Case Report

Pulmonary Arterio-Venous Malformation

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Abstract

Pulmonary arteriovenous malformation (PAVM) is an abnormal communication between the pulmonary artery and the pulmonary vein. PAVMs are usually congenital in origin. In this study we reported a 32-year-old woman that was referred to the clinic with the chief complain of frequently coughs and hemoptysis for three months. Also, her past medical history revealed that she had a thoracotomy and decortication of the right lung due to clotted hemothorax, about 12 years ago while she had a term pregnancy. CT angiography and spiral CT-scan demonstrated cystic-masses in the right hemithorax. These masses were enhanced like pulmonary vessels and had feeding artery and evacuator vein. So, three PAVMs were diagnosed in the right lung. The patient underwent right postrolateral thoracotomy. Two of the PAVMs were in the lower lobe which treated by resection but the one in the middle lobe was completely intraparenchymal and it could not be localized during the surgery so the lobectomy of the right middle lobe was performed.

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Introduction

First described in 1897, pulmonary arteriovenous malformation (PAVM) is an abnormal communication between the pulmonary artery and the pulmonary vein. PAVMs are usually congenital in origin; however, they may be acquired in a variety of conditions, such as hepatic cirrhosis, schistosomiasis, mitral stenosis, trauma, actinomycosis, and metastatic thyroid carcinoma. Approximately 50-70% of PAVMs are located in the lower lobes. About 70% of patients have unilateral disease, 36% have multiple lesions, and 50% may have bilateral disease. The size of the PAVMs may vary from microscopic to the typical size of 1-5cm. Approximately 70% of PAVMs are associated with hereditary hemorrhagic telangiectasia (HHT), and about 15-30% of patients with HHT have a PAVM. 1 The most common clinical symptoms are epistaxis, dyspnea, hemoptysis, palpitations, hypoxemia and easy fatigability. The procedure of choice for treating a PAVM is therapeutic embolization rather than lung resection. Lung surgery is performed only when the PAVM is larger than 1cm or when the risk of systemic embolization is significant.1,2 In this study, in addition to review the articles, we reported a case of PAVM that was treated by resection and lobectomy. Also, the clinical, radiological and pathologic findings are included.

Case Report

A 32-year-old woman referred to the clinic with the chief complains of frequently coughs and hemoptysis for three months. The examinations did not reveal any specific positive findings, such as decrease of the lung sound, rale, murmur and stridor. Also, her past medical history revealed that 12 years ago, while she was pregnant and had only one week till her term pregnancy was brought to Emergency Department by Emergency Medical Services System with dizziness and dyspnea. O2 saturation with nasal oxygen was 88% and the auscultation revealed that the sound of the lungs was decreased in the right hemithorax. According to the patient’s unstable condition and due to her pregnancy, Chest X-ray was performed with shield. The chest X-ray demonstrated haziness in the right hemithorax with blunt costophrenic angle. Tube thoracostomy was performed due to the hemothorax. The patient admitted to the surgical ICU and at the end of the first week she had a normal vaginal delivery. After about one month CT-scan of the chest had been demonstrated collapse of right long due to clotted hemothorax. Then she underwent thoracotomy and decortication of the right lung. The patient was well after the appropriate postoperative management and discharged. But, recent chest X-ray demonstrated a radiopaque cystic-mass with approximate size of 27*28 mm in the right hemithorax, near the right side of the heart. Spiral CT-scan of the chest with IV contrast was demonstrated cystic-masses with soft tissues that their size and location were as follows:
- 52*29 mm in the medial segment
- 27*28 mm in posterior segment of inferior lobe of the right lung
A smaller nodule with 10 mm diameter in central segment of middle lobe of the right lung CT angiography of the chest demonstrated cystic-masses that were enhanced like pulmonary vessels and had feeding artery and evacuator vein, similar to Arterio-Venous Malformation (Fig. 1).

Fig 1- CT Angiography of the Patient
The patient underwent right postrolateral thoracotomy with general anesthesia by double lumen endotracheal tube. First pneumolysis were performed. The bigger PAVM was subpleural and its artery and vein were ligated so, it was resected simply. The second one was intraparenchymal and found by deflating of right lung in addition to waiting and watching the suspicious area of the lung (Fig. 2). The Arterio-Venous Malformation of the middle lobe was completely intraparenchymal and it could not be found during the surgery so the lobectomy of the right middle lobe was performed.

Discussion

Although the PAVMs pathogenesis is not well delineated, they are considered to result from incomplete resorption of the vascular septa. These vascular septa separate the arterial plexus and the venous plexus, which normally anastomose during fetal development. Progressive dilatation of the smaller plexus leads to the formation of tortuous loops and multiloculated sacs. With rupture of intervening vascular walls, a single large, saccular PAVM develops. PAVM occurs twice as often in women as in men, but a male predominance exists in newborns. Approximately 10% of PAVM cases are identified in infancy or childhood; however, the incidence gradually increases through the fifth and sixth decades of life. PAVMs can be classified as simple or complex types, based on their architecture. Simple PAVMs have a single feeding segmental artery leading to single draining pulmonary vein. Approximately 79% of PAVMs are of the simple type and occur in lower lobes; these are associated with nonseptate aneurysms. Approximately 21% of PAVMs are complex; these have 2 or more feeding arteries or draining veins. They often occur in the distributions of the lingula and the right middle lobe. Whenever a PAVM is suspected, the presence of a right-to-left shunt should be confirmed by the performance of a 100% oxygen study, contrast-enhanced echocardiography, or radionuclide perfusion lung scanning. A definitive diagnosis is established by means of direct imaging of the PAVM with a contrast-enhanced study, such as a computed tomography (CT) scan or a pulmonary angiogram. Three-dimensional (3D), spiral CT scanning produces images of vascular structures that are continuously reconstructed. In one study, spiral CT scanning proved to be a better investigative tool than unilateral pulmonary angiography. The accuracy of 3D spiral CT scanning is reported to be more than 95%, and it may also be useful in identifying smaller PAVMs. Surgical excision of an isolated, single PAVM is successful, with minimal mortality and morbidity and little chance of recurrence of the lesion. Surgical excision is the procedure of choice when it can be accomplished. Because most fistulas are located subpleurally, they can be removed with conservative local resection. In difficult cases in which the feeding vessel is not definitely localized preoperatively, Almeida and colleagues (1998) suggest the use of transesophageal echocardiography with agitated blood and saline contrast during the procedure in order to assist in localizing the vessel. Puskas and associates (1994) reported the successful excision of a solitary arteriovenous fistula in nine patients; one patient had staged excision of bilateral lesions. All patients were relieved of dyspnea, and no recurrences or neurologic complications occurred in the follow-up of these patients. Also, multiple fistulas are occasionally suitable for resection. However, Sperling and colleagues (1977) reported that the combination of pulmonary hypertension and arteriovenous fistulas is a contraindication for surgical excision. But the procedure of choice for treating a PAVM is therapeutic embolization rather than lung resection. Lung surgery is performed only when the PAVM is larger than 1 cm or when the risk of systemic embolization is significant.
References

