Surgically Treatable Pulmonary Arteriovenous Fistula

Saviz Pejhan, MD,1 Nasrin Rahmanijoo, MD,2 Roya Farzanegan, MD,1 and Mostafa Rahimi, MD3

Arteriovenous fistuli are congenital malformations. Usually symptoms depend on size of the lesion. Lesions smaller than 2 cm are often asymptomatic. The most common symptoms are dyspnea, palpitation and fatigue. Cyanosis is indicative of right to left shunt. Helical computed tomography (CT) scan is a helpful diagnostic tool in this case. Surgery is the treatment of choice in patients with isolated lesions. Embolization is a selective method in patients with multiple or bilateral lesions.

The patient was a 13-year–old boy complaining of cyanosis of lips and nails as well as dyspnea for 5 years. Definite diagnosis of pulmonary arteriovenous malformation (PA VM) in the right middle lobe was based on CT angiography. The patient underwent a thoracotomy and lobectomy of the right middle lobe. After surgery cyanosis and dyspnea were completely resolved.

Keywords: pulmonary arteriovenous fistula, surgery, treatment

Introduction

Pulmonary arteriovenous fistuli are known as an abnormal connection between the pulmonary artery and vein which is usually congenital, but in special conditions, such as cirrhosis, trauma and schistozomiasis they can be acquired too.1) It is believed to be the most prevalent malformation of pulmonary vascular system.2) These fistuli are usually seen in the 3rd or 4th decade of life and more commonly among women. They disturb the filtering action of pulmonary capillaries and cause thromboembolic event in systemic circulation.3) Approximately 50%-70% of these fistuli exist in the lower lobes of lungs. Fistuli are isolated in half or two thirds of patients.4) Seventy percent of fistuli are unilateral, while only 36% of them are multiple and 50% are bilateral. Size of the lesion varies from microscopic to 1–5 cm.5)

Dyspnea, palpitation, fatigue and epistaxia, as well as hemoptysis, are common symptoms and superficial telangiectasia is a common sign.6)

Chest X-ray is abnormal in 98% of patients, and the lesion is seen as lobulated density with indistinctive margin connected to the hilum.

Helical CT scan is a helpful diagnostic tool in this case.5) Most of patients with one or more PAVF are candidates for resection of the lesion. Patients with Osler-Weber-Rendu syndrome are indicated for treatment, since the rate of complication is high in this group; asymptomatic patients, those with small lesions (10–15 mm) or who have a small shunt are not treated, though their condition is followed regularly.6,7)
An intervention is necessary in all symptomatic patients even with mild symptoms whose lesions are visible in a chest X-ray or CT scan.

Excision is a highly successful procedure in isolated fistuli, which is recommended in patients with permanent bleeding secondary to intrapleural rupture or hemoptysis despite embolization. Also mortality and morbidity and the recurrence rate are low.6)

Since most of the fistuli are sub-pleural, they are curable by resection. In Japan 2001, limited resection in 3 PAVF patients was a successful procedure for treatment of such fistula.8)

Case

A 13-year-old male patient presented by cyanosis of lips and nails and dyspnea for 5 years. The symptoms had recently exacerbated. In physical examination, cyanosis of lips and nails and clubbing of fingers were found, but no skin telangiectasia was detected. Other organs were normal. Oxygen saturation was 68%, which reached 91/8% by the administration of 100% oxygen. In lab tests, the positive point was polycytemia (Hb = 18.5, RBC = 6.56 × 10⁶). LVEF was reported 67% in echocardiography. Chest CT scan with contrast showed opacity with vascular changes in the right middle lobe. In CT angiography, typical AVM was clearly found in the right middle lobe (Fig. 1), while in bronchoscopy, there was no endobronchial lesion. The patient underwent a thoracotomy and lobectomy with diagnosis of isolated right lung AVM.

In left lateral decubitus position, we accessed thorax through the 5th intercostal space by posterolateral incision. The right middle lobe was completely occupied by AVM and totally enlarged and tortuous, which was double ligated; also, the right middle lobe lobectomy was classically performed (Fig. 2a and 2b).

Immediately after ligation of fistula, O₂ saturation rose to 98% and cyanosis was completely resolved. There were no complications, and in the follow-up, the patient was free of symptoms. Pathologic findings were reported as “lung tissue with focal abnormal proliferation of vascular channels with thick walls along with diffuse congestion and alveolar hemorrhage of surrounding lung tissue and diffuse interstitial angiomatosis” (Fig. 3).

Discussion

Pulmonary Arteriovenous fistuli are congenital malformations, which can grow bigger and develop...
complications, in particular symptomatic fistula and the ones larger than 2cm. So treatment is seriously recommended. The appropriate method of treatment depends on the number and location of fistula. The excision is highly successful for an isolated fistula, and the mortality rate is low in this surgery.

Also, an excision is indicated in patients whose hemoptysis continues after embolization or in cases of rupture of fistula to the pleural cavity. Subpleural malformations can be removed by local resection. Embolization is an appropriate treatment modality in multiple arteriovenous fistulae which are not suitable for surgery. Since these patients have massive right to left shunt and are disabled. In one report, a right to left shunt in the lower lobe of the left lung with multiple coil embolizations has been completely cured in 6 months.9)

**Conclusion**

Surgical treatment (resection) in isolated pulmonary AVF is recommended due to its safety and low recurrence and mortality rate.

**References**

1) Khurshid I, Downie GH. Pulmonary arteriovenous malformation. Postgrad Med J 2002; 78: 191-7. doi:10.1136/pmj.78.918.191

**Fig. 3** Pathology findings.