CASE REPORT

PULMONARY ARTERIOVENOUS MALFORMATION TREATED BY LOBECTOMY

Mustafa Yüksel1, Mehmet Oğuzhan Özyurtkan2, Nurhayat Topaloğlu3, Tunç Laçın1

1Department of Thoracic Surgery, School of Medicine, Marmara University, İstanbul, Turkey 2Department of Thoracic Surgery, School of Medicine, Fırat University, Elazığ, Turkey 3Department of Pulmonology, School of Medicine, Marmara University, İstanbul, Turkey

ABSTRACT

Pulmonary arteriovenous malformation (PAVM) is an abnormal communication between pulmonary arteries and pulmonary veins. These lesions are quite uncommon; they are an important part of the differential diagnosis of common pulmonary problems such as hypoxemia and pulmonary nodules. PAVM was observed in a 32-year-old woman who had the classical triad of cyanosis, polycythemia, and clubbing of the fingers and toes. She had also a brain abscess treated by drainage 10 years ago. Pulmonary angiography revealed two PAVMs located in the left upper lobe. She was treated by lobectomy.

Keywords: Arteriovenous malformations, Angiography, Clubbing, Polycythemia, Lobectomy

INTRODUCTION

PAVM is caused by abnormal communication between pulmonary arteries and pulmonary veins, which are most commonly congenital in nature. These lesions are quite uncommon; they are an important part of the differential diagnosis of common pulmonary problems such as hypoxemia and pulmonary nodules. PAVM may also be found in a variety of acquired conditions such as hepatic cirrhosis, schistosomiasis, mitral stenosis, actinomycosis, trauma, Fanconi’s syndrome, and metastatic thyroid carcinoma. Approximately 70% of the cases of PAVM are associated with hereditary hemorrhagic telangiectasia (HHT). Conversely, 15 to 35% of the patients with HHT have PAVM.

Here in this paper, we reported a case of PAVM in a 32-year-old woman who had the classical triad of cyanosis, polycythemia, and clubbing of the fingers, and treated by lobectomy.

CASE REPORT

A 32-year-old woman presented with a 31 year history of cyanosis of nails and lips. Her medical history was significant for acute rheumatic fever at 16 years of age, and a right temporal brain abscess drainage at 22. She had no family history of hereditary hemorrhagic telangiectasia (HHT). Remarkable signs on laboratory work-up included low arterial blood oxygen saturation of 83%, cyanosis and polycythemia, hemoglobin 20.4g/dl. On physical examination she had clubbing of fingers and an upper lobe chest bruit over posterior left hemithorax, which was increasing in pitch and volume with inspiration. A spiral chest computed tomography with intravenous contrast revealed two high-density large nodular lesions.
located in the apical and lingular segments of the upper lobe. Pulmonary angiography revealed two PAVMs in the left upper lobe, apical one being larger than 2cm in diameter, and the lingular one being localized centrally (Fig. 1).

Fig. 1: Pulmonary angiography showing two PAVMs located in the left upper lobe.

With the patient under general anesthesia and double lumen endotracheal intubation and a thoracic epidural catheter, the patient underwent left posterolateral thoracotomy. Before entering the left thoracic cavity through the fifth intercostal space, the affected lung was deflated. Following deflation, the arterial blood oxygen saturation of the patient suddenly increased from 85% to 95%, and remained constant during the rest of the operation. Visual and manual examination revealed two nodular lesions, located one in the apical segment, and the other one in the lingular segment. A left upper lobectomy was performed without complication. At the end of the procedure, she was extubated and transferred to the ward. She had an uneventful postoperative course, and was discharged on 10th postoperative day. Her follow-up visit at 3rd months showed no cyanosis, no dyspnea on exertion, and she had an arterial blood oxygen saturation of 97% at room air. Histolopathologic study of the resected lobe revealed, two plexiform masses of dilated vessels with feeding vessels. Of them, the one in the apical segment measured 3x3 cm, and the other one in the lingular segment measured 2x1.5 cm.

DISCUSSION

PAVMs occur more frequently in women and are transmitted as a dominant gene with incomplete penetrance. 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. Epistaxis, dyspnea, and haemoptysis are the commonest symptoms. Other frequent extra pulmonary symptoms and signs are headache in 43%, transient ischemic attacks in 57%, and cerebrovascular accidents in 18%.

Intracerebral abscess occurs in 33% of the cases. A typical bruit, present in 38% of those who have no HHT, has a rough, humming, continuous sound accentuated in systole and with deep inspiration. The classical triad of cyanosis, polycythemia, and clubbing has been noted in approximately 20% of the patients.

Contrast echography is an excellent tool for evaluation of cardiac and intrapulmonary shunts. Radionuclide perfusion scan is also useful in the diagnosis of PAVMs, and may be used as an adjunct to the contrast echocardiography or alone. The presence of a PAVM and its vascular anatomy can also be evaluated by contrast-enhanced ultrafast chest tomography. Magnetic resonance imaging may be useful in differentiating PAVM from various types of pulmonary nodules, but the main limitations include limited availability, and the highly specialized techniques required for accurate interpretation. Despite advances in the different techniques, contrast pulmonary angiography remains the gold standard in the diagnosis of PAVM, and is usually necessary if resectional or obliterator therapy is being considered.

It is recommended that all symptomatic PAVM and PAVM> 2 cm in diameter be treated with either surgery or embolotherapy, and nowadays embolotherapy (ballon and coil embolization) is considered first-line therapy in the treatment of most PAVMs since it is a less invasive method than surgery and has a lower complication ratio. White reported 276 cases of PAVMs successfully treated by balloon embolotherapy with an overall complication ratio of 1%. The complications of embolotherapy, such as pleuritic chest pain, pulmonary infarction, and deep venous thrombosis, have in general been infrequent and self-limited. Such complications, as well as paradoxical embolization of coils and balloons may occur especially in patients with PAVMs with large feeding arteries. For large and centrally localized lesions, or for lesions with large feeding vessels, surgery is still a proposed treatment modality since it carries no additional risks than other thoracic operations. In the study of Swanson, surgical resection alone was carried out in 19%, embolization therapy alone in 44%, and both therapies in 8%, and frequent follow-up of treated patients is advised because PAVMs tend to increase both in number and in size over time. Embolotherapy was not advised in our patient, since both lesions were located at the left
upper lobe; apical one was large in diameter, and the lingular one was central in localization.

We successfully treated a PAVM of the left upper lobe by lobectomy in a 32-year-old woman who had the classical triad of cyanosis, polycythemia, and clubbing. A sudden improvement in the arterial blood oxygen saturation of the patient following lung deflation was a remarkable finding. In selected cases, where PAVM is localized and large in diameter, or when the embolotherapy is not suitable, surgical resection is a safe method in the treatment of PAVM.

REFERENCES