the development of the haemothorax. Cyanosis was hidden by anaemia and digital clubbing was absent. Although dyspnoea, cyanosis and clubbing are regarded as the classic triad of PAVM, they were found in only 10% of patients in one study.\(^1\) Murmurs or bruits over the site of PAVMs are heard in 46% of patients,\(^1\) and become more audible during inspiration. In our patient the pulmonary bruit was noticed after tube drainage of the right pleural space.

The most frequently reported emergencies in PAVMs are related to the central nervous system (30% - 50%,\(^1,3\)) and predominant AVM in our patient was in the upper lobe.

Less common but life-threatening complications of PAVMs include haemothorax and haemoptysis.\(^1,2,3,8\) Haemothorax is reported as the rarest of the dangerous complications of PAVM and a third of them occurred during pregnancy.\(^1\) Pulmonary hypertension resulting from increased cardiac output and hyperdynamic state or from pulmonary arterial hypertension (HHT being an independent risk factor) may complicate PAVM by raising the PAVM sac filling pressure thereby increasing the risk of a fatal haemothorax as reported recently.\(^7\)

The classic roentgenographic sign of PAVM is a round or oval sharply-defined mass of uniform density, often lobulated, 1-5 cm in size, \(^{3/4}\) in lower lobes \(^1,2\) with feeding vessels radiating to the hilum. However the shadow may be obscured by haemothorax as seen in the present case (Fig. 2).

Shunt fraction is most accurately assessed by the 100% O\(_2\) method, more than 5% being considered abnormal.\(^1,2,4\) In the ‘room air’ method, PaO\(_2\) <85 mmHg or SaO\(_2\) <96% indicates a potential shunt fraction of >5%,\(^1\) though the method does not differentiate shunt from hypoxaemia due to ventilation perfusion mismatch. In our patient SaO\(_2\) was 79.6% and PaO\(_2\) 39.7 mmHg while breathing room air. However there was no significant orthodeoxia (fall of SaO\(_2\) on assuming upright posture) as the predominant AVM in our patient was in the upper lobe.

Ultra-fast contrast enhanced CT has been shown to be more sensitive than conventional pulmonary angiogram for PAVMs and better in defining their architecture.\(^6\) Pulmonary angiography remains the gold standard especially when a therapeutic intervention is planned.\(^1,8\)

CT pulmonary angiography showed a large AVM in the right upper lobe and a smaller PAVM in the right lower lobe. Approximately 53-75% of PAVMs are found in the lower lobe, 75% of patients have unilateral disease, 36% have multiple lesions and half of those with multiple lesions have bilateral disease.\(^1\)

The current preferred treatment for the majority of patients with PAVMs is percutaneous embolotherapy using coils or balloons and this has largely replaced surgical therapy.\(^1,2,8\)

Since our patient had a significant residual haemothorax with pleural clots and pleural thickening and was haemodynamically unstable, early thoracotomy with evacuation of the pleural space, decortication and surgical resection of the PAVM were decided upon. There was torrential bleed from the abnormal efferent vessels of the large arteriovenous aneurysm, hence the second PAVM in the right lower lobe was not dealt with.

Evaluation of the brother, sister and daughter of the patient clinically, radiologically (CT thorax in case of the sister) and by pulse oximetry was negative for HHT and PAVM although the sister had frequent nose bleeds.

Though uncommon, PAVMs are an important differential diagnosis of common pulmonary problems like haemoptysis, pulmonary nodule and hypoxaemia. A haemorrhagic pleural effusion, with a nodular opacity, commonly raises the clinical spectre of malignancy – however, clues like unexplained and profound hypoxaemia, a pulmonary bruit and a close reading of the chest X ray can lead to the correct diagnosis.

All patients with PAVMs should probably be screened for CAVMs and HHT. Even clinically silent PAVMs and CAVMs may cause considerable morbidity and mortality if kept untreated; so once identified intervention is required in most cases.

The risk of growth of occult lesions requires the patient to be on a regular follow-up.

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References