Case report

Pulmonary arteriovenous malformation treated by lobectomy

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Abstract

Pulmonary arteriovenous malformation (PAVM) may occur primarily or in association with hereditary hemorrhagic telangiectasia. We present a case of PAVM in the central lower lobe of the left lung of a 75-year-old woman, which was successfully treated by lobectomy. Contrast echocardiography is an excellent tool for evaluation of this uncommon lesion. Advances in interventional radiology have led to the introduction of obliterative techniques for the treatment of PAVM. However, in the presence of a large solitary malformation centrally located, as in our case, and in high-risk patients, surgery is still a safe and effective first option.

Keywords: Pulmonary arteriovenous malformation; Hereditary telangiectasia; Surgery

1. Introduction

Pulmonary arteriovenous malformations (PAVMs) are caused by an abnormal communication between the pulmonary arteries and veins, and are usually congenital. They may occur as an isolated anomaly or in association with hereditary hemorrhagic telangiectasia (HHT) (Rendu-Osler-Weber syndrome). The right-to-left shunting induces chronic hypoxemia or paradoxical embolism (often infections). We describe a 75-year-old woman with a large PAVM on the left lower lobe of long duration, which had recently become symptomatic (dyspnea on exertion, cyanosis and polycythemia). She underwent successful left lower lobectomy without complications.

2. Case report

A 75-year-old woman with a PAVM on the left lower lobe, which was known to be present for years but had only recently become symptomatic (dyspnea on exertion), was admitted to our department for elective left lower lobectomy. There was no family history of HHT. Remarkable signs on laboratory work-up included low arterial blood oxygen saturation of 65–70%, cyanosis and polycythemia, hemoglobin 16.4 g/dl. Physical examination revealed a left lower posterior chest bruit, which increased in pitch and volume with inspiration. Chest computed tomography (CT) with contrast enhancement showed a high-density large nodular lesion centrally located with connecting vessels in the subpleural region of the left lower lung (Fig. 1a). Contrast echocardiography by injection of 5 ml saline (agitated with a small amount of air) into a peripheral vein showed microbubbles in the left atrium after 5 s, thus confirming intrapulmonary shunt.

The patient underwent left posterolateral thoracotomy. Findings at surgery were a large pulsating complex PAVM (Fig. 1b) measuring 8.0 £ 8.0 cm, almost completely occupying the left lobe, which was supplied by both the superior segmental and main basal pulmonary arteries. In addition, there were large systemic collaterals from the aorta. The pulmonary artery and vein were ectatic.

Following resection, blood gases in room air showed a dramatic improvement from baseline (Table 1). On pathohistologic study of the resected lobe, a plexiform mass of dilated vessels with feeding vessels were noted. The postoperative course was uneventful, and the patient required only minimal administration of analgesics with epidural analgesia. The patient was acyanotic and was discharged after 4 days. Follow-up visit at 12 months showed acyanosis, no dyspnea on exertion and room air arterial blood oxygen saturation of 97%. Blood gas analysis.
in room air showed a dramatic improvement from baseline (Table 1).

3. Discussion

PAVM was first described in 1897 by Churton [1], in a 12-year-old boy found to have multiple bilateral PAVMs on postmortem examination. Although these lesions are quite uncommon, they are an important part of the differential diagnosis of common pulmonary problems such as hypoxemia and pulmonary nodules. They have been reported in the literature under various names, including pulmonary arteriovenous aneurysms, hemangiomas of the lung, cavernous angiomas of the lung, pulmonary telangiectases, and pulmonary arteriovenous malformations [2]. PAVM may be simple (single pulmonary artery-to-pulmonary vein communication) or complex (multiple feeding arteries and draining veins) [3], and single (40%), multiple (40%), or bilateral (20%). The vast majority are congenital; 36% of single lesions and 57% of multiple lesions are associated with HHT [4]. Fifty-three to 70% of PAVMs are found in the lower lobes [2]. Women are affected twice as often as men, but there is a male predominance in newborns [5].

The exact pathogenesis of PAVM is unknown. Some investigators have hypothesized that the cause is a defect in the terminal arterial loops, which allows dilatation of the thin-walled capillary sacs [6]. Others have argued that PAVM is the result of incomplete resorption of the vascular septae that separate the arterial and venous plexuses which normally anastomose during fetal development [6]. It has also been suggested that multiple small PAVMs are induced by a failure of capillary development in the fetal stage [6].

The work-up of patients with suspected PAVM should start with a detailed history of epistaxis, dyspnea on exertion, or family history of telangiectasia. Many patients are asymptomatic, though 6% have brain abscesses [7] due to bacteria bypassing the natural pulmonary vascular filter and lodging in the cerebral circulation. A detailed physical examination, with a specific search for chest bruits (30%), nasopharyngeal telangiectasia (33%), clubbing, or cyanosis is required. In 95% of patients, chest roentgenogram [8] shows peripheral, circumscribed, non-calcified lesions that can be misinterpreted as infiltrates, metastatic lesions, or coin lesions. Contrast-enhanced CT scan and magnetic resonance imaging may be useful, if other techniques are inconclusive, and arterial blood gas evaluation may help to establish the diagnosis. Most patients have hypoxemia with a high incidence of desaturation in the upright position (orthodexia). Oxygenation does not improve significantly with supplemental oxygen because of the fixed right-to-left shunt. Contrast echocardiography is an excellent tool to evaluate cardiac and intrapulmonary shunts, and it can identify small right-to-left shunts even when they are not suggested by the gas exchange data [6]. In the case of PAVM, there is nearly always a delay of three to eight
cardiac cycles (2–5 s) before contrast is visualized in the left atrium, due to the time required for the contrast material to traverse the pulmonary vasculature [6]. Pulmonary angiography is helpful in differentiating the origin of the arterial supply to the AVM (pulmonary or systemic).

Treatment of PAVM should be based on the size, number, and location of the lesions and the specific complications, as well as the patient’s general condition. Traditional indications for treatment have been progressive PAVM enlargement, paradoxical embolization, and symptomatic hypoxemia [6]. As PAVM has a tendency to progression and complications, the most radical and least invasive method of treatment should be used. Before 1978, surgical resection was the only method available; ligation, local excision, segmentectomy, lobectomy, or pneumonectomy was performed in most cases, though some clinicians opted for conservative management and observation in some asymptomatic patients. Since then, recent advances in interventional radiology have led to the successful application of percutaneous transcatheter embolization by coil or balloon for multiple PAVMs. This method is less invasive than surgery and has a lower complication rate. Today, it is the first choice in most cases of PAVM in institutions with the necessary expertise and facilities [9]. Nevertheless, for large, centrally localized lesions, as in our case, lobectomy is still required. Therapeutic surgery for PAVM carries at least the same risk as any other thoracic operation, but when properly performed in well-selected patients, it is associated with minimal morbidity and mortality, with only rare postoperative recurrences [4].

We conclude that surgery is a safe method of treatment of PAVMs in selected cases, i.e. when the PAVM is solitary and large (> 2 cm diameter), and the risks of embolotherapy are high.

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**References**


**Table 1**

Patient’s arterial blood gases

<table>
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<tr>
<th></th>
<th>FIO2 (%)</th>
<th>pH</th>
<th>PCO2 (mmHg)</th>
<th>PO2 (mmHg)</th>
<th>HCO3 (mEq/l)</th>
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<td>Preoperative (baseline)</td>
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<tr>
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<td>80</td>
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