ECRONICON OPEN ACCESS

EC PULMONOLOGY AND RESPIRATORY MEDICINE

Case Report

Haemothorax and Refractory Hypoxemia: A Rare Presentation of Pulmonary Arteriovenous Malformation

Sushil Upadhay*

Holy Family Hospital, New Delhi, India

***Corresponding Author:** Sushil Upadhay, Holy Family Hospital, New Delhi, India.

Received: September 03, 2019; **Published:** October 17, 2019

Abstract

Pulmonary arteriovenous malformation is a rare vascular anomaly. It bears strong association with hereditary hemorrhagic telangiectasia. The presentation varies from incidental radiological abnormality in asymptomatic subject to life threatening complications in the form exsanguinating hemoptysis or hemothorax. The case discussed is a young male who presented in emergency department with refractory hypoxemia and unilateral hemothorax found to be due to pulmonary AVM who was saved by timely institution of mechanical ventilation followed by angiographic embolization.

Keywords: *Pulmonary Arteriovenous Malformation (PAVM); Hereditary Hemorrhagic Telangiectasia (HHT); Percutaneous Embolization*

Introduction

Pulmonary arteriovenous malformations (PAVMs) are abnormal direct communications between pulmonary arteries and pulmonary veins, resulting in a low resistance right to left shunt [1]. Nearly seventy percent of patients with PAVMs are associated with hereditary hemorrhagic telangiectasia (HHT). Conversely 15 - 35% with HHT have pulmonary AVMs. Dyspnoea, cyanosis and clubbing represent the classical triad of PAVMs. Neurological complications in the form of embolic cerebrovascular accidents, migraine and brain abscess are reported in approximately 50% of patients. Life threatening hemoptysis is rare (11%) and hemothorax is very rare (< 1%) presentation of pulmonary AVM [19].

We describe a case of young male who presented with life threatening hemothorax and refractory hypoxemia due to pulmonary AVM who was successfully treated with timely institution of mechanical ventilation and subsequently angiographic embolization.

Case Report

Twenty eight year old male presented in emergency department (ED) at mid of night with severe breathlessness. Attending friends informed that he visited a hill station for holiday. There, he felt breathless while walking up a slope. He also complained of shooting pain in right side of his chest which subsided after taking an analgesic. Next day he returned to planes by bus. He continued to have mild right side chest discomfort. In the night, he experienced severe pain and breathlessness which was now unbearable and he was brought to ED at odd hours of night. He was conscious, restless, severely breathless and cyanosed. Oxygen saturation (SpO_2) was 55% on ambient air. He was immediately intubated and transferred to intensive care unit for mechanical ventilation. However, even on FiO_2 of 1.0 his spO_2 was

barely touching 90%.ABG was also consistent with severe hypoxemia. Physical examination was marked by cyanosis and clubbing in all the fingers. Breath sounds were absent on right side of chest. Chest X-ray (Figure 1a) showed completely white right hemithorax. Left lung field was apparently clear. Ultrasound of chest revealed large collection in right pleural space. Needle aspiration revealed bloody fluid. Intercostal drain was placed and one and half litre of fluid was drained slowly over half an hour. The fluid was similar to mixed sample of arterial and venous blood. Leading question about any trauma was replied in the negative by the attendants. There was no history of any drug abuse. Repeat chest x-ray (Figure 1b) showed expanded right lung. However, contrary to our expectation the oxygenation did not improve. ABG on Mechanical ventilation with FiO_2 of 1.0 was pH:7.42/$PaCO_2$:42/PO_2:49.7/HCO_3:27.3. It appeared unusual and therefore other possibilities were considered. Closer look at the chest x-ray (Figure 1b) revealed faint round opacity at the base of right lung which was masked by the effusion. Contrast enhanced CT scan of thorax (Figure 2a and 2b) revealed vascular mass suggestive of pulmonary arteriovenous malformation. Patient was hemodynamically stable. Bed side echocardiogram revealed normal cardiac function. The laboratory findings showed haemoglobin of 14 gm%. Liver and renal biochemistries, urine examination, platelet counts and coagulation parameters were normal. Pleural fluid haematocrit was 24% consistent with hemothorax.

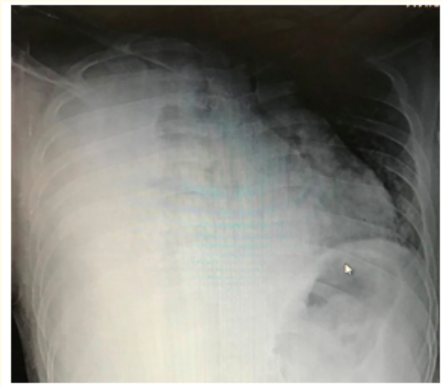

Figure 1a: *Chest X-ray (at presentation).*

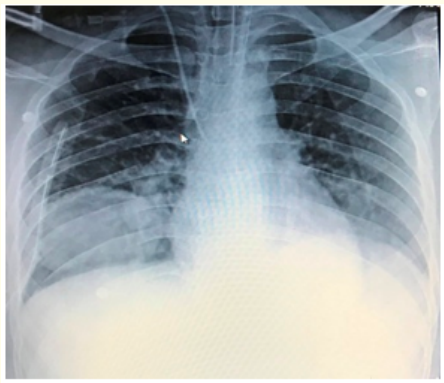

Figure 1b: *Chest X-ray (Post ICD).*

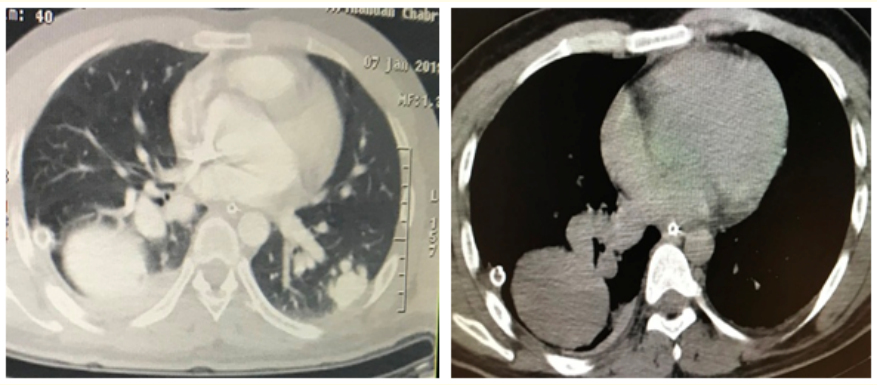

Figure 2a and 2b: *Contrast enhanced CT scan of thorax.*

Multidetector computed tomography (MDCT) thorax angiography was performed for confirmation. A lobulated vascular mass of 30 x 15 mm in right anterior lower chest with single feeder vessel of diameter 5.5 mm was noted. Efferent was seen draining into inferior pulmonary vein. There were smaller vascular masses on left side. Interventional radiologist was called for and embolization was planned. Patient was transferred to cath lab and percutaneous angiography was performed. The malformations were embolized with metallic coils (Figure 3). Post embolization oxygenation improved and the patient could be extubated next day. ABG on 6LPM oxygen by mask pH:7.41/$PaCO_2$:40/PO_2:63/HCO_3:25. ABG on Ambient air was pH:7.45/$PaCO_2$:34/PO_2:55.5/HCO_3:23. History was taken from the patient himself after extubation. He revealed that there were infrequent incidence of epistaxis during his childhood. He also informed that his mother and grand mother suffered frequent minor nose bleeds but did never seek any treatment. CT brain was done and it ruled out any evidence for cerebral AVMs. He was discharged to home and was followed up on out patient basis. He resumed his daily activities without any discomfort. Chest X-ray (Figure 4) done at 6month follow up showed marked reduction in size of AVMs with coils *in situ*.

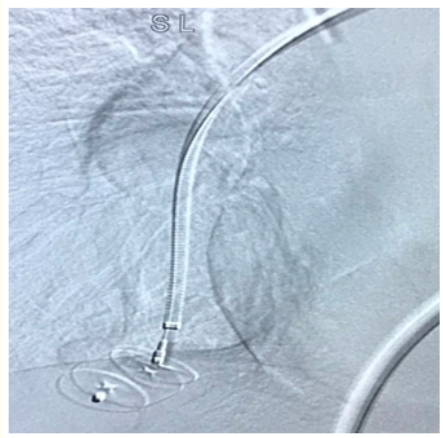

Figure 3: *Coil embolization.*

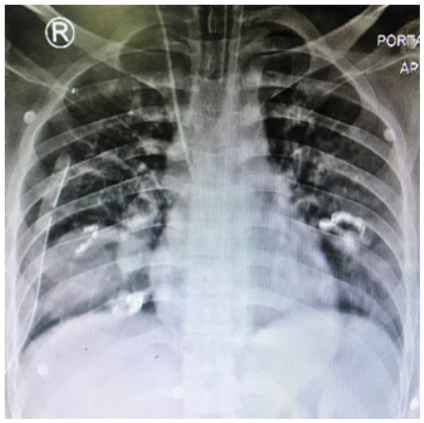

Figure 4a: *Chest X-ray (post embolization).*

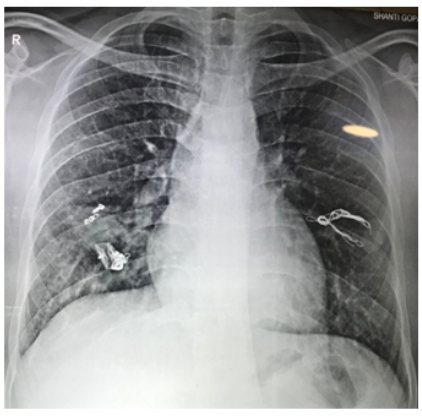

Figure 4b: *Follow up Chest X-ray.*

Discussion and Conclusion

Direct communications between the branches of pulmonary artery and pulmonary veins, without an intervening capillary bed, have been variously called as pulmonary arteriovenous fistula, pulmonary arteriovenous malformation (PAVM), pulmonary arteriovenous aneurysm (PAVA), pulmonary angioma, arteriovenous angiomatosis, cavernous haemangiomas, and pulmonary hamartomas [1,2]. PAVM is the most preferred term as it represents the developmental anomaly.

More than 80% of PAVMs are congenital, and of these 47% - 80% are associated with Osler-Weber-Render disease or hereditary haemorrhagic telangiectasia (HHT) [3]. Exact etiology is unknown but mutation of endoglin gene can cause vascular dysplasia and is seen more often in patients with genetic linkage to chromosome 9q3 [5].

PAVMs are rare vascular anomalies. The incidence of PAVM is 2 - 3 per 100,000 population [6]. Male to female ratio varies from 1:1.5 to 1.8, in several series [4].

The age at the first presentation ranges from newborn to 70 but the majority of cases are diagnosed in the first three decades of life [3].

Asymptomatic patients are common and account for 13% to 55% of patients in different series [7]. The most common presentation is epistaxis followed by dyspnea. Hemoptysis being third most commonly reported symptom. Dyspnoea on exertion is reported by 31% to 67% of patients. The majority of the patients with PAVMs tolerate hypoxaemia very well and are relatively or completely asymptomatic unless the arterial oxygen pressure is less than 8.0 kPa (60 mm Hg) [3,4]. The classic triad of dyspnoea, cyanosis, and clubbing is found in only 10% of patients with a PAVM [8]. Our patient did not have any history prior to this presentation except for occasional nose bleeds in winter months that too in early age.

Meticulous physical examination may detect abnormal signs in 75% of patients. The most common physical findings are cyanosis, clubbing, and pulmonary vascular bruit. Mucocutaneous telangiectasias have been reported in up to two thirds of HHT patients with a PAVM [4]. The reported subject had cyanosis and clubbing of the fingers with no other signs suggestive of HHT.

Massive haemoptysis after intrabronchial rupture of a lesion or haemothorax after rupture of a subpleural lesion is rare but potentially a fatal complication of PAVM [7,9]. Our patient presented with severe hypoxemic respiratory failure requiring immediate intubation and mechanical ventilation. Such a presentation is rare to encounter in literature.

In 1917, Wilkins described the necropsy findings in a 23 year old women with cyanosis, clubbing, telangiectasia, and bilateral axillary bruits who died from haemothorax after rupture of a PAVA into the pleural cavity [10]. The presence of symptoms usually correlates with the size of lesion, a single PAVM less than 2 cm in diameter on chest radiography usually does not cause symptoms [3].

The most commonly reported complications relate to the central nervous system. Incidence of neurological events has been reported as: migraine 43%, transient ischaemic attack 37%, stroke 18%, brain abscess 9%, and seizure 8%. The most likely mechanism for these neurological events is paradoxical embolism across the PAVM or across a coexisting cerebral arteriovenous malformation in patients with HHT [18].

Chest radiography is an important diagnostic tool not only in diagnosis but also in the follow up of patients with a PAVM. A plain chest radiograph shows abnormalities in about 98% of patients. The classic radiographic features of PAVM is a round or oval sharply defined mass of uniform density, frequently lobulated, and ranging in size from 1 - 5 cm in diameter.; two thirds are located in the lower lobes and in nearly 30% they are bilateral [11].

The shunt fraction (fraction of cardiac output that shunts from right-to-left through a PAVM) is raised in 88% to 100% of patients with a PAVM. Contrast echocardiography is almost 100% sensitive in detecting intrapulmonary shunts in PAVM and also helps to monitor the therapeutic efficacy following embolization [11,14]. Shunt fraction is most accurately measured by the 100% oxygen method [12]. Radionuclide perfusion lung scan is a useful adjunct in the diagnosis and quantification of PAVM. Although, a positive result is not specific for PAVM, a negative result essentially excludes the diagnosis [13]. Ultra-fast contrast enhanced CT has been shown to be more sensitive than conventional pulmonary angiogram for PAVMs and better in defining their architecture [11]. Pulmonary angiography remains the gold standard in the diagnosis of PAVM and is justified to confirm the diagnosis in virtually all cases. A pulmonary angiogram not only identifies the PAVM but also further defines the angioarchitecture of pulmonary vasculature, which is necessary before therapeutic embolisation or urgical resection [1,3,4,11].

There is evidence that PAVMs progressively enlarge over a period of time and incidence of progression is higher in patients with untreated PAVM. Treatment should be offered to all symptomatic patients and also to asymptomatic patients with lesions less than 2 cm in

diameter on chest radiography. The purpose of treatment includes prevention of neurological complications, progressive hypoxia and its resultant effects, and high output cardiac failure [3].

Since the first successful resection of PAVM in 1942, surgery was the only treatment available until 1978, when Taylor., *et al.* reported the first successful percutaneous embolization [15,16]. The current preferred treatment for the majority of patients with a PAVM is percutaneous embolotherapy using coils or balloons [17] and it has practically replaced surgical intervention.

The overall success rate of coil embolization was reported as 99% in one series. The procedure is safe with minimal morbidity and no mortality. Most common complication being self limited pleuritic chest pain reported by 13% of patients which may be more (i.e. 31%) in those with larger AVMs (feeding vessel > 8 mm) [14,18].

We chose percutaneous embolization as treatment option for this patient as he was extremely sick requiring mechanical ventilation with 100% FiO_2. Moreover he had bilateral PAVMs where surgical treatment was not feasible.

This case is presented in view of its rarity with rare complication and successful management.

Bibliography

1. Stringer CJ., *et al*. "Pulmonary arteriovenous fistulas". *American Journal of Surgery* 89.5 (1955): 1054-1080.
2. Le Roux BT. "Pulmonary hamartomas". *Thorax* 19 (1964): 236-243.
3. Dines DE., *et al*. "Pulmonary arteriovenous fistula". *Mayo Clinic Proceedings* 58 (1983): 176-181.
4. Sluiter-Eringa H., *et al*. "Pulmonary arteriovenous fistula: diagnosis and prognosis in non-complaint patients". *American Review of Respiratory Disease* 100.2 (1969): 177-184.
5. Porteous MEM., *et al*. "Genetic heterogeneity in hereditary haemorrhagic telangiectasia". *Journal of Medical Genetics* 31.12 (1994): 925-926.
6. Hodgson CH and Kaye RL. "Pulmonary arteriovenous fistula and hereditary telangiectasia". *Diseases of the Chest* 43 (1963): 449-455.
7. Esplin MS and Varner MW. "Progression of pulmonary arteriovenous malformation during pregnancy". *Obstetrical and Gynecological Survey* 52.4 (1997): 248-253.
8. Puskas JD., *et al*. "Pulmonary arteriovenous malformations: therapeutic options". *Annals of Thoracic Surgery* 56.2 (1993): 253-258.
9. Ference BA., *et al*. "Life-threatening pulmonary hemorrhage with pulmonary arteriovenous malformations and hereditary hemorrhagic telangiectasia". *Chest* 106.5 (1994): 1387-1390.
10. Wilkins GD. "Ein Fall von multiplen pulmonalis anerysmen". *Beitr Klinis Tuberk* 38 (1917): 1-10.
11. Prager RL., *et al*. "Arteriovenous fistula of the lung". *Annals of Thoracic Surgery* 36.2 (1983): 231-239.
12. Haitjema TJ., *et al*. "Embolization of pulmonary arteriovenous malformations: results and follow up in 32 patients". *Thorax* 50.7 (1995): 719-723.
13. Whyte MKB., *et al*. "Quantification of right-to-left shunt at rest and during exercise in patients with pulmonary arteriovenous malformations". *Thorax* 47.10 (1992): 790-796.

14. Remy J., *et al*. "Angioarchitecture of pulmonary arteriovenous malformations: clinical utilities of three-dimensional helical CT". *Radiology* 191.3 (1994): 657-664.

15. Taylor BG., *et al*. "Therapeutic embolization of the pulmonary artery in pulmonary arteriovenous fistula". *American Journal of Medicine* 64.2 (1978): 360-365.

16. Hepburn J and Dauphinee JA. "Successful removal of hemangioma of lung followed by disapperance of polycythemia". *American Journal of the Medical Sciences* 204 (1942): 681-687.

17. Dutton JAE., *et al*. "Pulmonary arteriovenous malformations: results of treatment with coil embolization in 53 patients". *American Journal of Roentgenology* 165.5 (1995): 1119-1125.

18. White RI Jr., *et al*. "Pulmonary arteriovenous malformations: techniques and long-term outcome of embolotherapy". *Radiology* 169.3 (1988): 663-669.

19. Khurshid I and Downie GH. "Pulmonary arteriovenous malformation". *Postgraduate Medical Journal* 78.918 (2002): 191-197.

Volume 8 Issue 11 November 2019