

Pathology - Case Report

A Case of Right Middle Lobe Pulmonary Arteriovenous Malformation Presenting as Non-Resolving Pneumonia

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Background --- Pulmonary arteriovenous malformations (PAVMs) are lesions in the vasculature that allow blood to bypass the capillary system, flowing from arteries directly into veins.

Case --- Patient is a 50 years old male, with recurrent episodes of hemoptysis was initially treated as a case of pneumonia and PTB. CT scan with contrast and CT angiogram revealed AV malformation.

Histopathologic Findings --- The specimen showed lung tissue exhibiting areas of varisized thin and partially thick vascular channels lined by endothelial cells surrounded by extensive intraalveolar hemorrhage and focal areas of atelectasis. It was seen in close proximity to a bronchus and accompanying medium-sized arteries, predominantly extending to the pleural lining.

Discussion --- The definitive diagnosis is established by means of direct imaging study such as a CT scan with contrast, pulmonary angiogram and currently by CT angiogram. Our case presented with pneumonia, later developed hemoptysis and was confirmed with PAVM by CT scan and CT angiogram, and underwent right middle lobectomy. Other treatment options are embolization and lung transplantation.

Conclusion --- The rarity of cases and the variability of presentation posed a great challenge. Its rarity should alert clinicians to include PAVM's in the differential diagnosis of patients with pneumonia and hemoptysis. *Phil Heart Center J 2012;16:58-62.*

Key Words: Pulmonary Arteriovenous Malformation

Pulmonary arteriovenous malformation (PAVM) is a rare clinical entity wherein there is an abnormal communications between the pulmonary artery and pulmonary veins. The etiology of this disease is usually congenital; however, they may be acquired in certain conditions, such as mitral stenosis, schistosomiasis, tuberculosis, trauma and metastatic thyroid carcinoma among others. Some patients have the hereditary hemorrhagic telangiectasia (HHT) also known as Rendu-Osler-Weber syndrome.¹

Congenital cases are caused by failure of differentiation of the embryonic vascular plexus. These vessels expose the low resistance venous system to systemic pressures leading to abnormal vascular formations. Such lesions may be solitary and discrete or generally diffuse. Based on their architecture, they can be classified as simple PAVM in 80% of cases, wherein there is a single feeding segmental artery leading to a single draining pulmonary vein, which usually occurs

in lower lobes; and 20% are complex PAVMs; wherein there are often 2 or more feeding arteries or draining veins and often occur in the distributions of the lingual, right lower lobe and the right middle lobe.²⁻³

Milder forms are usually asymptomatic, whereas, those with severe disease have respiratory distress and hemoptysis. The presence of right to left shunt can be complicated by cyanosis, brain abscess and polycythemia.⁴

Pulmonary angiogram, which is the gold standard in the diagnosis of PAVM, usually shows the abnormal peripheral vascular formations, which may either be localized or diffused. Contrast echocardiography has been proven useful and accurate in screening for PAVM.⁵ Likewise multi-slice computed tomography is also useful.⁶

The reported incidence is 3.2 -4.5 cases per year.⁷ In our institution, this is the 6th diagnosed case since 1976 to the present time.

CASE SUMMARY

A 50 year old male, asthmatic, without any significant past or family history, consulted for hemoptysis. Seven months prior to admission, the patient suddenly developed cough with whitish phlegm, accompanied by undocumented low grade fever and was treated as a case of pneumonia.

Six months prior to admission, he had blood streaked sputum but with no fever nor weight loss noted. Chest film showed an inflammatory process on the right mid lung field. Plain Chest CT scan showed inflammatory process with necrosis, and fibrosis in the lingular division of the upper lobe. He was treated as a case of PTB with quadruple anti-Koch's medications.

Three months prior to admission, he developed massive hemoptysis. Chest film revealed pneumonia, right middle lobe with beginning consolidation, resolving. Chest CT Scan result revealed an inflammatory process, probably chronic. Echocardiography showed normal left ventricular dimension with normal wall motion contractility and systolic function but with Doppler evidence of impaired relaxation, dilated aortic root with no evidence of dissection, normal pulmonary artery pressure with pulmonic regurgitation. Sputum AFB smears and blood culture were negative.

One month prior to admission, due to persistence of hemoptysis, a CT angiogram and a repeat CT scan with contrast (*Figure 1a and 1b*) revealed a focal consolidation with small necrosis in the lateral segment of the right middle lobe. Impression was an arterial fistula vs. arteriovenous malformation. An incidental finding of a benign granuloma was also seen. Patient was advised surgery.

On admission, the patient was apparently well. The rest of the PE findings were also unremarkable. On the 3rd hospital day, bronchoscopy showed no intraluminal masses with bronchial walls noted to have mucosal edema. Carina was sharp and left main bronchus revealed pits and depression. The patient then underwent elective thoracotomy and right middle lobe lobectomy on 4th hospital day. The operation was unremarkable and on the tenth hospital day patient was discharged improved.

HISTOPATHOLOGY

The specimen submitted for histopathologic examination consisted of a right middle lobectomy specimen (8.5 x 6.0 x 4.0 cm) with bronchial remnant (1 cm in length, 1 cm in diameter). The entire specimen weighed 60 g. The pleura is smooth with an ill-defined thickening (2 x 2 x 1.5 cm) on the superior portion of the middle lobe (2.5 cm from the bronchial line of resection). A soft area measuring 3 x 2.5 x 1.5 cm is noted on the anterior, middle portion of the middle lobe (2.5 cm from the bronchial line of resection). Cut sections showed tan-red to tan-brown spongy cut-surfaces with a pale firm area measuring 2 cm corresponding to the thickened pleural surface.

Microscopic sections showed lung tissue exhibiting areas of varisized thin and partially thick vascular channels lined by endothelial cells and filled with red blood cells surrounded by extensive intraalveolar hemorrhages and focal areas of atelectasis. This area is in close proximity to the bronchus and accompanying medium-sized arteries, predominantly closer to the thick pleural lining. Elastic stain (von Gieson) highlights the elastic lamina of the thin-walled and thick-walled vessels. Moderate plasmalymphocytic and few neutrophilic infiltrates are noted within the peribronchial and some alveolar spaces indicative of bronchopneumonia. No fibro-calcific nodules nor evidence of chronic granulomatous disease identified.

DISCUSSION

Although PAVM is considered as an unusual lesion, cases have been reported as early as 1897. Single and isolated cases are not as uncommon as small diffuse and bilateral cases, which usually presents at an earlier age.⁹

PAVMs occur more frequently in women and are transmitted as a dominant gene with incomplete penetrance.¹⁰ Around 10% of the cases are identified in infancy or childhood, followed by a gradual increase in the incidence through the fifth and sixth decades. More than 80% of PAVM's are congenital, the remaining being acquired. The acquired PAVM are very rare and are caused by chest trauma, surgery cirrhosis of liver, metastatic carcinoma and infections (actinomycosis,

mucormycosis).¹¹ Seventy percent of PAVM are associated with Osler-Weber-Rendu syndrome or hereditary hemorrhagic telangiectasia (HHT) and conversely 30% of patients with HHT have PAVM.¹¹

Lesions less than 2 cm are usually asymptomatic.⁷ PAVM usually occur in the lower lobe in 53%-70% of the cases.¹² The diagnosis of PAVM should be suspected in patients with any of the following presentations:⁹ (1) presence of pulmo-

nary nodules with typical radiographic findings; (2) mucocutaneous telangiectasias; and (3) unexplained findings such as dyspnea, hemoptysis, hypoxemia, polycythemia, clubbing, cyanosis, cerebral embolism, or brain abscess. Majority presents with symptoms when they are between the fourth to sixth decades of life, although some patients present in early life with severe cyanosis, congestive heart failure or respiratory failure.⁹

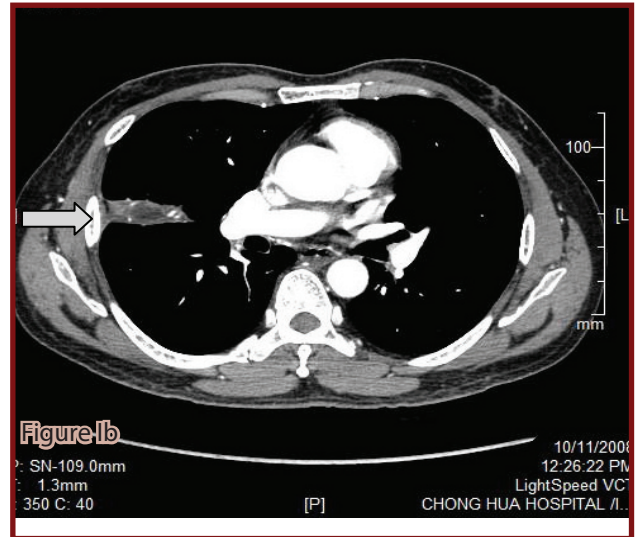
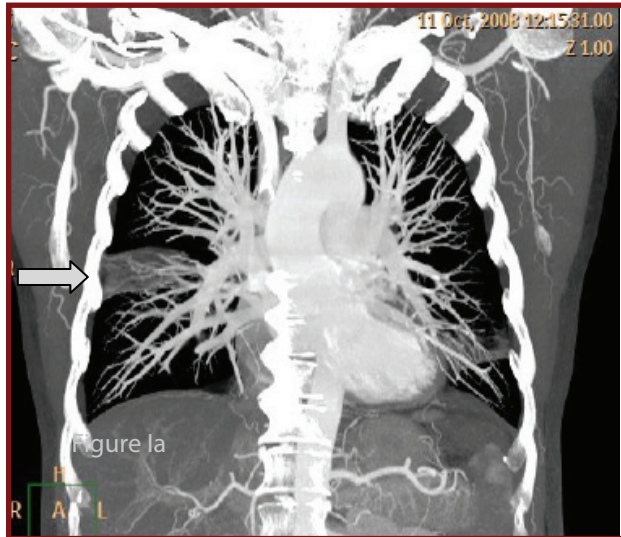


Figure 1: Chest CT Scan of a 50 year old male with recurrent hemoptysis.

Figure 1a: CT scan Coronal - Maximum Intensity Projection view with contrast shows area with wedge shape (grey arrow), intensely enhancing lesion in the right middle lung field with feeding vessel suggestive of AV malformation.

Figure 1b: CT scan Axial - Maximum Intensity Projection view with contrast shows area with wedge shape (grey arrow), intensely enhancing lesion in the right middle lung field with feeding vessel suggestive of AV malformation.

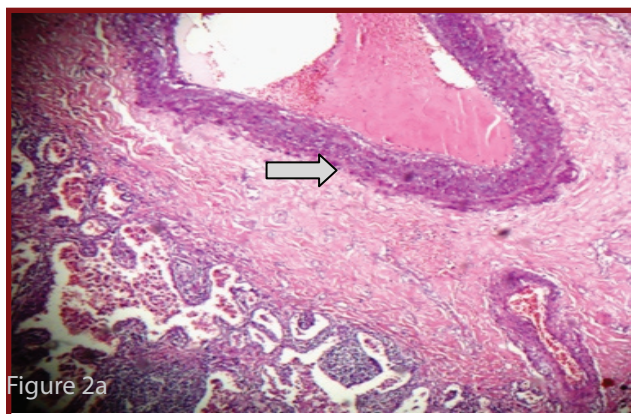


Figure 2: Histopathology of right middle lobectomy specimen of a 50 year old male with recurrent hemoptysis.

Figure 2a: Hematoxylin-eosin section showing an area with thick and thin walled vascular channel adjacent to each other (grey arrow).

Figure 2b: Elastic stain (von Geison) highlights the elastic lamina of the vascular channel (grey arrow).

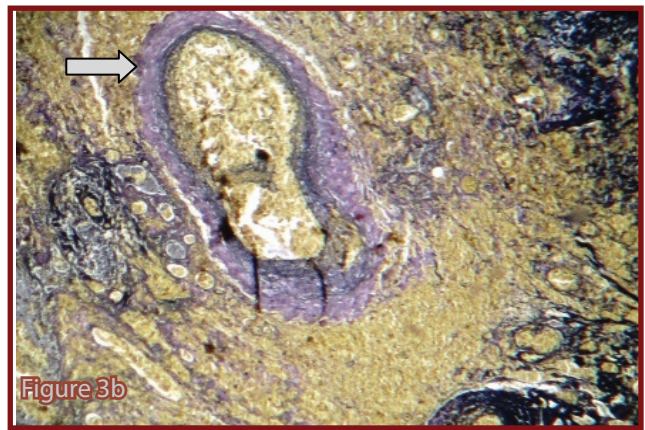
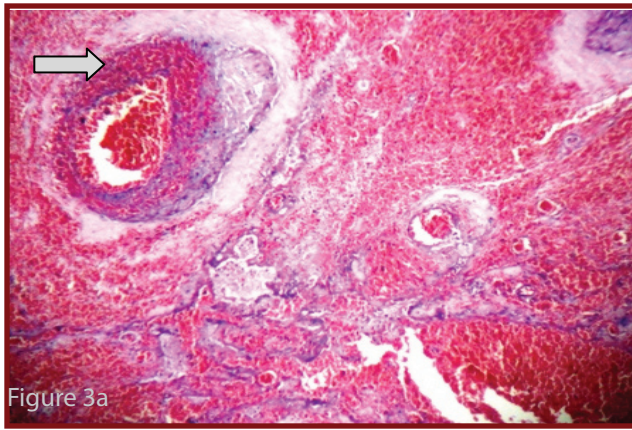


Figure 3: Histopathology of right middle lobectomy specimen of a 50 year old male with recurrent hemoptysis.

Figure 3a: Hematoxylin-eosin section shows lung tissue exhibiting areas of varicose thin and thick walled vascular channels extending up to the pleura (grey arrow).

Figure 3b: Elastic stain (von Geison) highlights the elastic lamina of the vascular channel (grey arrow).

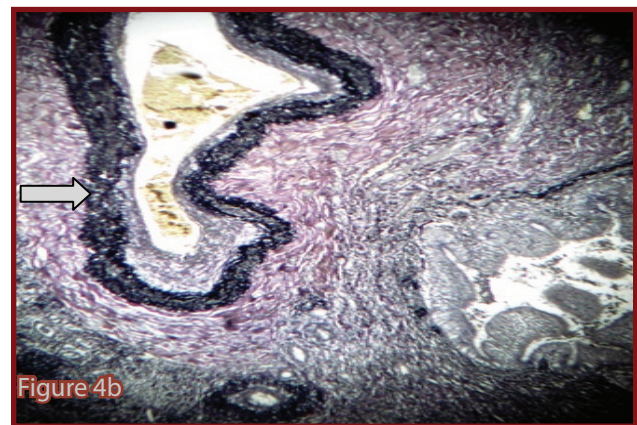
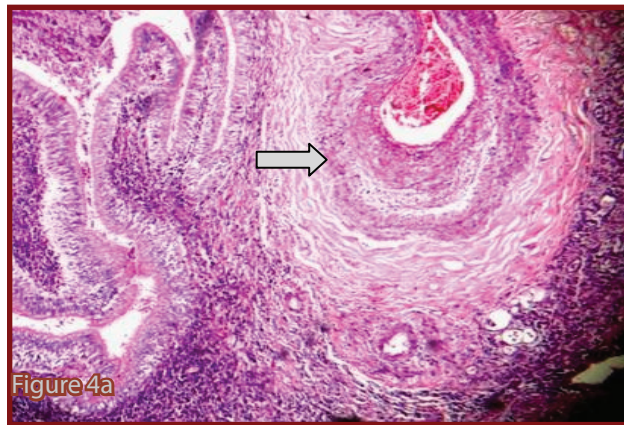


Figure 4: Histopathology of right middle lobectomy specimen of a 50 year old male with recurrent hemoptysis.

Figure 4a: Hematoxylin-eosin section shows area with thick walled vascular channel adjacent to a bronchus (grey arrow).

Figure 4b: Elastic stain (von Geison) highlights the elastic lamina of the vascular channel (grey arrow).

In patients with large or multiple PAVMs, they can present with platypnea; while some may present with stroke, massive fatal hemoptysis¹³ or brain abscess.¹⁴ Cryptogenic stroke in a young adult necessitates evaluation for the possibility of PAVMs in a previously undiagnosed patient.¹⁵

Our patient, with presented with a clinical picture of non-resolving pneumonia and later developed hemoptysis. The lesion was a 2 cm solid ill-defined mass that occupied the area between the bronchus and the pleura. It was in the right middle lobe in contrast to the more frequent locations, which are the lingula and right lower lobe. Unlike in classic case which presents with prominent thick vessels, the AV presents with prominent thick vessels, the AV malforma-

tion in our case was not grossly identifiable. In classic cases, the malformation was more appreciated on microscopic sections.

The practical approach in patients suspected with PAVM is to do a chest radiograph first, followed by shunt measurement by 100% oxygen method to confirm the presence of a right-to-left shunt.¹⁴ Definitive diagnosis is established by means of direct imaging with a contrast-enhanced study, such as computed tomography or pulmonary angiogram.¹

Contrast-enhanced CT scanning remains one of the standards in the diagnosis of PAVM. However, pulmonary angiography is performed to accurately define the anatomy, specifically

before therapeutic embolization is performed and when further intervention is planned.¹ In most situations, contrast-enhanced CT scanning is sufficed to confirm the diagnosis of PAVM. Therefore, if the plain chest radiographs suggest a PAVM, contrast-enhanced CT scan remains the preferred examination for confirming its presence.¹ Currently, CT angiography is 98% sensitive, 54% specific 50%.¹⁶ In our patient, the persistence of his hemoptysis warranted a CT angiography aside from the CT scan that confirmed a most-likely vascular lesion like AV-malformation.

It is important to understand the natural history of an illness in order to approach its treatment. Unfortunately, the natural history of untreated PAVM is not well delineated as there are no prospective reports of patients who were randomized to treatment versus observation only.⁹

When followed over time, PAVM typically remain unchanged in size, although about 25% enlarge gradually.¹² Because PAVM cause significant morbidity and mortality if lesions are untreated, radical treatment is generally mandatory as soon as PAVM is detected, even in asymptomatic patients. The aims of theaposto prevent bleeding episodes and neurological events following repeated cerebral embolism."¹⁷ Therapeutic options includes surgical removal, occlusion of fistulas by embolization, video-assisted thoracoscopic surgery¹⁸ and lung transplantation.¹⁷

CONCLUSION

In conclusion, since PAVMs are rare and unusual cases, there is a need for an updated multidisciplinary approach. The rarity of cases and the variability of presentation poised a great challenge. The advent of sophisticated imaging like CT angiogram provided our opportunity to deliver timely treatment. This case in particular which presented as a non-resolving pneumonia with an unusual location in the right middle lobe is a significant contribution to our registry of only 5 cases since 1976. As of the moment, we are successful in the management of this particular case, and its rarity should alert clinicians to include PAVM in the differential diagnosis of patients with pneumonia and hemoptysis.

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