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Endovascular Management of Pulmonary Arteriovenous Malformation

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ABSTRACT

Although many studies showed that pulmonary arteriovenous malformations (PAVM) are relatively rare disorders, they are still an important part of differential diagnosis of common pulmonary problems such as hypoxemia, dyspnea on exertion and pulmonary nodules.

A thirty-five-year-old female patient presented with low oxygen saturation six hours post simple vaginal delivery. The patient was investigated by echocardiography, chest x-ray and CT pulmonary angiography, which confirmed the diagnosis of pulmonary arteriovenous malformations (PAVM). Pulmonary conventional angiogram with transcatheter coil embolization was performed to treat the condition. The patient had uneventful recovery following coil embolization of pulmonary arteriovenous malformations.

INTRODUCTION

Direct communications between the branches of pulmonary arteries and pulmonary veins without an intervening pulmonary bed could result in formation of pulmonary arteriovenous malformations. Congenital cause is usually the source of these malformations except in very rare acquired cases[1,2].

Patient may present with symptoms related to PAVM, such as dyspnea on exertion, finger clubbing, cyanosis, hemoptysis and chest pain or underlying hereditary hemorrhagic telangiectasia (HHT), epistaxis and skin telangiectasia[3-6].

The aim of this report is to present a case report of pulmonary arteriovenous malformation which was treated by transcatheter coil embolization.

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THE CASE

A thirty-five-year-old female presented to Obstetrics and Gynecology Department with early labor pain. She is G3P2+0, previous deliveries were spontaneous vaginal deliveries, last delivery was five years ago. No significant medical or family history was revealed.

Six hours post-natal, she was desaturated (O_2 Saturation=86%) although her respiratory rate was 20. Her vital signs were normal. The uterus was contracted with only mild vaginal bleeding. General examination revealed clubbing of fingers and no cyanosis or skin telangiectasia.

Her laboratory investigations showed WBCs 1600, Neutrophils 82%, Hb 14.5 g/dl, Creatinine 69.5 umol/L, Urea 3.4 mmol/L, D-DIMER 0.19 mg/L, Prothrombin Time 12.2 seconds, INR 1.04 and APTT 27.4 seconds.

Echocardiography revealed normal cardiac structures, no evidence of intra-cardiac right-to-left shunt and no pulmonary hypertension.

Chest x-ray showed increased vascularity of right mid-lung zone assuming tubular and small nodular fashion, see figure 1. CT pulmonary angiography revealed multiple serpiginous vessels predominantly involving apical segment of right lower lobe, see figure 2.

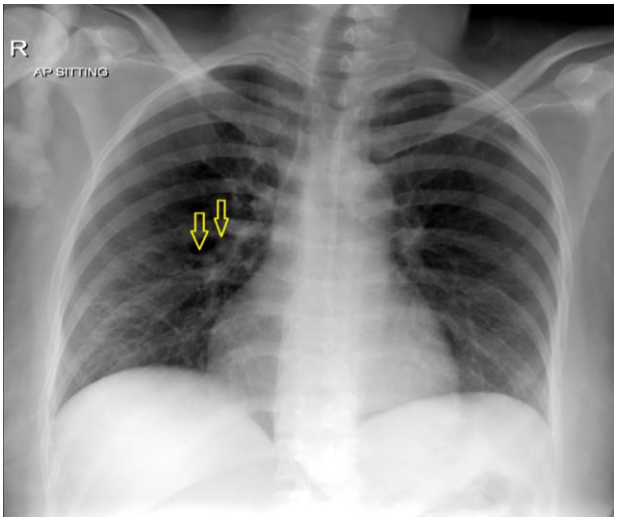

Figure 1: **Increased Vascularity of Right mid-Lung Zone Assuming Tubular and Small Nodular Fashion**

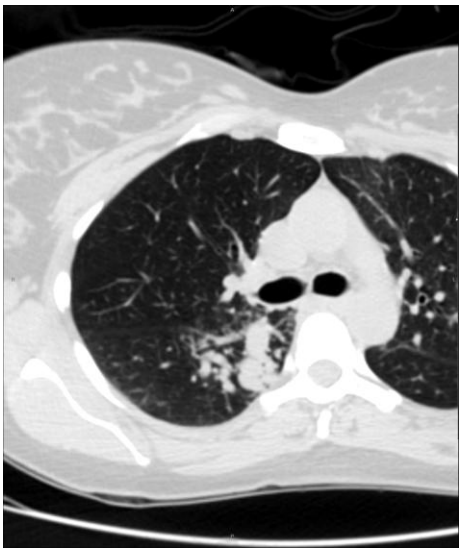

Figure 2: CT Pulmonary Angiography Revealed Multiple Serpiginous Vessels Predominantly Involving Apical Segment of Right Lower Lobe

The largest of the malformations had a feeder vessel from the right pulmonary artery and drained through the superior pulmonary vein into the left atrium. Small vascular malformations were seen in relation to the right inferior pulmonary vein and segmental branches of the right pulmonary artery. A hypertrophied bronchial artery was seen; no pulmonary embolic changes were noted. Treatment using transcatheter coil embolization was planned.

Pulmonary conventional angiogram was performed. Local anesthesia was used; the right common femoral vein approach was secured by 5F sheath selective cannulation, 5F CII cobra catheter was manipulated over guide wire and selective cannulation of right pulmonary trunk was done followed by selective right-sided pulmonary angiogram revealing two focal areas of AVM with multiple feeders seen at the right middle and lower lung fields which correspond to previously detected PAVM at CT scan, see figure 3.

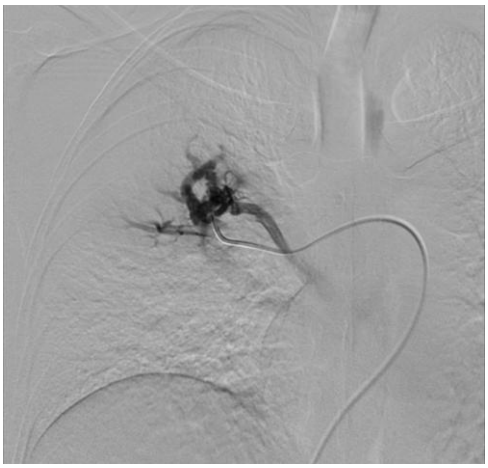

Figure 3: Pulmonary Angiogram Revealing Two Focal Areas of AVM with Multiple Feeders Seen at Right Middle and Lower Lung Fields

Super selective cannulation of the main feeder was done followed by coil embolization using single detachable coil (6mmx50cm Axium coil). Post-embolization angiogram revealed good response, see figure 4. No acute complications were encountered during the procedure. Follow-up revealed patient's clinical improvement confirmed by improvement of her respiratory rate (14/minute) as well as the O_2 saturation level which had increased to 94% after procedure.

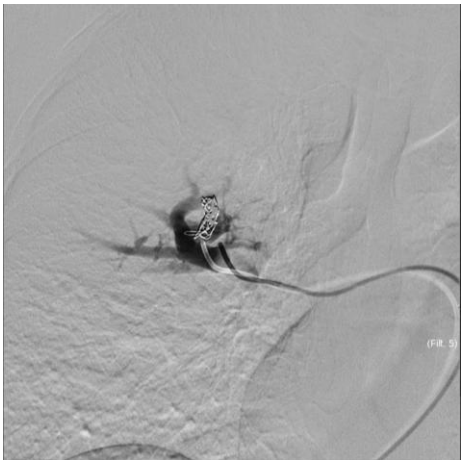

Figure 4: Post-embolization Angiogram Showing No More Opacification of Previously Detected AVM

DISCUSSION

Pulmonary arteriovenous malformation is a rare condition. The first reported case was in 1897 and since then, only three cases of PAVM were detected in 15,000 consecutive autopsies performed by Sloan and Cooley in 1953[7].

Mayo Clinic reported 194 cases of PAVM giving an annual incidence of 4.3 cases per year[8-10]. More than 80% of PAVMs are congenital and approximately 70% of those cases are associated with hereditary hemorrhagic telangiectasia (HHT). It is estimated that approximately 5%-15% of patients with HHT have PAVM.

The genetic linkages to HHT are located on chromosome 9 in some families and chromosome 12 in others[11]. Acquired pulmonary AVM has been reported in hepatic cirrhosis, schistosomiasis, mitral stenosis, metastatic thyroid carcinoma, trauma and chest surgery.

Increased blood volume, increased cardiac output and progesterone-induced venous distensibility during pregnancy has been associated with an increased rate of PAVM growth[12]. Increased steroid hormone synthesis during pregnancy results in an increased incidence of spontaneous hemothorax due to intrapleural rupture of PAVM[13].

PAVM has a male to female ratio of 1:2. PAVM may be single or multiple, the left lower lobe being the most common location for single PAVM while the majority of multiple PAVMs are confined to bilateral lower lobes[14].

There are two different types of PAVM either simple or complex, which is classified according to feeding segmental artery and draining vein. In case there is a single feeding segmental artery as well as single draining vein, it is named as simple PAVM which could be seen in 80% of cases; however, if there are two or more segmental feeding arteries or draining veins, it is named as complex type which could be seen in 20% of cases[15].

The pathogenesis of PAVM is not yet determined. Some authors suggest that PAVM is due to a defect in terminal arterial loops which allow dilatation of thin-walled capillary sacs. Asymptomatic patients are common and correlate with lesion size where a single PAVM <2 cm in diameter does not cause symptoms.

Patients with PAVM usually have symptoms referable to PAVM or underlying HHT in approximately 72% of cases. PAVM symptoms often develop between the fourth and sixth decades of life, which includes dyspnea and hemoptysis. HHT symptoms are noticeable before the age of 20 years including epistaxis and skin telangiectasia. Signs associated with PAVM include clubbing of fingers, cyanosis and pulmonary vascular bruit[11].

Chest x-rays could show a round or oval sharply defined mass of uniform density, frequently lobulated ranging from 1-5 cm in size and in two-thirds of cases located in the lower lobes[14]. CT angiography is significantly better than conventional angiography in detecting PAVM with a sensitivity of 97%. However, angiography was better in determining the angio-architecture of individual PAVMs than CT[16].

Pulmonary angiography is justified to confirm the diagnosis in virtually all cases and define the angio-architecture of pulmonary vasculature which is necessary before therapeutic embolization or surgery. Pulmonary angiography should be performed on all long portions to discover any unsuspected PAVM and source of intra or extra-thoracic vascular communications. Other diagnostic modalities include contrast echocardiography, MRI and isotope scan.

The natural course of PAVM is not benign. These lesions can be associated with many complications, such as massive hemoptysis due to intra-bronchial rupture, hemothorax due to intra-pleural rupture, stroke, brain abscess, paradoxical embolism and congestive heart failure[11]. Spontaneous hemothorax during pregnancy has been reported secondary to increased steroid hormone synthesis[13].

Treatment should be offered to all symptomatic patients and asymptomatic patients with lesions larger than 2 cm in diameter.

Embolotherapy, using stainless steel coils and vascular plugs, seems to be preferable in most cases especially in patients with multiple or bilateral PAVM and in patients who are poor operative risks.

Surgery is the best choice for fail embolotherapy, who develop serious bleeding despite embolotherapy or have untreatable allergy to contrast material[17,18].

CONCLUSION

A thirty-five-year old female patient with right pulmonary AVM treated successfully with endovascular coil embolization. Postoperative period was uneventful.

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