International Journal of Cardiovascular and Thoracic Surgery
2015; 1(1): 1-4
Published online June 30, 2015 (http://www.sciencepublishinggroup.com/j/ijcts)
doi: 10.11648/j.ijcts.20150101.11

Pulmonary Arterio-Venous Malformations: Is it Easy to Diagnose and Treat Early

Waheed Etman[1], Walid Abu Arab[1, 2]

[1]Cardiothoracic Surgery Department, University of Alexandria, Alexandria, Egypt
[2]Service of Cardiothoracic Surgery, University of Sherbrooke, Quebec, Canada

Email address:
watman@yahoo.com (W. Etman), walidabuarab@yahoo.com (W. Abu Arab)

To cite this article:
Waheed Etman, Walid Abu Arab. Pulmonary Arterio-Venous Malformations: Is it Easy to Diagnose and Treat Early. *International Journal of Cardiovascular and Thoracic Surgery*. Vol. 1, No. 1, 2015, pp. 1-4. doi: 10.11648/j.ijcts.20150101.11

Abstract: *Background:* Pulmonary arterio-venous malformation (PAVM) is a rare vascular anomaly that can be confronted with in cardiothoracic surgery patients. This study is a retrospective one that analyses the data of our experience with this entity at cardiothoracic surgery department of Alexandria University during nine years. *Patients and methods:* This is a retrospective study. The files of patients who presented with PAVMs were studied with special attention to the symptoms, signs, investigations that were performed, management and outcome. *Results:* Eleven patients were included in this study (seven males and four females). They were presented with various symptoms and were investigated mostly by plain x-ray chest and CT-Chest. CT scan with contrast or recently multi-slice CT was a good investigative tool that we depended on it without need for other diagnostic tool. The management was in the form of surgical resection in seven patients and embolo-therapy in four patients. Follow-up with plain x-ray chest and CT-Chest was completed for seven patients where there was no recurrence in those patients underwent embolo-therapy and no new development of PAVM in those underwent surgical resection *Conclusion:* We concluded that PAVM is an easy entity to diagnose, treat and manage but needs high clinical suspicion.

Keywords: Pulmonary Arterio-Venous Fistula, Vascular Malformation, PAVMs

1. Introduction

Pulmonary arterio-venous malformation (PAVM) is a rare entity of pulmonary vascular anomalies. It includes a direct communication between the branches of pulmonary artery and that of the pulmonary vein (1-4). In some occasions, the arterial supply is derived from systemic circulation like aorta, intercostal artery, or bronchial artery. Even more, the venous drainage may be to the systemic circulation in some cases (5, 6). PAVM is mostly difficult to diagnose as many patients are asymptomatic. Patients are usually presented late with complications. In 1897; Churton described the first case of PAVM (3). PAVM can be either congenital (80%), or acquired (20%). Congenital cases of PAVMs are associated with Osler-Weber-Render disease (7) or Hereditary Hemorrhagic Telangectasia (HHT) (8, 9).

Recently, many studies attributed PAVM to gene alteration (10, 11).The gene alteration linkage was located on chromosome 9 (9q33-34 or OWR-1) in some families and chromosome 12 (12q or OWR-2) in others (10, 11). While acquired PAVM was attributed to thoracic surgery, long standing hepatic cirrhosis, infections, mitral stenosis and pregnancy (6, 11-14). The incidence of PAVM was reported to be 2-3/100 000 of population (15). Male to female ratio was found to be 1:1.8 (3). Most of PAVMs were located in the lower lobes of the lungs (5, 16).

The clinical presentation is variable. Patients are usually presented late as most of them are asymptomatic (14). Majority of patients are presenting with variable degrees of dyspnea which is usually attributed to any other causes (3, 7). Other presenting symptoms are epistaxis, melena, or neurological symptoms (7, 16). Physical findings are also not so prominent in early course of the disease and usually evident in complicated disease or during the late course of the disease including cyanosis, clubbing, and pulmonary vascular bruit (3, 17).

2. Patients and Methods

This is a retrospective study that was performed at Cardiothoracic Surgery Department, Faculty of Medicine of Alexandria. Analysis of the PAVM patients' files during a previous nine years was done. Parameters of the study included age, gender, symptoms and signs, methods of

investigations, ways of treatment and follow-up.

3. Results

Study included eleven patients. There were seven males (63.6%) and four females (36.4%). All patients included in the study were adults. Age ranged from 32- 55 years old (37.7± 8.5 years). Eight patients (73%) were symptomatic. PAVM was detected accidently in the other three patients at chest x-ray that was performed for anther reasons (Table 1).

There was a family history of Hereditary Hemorrhagic Telangectasis in 20% of patients; while it was irrelevant in the remaining patients (80%). Clinical signs elicited were summarized in Table 2. The most frequent detected sign was cyanosis and clubbing *(n = 4)* followed by *anemia (n = 3)* and polycythemia *(n = 2)*. One female patient presented with embolic cerebral embolisation.

Table 1. *Clinical symptoms among patients with PAVM.*

Symptom	Patient numbers (n)
Asymptomatic	3
Chest pain	3
Dyspnea & Cyanosis	2
Haemoptysis	2
Embolic manifestation	1

Table 2. *Clinical signs detected in patients with PAVM.*

Clinical symptoms	Patient numbers (n)
Clubbing	3
Cyanosis & Hypoxia	3
Polycythemia	2
Anemia	2
Transient Ischemic Attacks (TIA)	1

Diagnostic work-up started with plain chest x-ray (postero-anterior and lateral views). It revealed sharply defined mass either oval or rounded. The diameter of the mass ranged from 2-7 cm (3.7±1.6 cm). Sixty four percent of these lesions (64%) were located at the left lower lobe and the remaining (46%) were located at the right lower lobe (Fig.1). In patients who presented with cyanosis and clubbing; arterial blood gas analysis demonstrated PO_2 around 40 mmHg (40.8 ± 0.9).

The second tool for diagnosis was the CT- chest with contrast and recently multi-slice CT with three dimensional image reconstruction (Fig. 2). This investigative tool was able successfully to confirm the diagnosis of the PAVM and to delineate its size and location with accuracy. In the first five patients, diagnosis was established by CT chest and confirmed with pulmonary angiography. Later on, diagnosis was established on CT-chest only.

The management was in form of surgery in seven patients because embolo-therapy was not possible due to very large vessels. Embolo-therapy was used in four patients.

During surgery, mobilization of the peripheral adhesions was not performed except after controlling proximal vessels and securing the main pulmonary artery. This was to secure control in emergency during dissection for resection and to decrease the incidence of intra-operative bleeding and consequently avoiding post-operative complications.

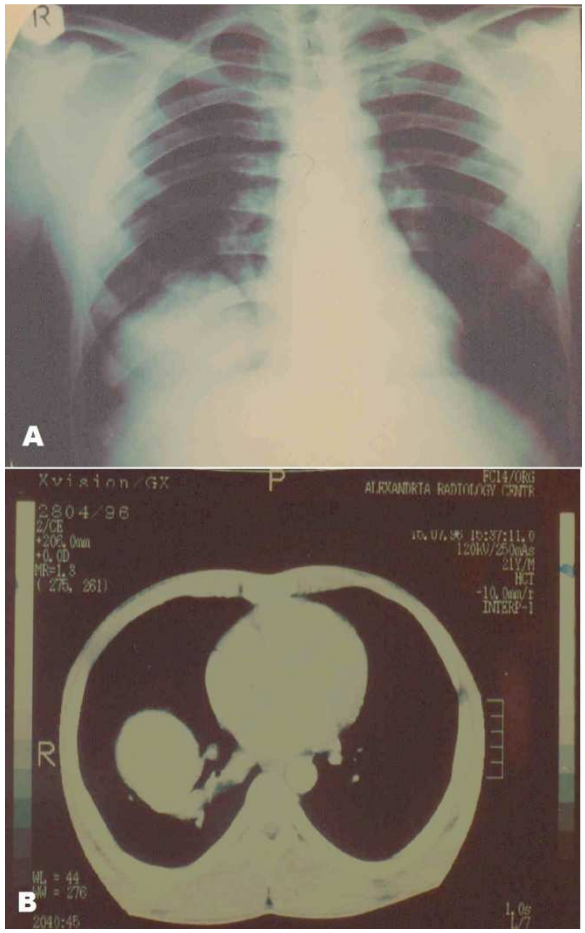

Fig. 1. *Fig. (1-A): Chest x-ray chest (P-A view) demonstrates a large mass located at the right lower lobe. Fig. (1-B): CT chest with contrast revealed vascular mass located at the right lower lobe with a feeding vessel originating from the right pulmonary artery.*

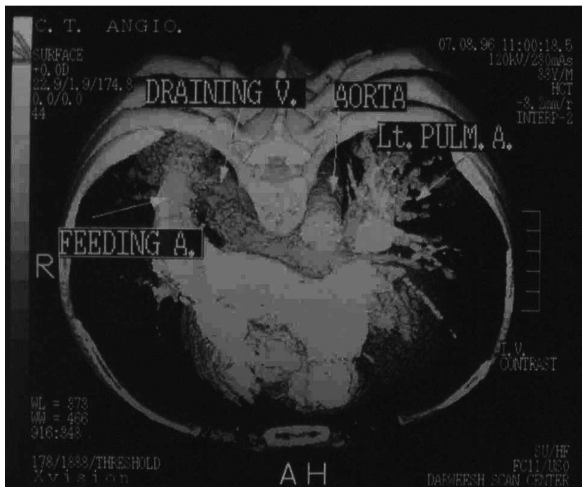

Fig. 2. *CT chest angiography shows the feeding and draining vessel at right lower lobe.*

Some complications were encountered. Post-operative lung collapse was detected (*n = 1*). It was managed by bronchoscopy and aspiration of sticky mucous plug. One patient required exploration for postoperative bleeding. Exploration revealed massive bleeding from previously dissected adhesions to chest wall. One patient died in the

immediate postoperative period due to persistent hypoxemia. Late complication in the form of spontaneous pneumothorax was detected in one patient one month later after surgery. It was managed with intercostal tube thoracostomy for three days. Recurrent attacks of mild haemoptysis are another late complication that was encountered in another patient who underwent lobectomy. Bronchoscopy revealed granuloma on suture line.

Follow-up CT was completed in seven patients (three of them underwent surgery and the other four patients underwent embolotherapy); no recurrence for PAVM could be detected in three of those who underwent embolotherapy. In one patient who complained of recurrent haemoptysis CT revealed recurrence of the PAVM. Patient underwent right lower lobectomy. In the resection group those who were followed with CT-chest; no obvious abnormalities could be detected.

4. Discussion

The first description of PAVM was done by Churton at autopsy in 1897 (3). Since that time; various terms have been used to describe these abnormal communications including pulmonary arteriovenous fistulas, pulmonary arteriovenous aneurysms, haemangiomas of the lung, cavernous angiomas of the lung, pulmonary telangiectases, and pulmonary arteriovenous malformations (5). PAVMs are caused by abnormal connections between pulmonary arteries and pulmonary veins through a thin-walled aneurysm, which are most commonly congenital in nature; although an acquired condition resulting from penetrating trauma has been described. They act as direct right-to-left shunts, resulting in dyspnea, fatigue, cyanosis, and polycythemia when the shunt is large. Moreover, the PAVM bypasses the capillary bed the lung loses its filter function thus allowing emboli and bacteria to pass directly into the systemic circulation, resulting in stroke or cerebral abscess (11).

This study stresses on the importance of clinical suspicion in diagnosis of this uncommon clinical problem (PAVM). Clinical presentation including dyspnea, cyanosis, hemoptysis, chest pain and clubbing should raise the suspicion of PAVM. A definitive diagnosis of PAVM was made conventionally by chest x-ray and angiography or multi-slice CT-angiography. CT- angiography remains the gold standard allowing; not only the location of the pathology but also; mapping of the feeding vessels before surgical treatment or embolotherapy. The goals of treatment are to improve symptoms of dyspnea/hypoxemia; to prevent lung hemorrhage; and most important to prevent neurologic complications.

One of the most vexing aspects of pulmonary AVMs is that there are few reliable treatment options. The most common indication for treatment of AVMs is cyanosis. However, it has been proposed that the risk of systemic embolisation justifies the treatment of AVMs even in asymptomatic cases if the diameter of the feeding vessel(s) is more than 3 mm (17). Shunt-induced hypoxemia is surgically correctable by ligation of the fistulas or lung resection (14). When operation is contraindicated, an alternative therapy may be balloon or coil embolisation of the PAVMs (18).

Studies have shown that CT with contrast was able to reach diagnosis easily without need to further investigations like pulmonary angiography (19) or shunt measurement (3, 18, 19). Moreover, meticulous history taking and clinical suspicion are very important in diagnosis as in 20% of patients there was a family history of Hereditary Hemorrhagic Telangectasis (HHT). In some instances, the presenting symptoms were only pleuritic pain (27%) which may be taken as pleurisy or myositis. Radiologists should be very careful in deciding to take a needle biopsy from lung nodule as this nodule may be PAVM and can cause catastrophic intra-thoracic bleeding. Management is either with surgical resection or embolotherapy or both. Any intervention is easy once you are aware of possible complications and trying to avoid them (14).

5. Conclusion

Finally; we concluded that early diagnosis of PAVM is not easy as most patients are presented late, or are presented with complications. However, it is easy to manage and to treat once diagnosis is established. Follow up is mandatory to detect recurrence or new development of new PAVM.

References

[1] Altin S, Dalar L, Karasulu L, Çetinjaya E, Timur S, Solmazer N. Resection of giant endobronchial hamartoma by electrocautery and cryotherapy via flexible bronchoscopy. Tüberküloz ve Toraks Dergisi. 2007; 55(4):390-4.

[2] Anabtawi IN, Ellison RG, Ellison LT. pulmonary arteriovenous aneurysms and fistulas. anatomical variations, embriology, and classification. The Annals of thoracic surgery. 1965;122:277-85.

[3] Khurshid I, Downie GH. Pulmonary arteriovenous malformation. Postgraduate medical journal. 2002;78(918):191-7.

[4] Remy J, Remy-Jardin M, Wattinne L, Deffontaines C. Pulmonary arteriovenous malformations: evaluation with CT of the chest before and after treatment. Radiology. 1992 March 1, 1992;182(3):809-16.

[5] Bosher Jr LH, Blake DA, Byrd BR. An analysis of the pathologic anatomy of pulmonary arteriovenous aneurysms with particular reference to the applicability of local excision. Surgery. 1959;45(1):91.

[6] Prager RL, Laws KH, Bender HW. Arteriovenous fistula of the lung. The Annals of thoracic surgery. 1983;36(2):231-9.

[7] Mansour KA, Hatcher Jr CR, Logan Jr WD, Abbott OA. Pulmonary arteriovenous fistula. The American surgeon. 1971;37(4):203.

[8] Gossage JR, Kanj G. Pulmonary arteriovenous malformations a state of the art review. American journal of respiratory and critical care medicine. 1998;158(2):643-61.

[9] Shovlin CL, Letarte M. Hereditary haemorrhagic telangiectasia and pulmonary arteriovenous malformations: issues in clinical management and review of pathogenic mechanisms. Thorax. 1999;54(8):714-29.

[10] Pagon RA, Bird TD, Dolan CR, Stephens K, McDonald J, Pyeritz RE. Hereditary Hemorrhagic Telangiectasia. 2000.

[11] Guttmacher AE, Marchuk DA, White Jr RI. Hereditary hemorrhagic telangiectasia. New England Journal of Medicine. 1995;333(14):918-24.

[12] Symbas PN, Goldman M, Erbesfeld MH, Vlasis SE. Pulmonary arteriovenous fistula, pulmonary artery aneurysm, and other vascular changes of the lung from penetrating trauma. Annals of surgery. 1980;191(3):336.

[13] Kamei K, Kusumoto K, Suzuki T. Pulmonary amyloidosis with pulmonary arteriovenous fistula. CHEST Journal. 1989;96(6):1435-6.

[14] Puskas JD, Allen MS, Moncure AC, Wain JC, Hilgenberg AD, Wright C, et al. Pulmonary arteriovenous malformations: therapeutic options. The Annals of thoracic surgery. 1993;56(2):253-8.

[15] Hodgson CH, Kaye RL. Pulmonary arteriovenous fistula and hereditary hemorrhagic telangiectasia: a review and report of 35 cases of fistula. CHEST Journal. 1963;43(5):449-55.

[16] Roman G, Fisher M, Perl DP, Poser CM. Neurological manifestations of hereditary hemorrhagic telangiectasia (rendu-osler-weber disease): Report of 2 cases and review of the literature. Annals of neurology. 2004; 4(2):130-44.

[17] Burke CM, Safai C, Nelson DP, Raffin TA. Pulmonary arteriovenous malformations: a critical update. The American review of respiratory disease. 1986;134(2):334.

[18] Dutton JA, Jackson JE, Hughes JM, Whyte MK, Peters AM, Ussov W, et al. Pulmonary arteriovenous malformations: results of treatment with coil embolization in 53 patients. American Journal of Roentgenology. 1995; 165(5):1119-25.

[19] Grahmann PR, Bonnet R, Swoboda L, Nöldge G, Hasse J. Pulmonary Arteriovenous Malformations Case Reports and Literature Review. Angiology. 1994; 45(5):399-404.