Case report - Pulmonary

Segmental arterial ligation under video-assisted thoracoscopy combined with transcatheter embolotherapy for multiple bilateral pulmonary arteriovenous malformations

Mitsutomo Kohno*, Kazumasa Noda, Hirokazu Moriyama, Yoshihiro Nishimura

Department of Surgery, Tokyo Metropolitan Komagome Hospital, 3-18-22 Honkomagome, Bunkyo-ku, Tokyo 113-0021, Japan

Received 15 March 2004; received in revised form 21 June 2004; accepted 5 July 2004

Abstract

Multiple bilateral pulmonary arterio-venous malformations (PAVM) were diagnosed in a 15-year-old girl with erythrocytosis. Although transcatheter embolotherapy is generally accepted as a treatment of PAVM, the largest lesion in the right lower lobe was fed by a 13-mm diameter feeding artery without downstream narrowing, which was unsafe for the embolization of coils or balloons. Instead, we surgically ligated the largest feeding artery under video-assisted thoracoscopy, without losing any lung parenchyma. The other PAVM were successfully treated by embolization of metallic coils. The arterial partial pressure of oxygen (O2) increased from 37 to 64 mmHg on room air, and from 46 to 410 mmHg on 100% O2.

Keywords: Pulmonary arteriovenous malformation; Rendu–Osler–Weber syndrome; Therapeutic embolization; Pulmonary artery ligation; Video-assisted thoracoscopic surgery

1. Introduction

Pulmonary arterio-venous malformations (PAVM) are abnormal direct communications between a pulmonary artery and a pulmonary vein through a thin-walled aneurysm. Though their etiology and prevalence have not been precisely characterized, PAVM have been reported in up to nearly 90% of patients suffering from Rendu–Osler–Weber hereditary, hemorrhagic telangiectasis [1]. Detailed studies revealed the presence of multiple lesions in between one- and two-third of patients [1,2]. Symptoms in early life may range from none to severe with cyanosis, congestive heart failure, and respiratory failure. Since patients with PAVM are at risk of several life-threatening complications, including neurologic complications, such as stroke, transient ischemic attack and brain abscess, or massive hemothorax or hemoptysis, it has been recommended to treat all PAVM with feeding vessels ≥3-mm in diameter, including in patients without symptoms [3].

This report describes a patient with multiple, bilateral PAVM treated by video-assisted thoracoscopic surgery combined with a percutaneous embolotherapy.

2. Case report

2.1. Clinical presentation

A 15-year-old girl was examined by a pediatrician at our hospital, after erythrocytosis was diagnosed at a high school health screening. Bilateral digital cyanosis without clubbing was noted, and a systolic bruit was audible in the right posterior hemithorax. The patient had no other abnormal physical finding and she reported no dyspnea related to her daily activities. An abnormal shadow was detected on chest roentgenogram, and bilateral PAVM were suspected by computed tomography (CT).

Arterial blood gas measurements revealed an arterial partial pressure of oxygen (PaO2) at 36 mmHg, and venous hemoglobin, hematocrit, and erythrocytes count were
15.8 g/dl, 50.3%, and 629 × 10^6/μl, respectively. Respiratory function tests and echocardiography were normal. The chest CT revealed a 3.5 × 2.5 cm nodule in the right anterior basal lung segment, preceded by a 13-mm diameter, dilated pulmonary artery. Thirteen additional malformations, <1 cm in diameter, were found over both lung fields.

The patient underwent pulmonary arteriography, which confirmed the presence of a 13-mm diameter arterial branch of the right lower lobe artery feeding the largest PAVM located in the anterior basal segment (Fig. 1). There were also a few other abnormal narrow arteries originating from A9, feeding the largest PAVM. The diameter of the effluent vein was also dilated up to 14 mm. The smaller PAVM were opacified in both lungs.

We considered performing percutaneous catheter embolization, though we were concerned that a coil or balloon may paradoxically embolize from the largest PAVM. Therefore, we chose to proceed with ligation of the largest feeding artery under video-assisted thoracoscopy, and embolize metallic coils into the smaller PAVM.

2.2. Procedures and outcome

Thoracoscopy was performed under one-lung ventilation in a left lateral decubitus position. After thoracoscopy via the seventh intercostal space to explore the thoracic cavity and confirm the absence of adhesions, we made other ports at the fourth intercostal space and opened the chest minimally. The interlobular lung tissue was dissected to reach the dilated right anterior basal feeding artery, which was twice ligated with 2-0 silk sutures.

Pulmonary arteriography performed after surgical ligation and first embolotherapy confirmed the occlusion of the right anterior basal artery, a reduced blood supply to the largest PAVM, and presence of occlusive coils embolized in other small feeding arteries of both lungs (Fig. 2). A few abnormal arteries, which originated from the right lateral basal artery and fed the largest PAVM, and other abnormal arteries at the right superior segment were initially left untreated, since their walls might have been weakened during the thorascopic intervention. These remaining arteries were successfully embolized in a second procedure 4 months later.

The thorascopic intervention and endovascular embolization procedures were uncomplicated. PaO2 on room air increased from 37 to 55 mmHg after surgical ligation only, and to 64 mmHg after the combined interventions. Likewise, PaO2 during exposure to 100% O2 increased from 46 to 98 mmHg after surgical ligation only, and to 410 mmHg after completion of both procedures. The arterial O2 saturation, which was 66% before treatment, had remained >90% at 4 months after combined therapy.

3. Discussion

Open-chest lesion excisions, segmentectomy, lobectomy or pneumonectomy have been performed to treat PAVM. The less invasive transcatheter embolotherapy to occlude feeding arteries with metallic coils or balloons can be performed repeatedly and has become the treatment of choice owing to advances in interventional radiology. However, rarely, large feeding arteries may be the source of various complications [1,2,4]. Coils or balloons may escape from the malformation and cause paradoxical embolization if the feeding artery does not narrow downstream. Premature autodeflation of balloons, recurrences from recanalization of occluded vessels, or post-procedural growth of accessory vessels may also occur. In our patient, the largest PAVM was fed by a huge artery, which appeared unsafe for the placement of coils or balloons. Therefore, we opted to proceed with surgical
ligation to allow a more precise occlusion of the feeding artery without loss of lung parenchyma. Furthermore, the use of video-assisted thoracoscopic surgery minimized the injury to respiratory muscles and the best cosmetic outcome in this 15-year-old girl.

In conclusion, surgical ligation under video-assisted thoracoscopy combined with transcatheter embolotherapy in an adolescent with multiple bilateral PAVM was safe and effective. This combined treatment should be considered for patients with multiple bilateral PAVM with large feeding arteries, who may be at high risk of paradoxical device embolization.

References