
Multiple Cavernous Hemangiomas of the Lung
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We present a rare case of multiple cavernous hemangiomas. A 19-year-old girl was seen with dyspnea and fatigue. Thoracic computed tomography showed multiple nodule shadows scattered in the lung. Lung biopsy was carried out. Postoperative histopathologic study identified the nodules as pulmonary cavernous hemangiomas.

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Cavernous hemangiomas are a variant of slow-flow pulmonary venous malformations. They can affect any visceral organ but rarely occur in the lung. According to a review of the international literature, only 10 cases of pulmonary cavernous hemangiomas (PCHs) were reported during the 60 years before 2010 [1]. We describe a patient with multiple PCHs verified by lung biopsy. We briefly discuss the clinical, radiographic, and pathologic features and the management strategies.

A 19-year-old girl sought medical attention at another hospital because of dyspnea and fatigue for 1 month on July 22, 2013. Laboratory analysis revealed hemoglobin 61 g/L, and further examination suggested iron-deficiency anemia. Thoracic computed tomography (CT) revealed multiple nodules with uniform density throughout both lung fields and left pleural effusion (Figs 1B, 1C). The clinical impression was malignancy. She was treated with iron supplementation and nutritional therapy. Two weeks later, her hemoglobin level had risen to 105 g/L. A chest radiograph revealed multiple nodules scattered throughout both lung fields, and the left pleural effusion was spontaneously absorbed (Fig 1A).

The girl was admitted to our hospital August 27, 2013, and she was then asymptomatic. Her family history was noncontributory. The result of physical examination was unremarkable except for scoliosis. Her hemoglobin level and platelet count were normal. Contrast-enhanced CT of the chest showed bilateral pulmonary nodules without prominent infiltration, and the left pleural effusion had disappeared. The radiologic findings of other organs revealed no abnormalities. Bronchoscopic examination demonstrated submucosal lesions at the right principal bronchus. Because it was not possible to rule out malignancy, lung biopsy by video-assisted thoracoscopic surgery was carried out. Examination of the right lung revealed numerous well-circumscribed, greyish red nodules of different sizes, diffusely distributed throughout the surface of all lobes (Fig 1D), especially the middle and lower lobes of the lung. Some of them merged together; they were soft and were easily collapsed with a probe. Biopsy samples were taken by wedge resection.

On gross inspection, the lesions involved lung parenchyma extending to subpleural tissue. Microscopically, the nodules were composed of large, dilated vascular channels separated from one another by only a scant connective tissue stroma, and the channels were filled with blood. Immunohistologic staining demonstrated that the lining cells of the cavernous structure stained positively for CD31 and CD34, which suggested that the lesions were of vascular origin. All the above features taken together led to a diagnosis of multiple PCHs.

The patient was discharged home on the fourth postoperative day without complications. At her monthly follow-up visit, she had no apparent symptoms.

Comment
Cavernous hemangiomas can occur throughout the body. They can be superficial, deep, or visceral, but they are predominantly found in the skin. Visceral hemangiomas rarely occur in the lung; they are frequently located in deeper organs such as the liver, kidney, colon, brain, or bones. A PCH can be found in a patient of any age, with the youngest reported patient being 10 weeks old [2] and the oldest 84 years old [3]. A PCH may be localized or diffuse. Clinical manifestations are dependent on the lesion’s location, number, and size. Some patients present clinical respiratory distress, cyanosis, hemoptysis, and even heart failure. Some patients show no apparent adverse symptoms even after death resulting from other causes [4]. In our case, the girl was dyspneic, with pleural effusion and low hemoglobin, and we speculate that the pleural effusion was caused by spontaneous tumor rupture of the subpleural lesions. The spontaneous absorption also supported the diagnosis of spontaneous hemotorax.

PCHs may involve the parenchyma and the airway. Airway PCHs are rare but may be life-threatening as a
result of their potential for airway obstruction or fatal hemoptysis. In the case reported here, PCHs invaded the large airway. If the lesions break through the mucous layer, there will be hemoptysis, even leading to lethal blood aspiration [1]. In general, PCHs seldom regress, nor do they rapidly progress. But in our case, the lesions indeed changed slightly in size and number, and the pleural effusion was spontaneously absorbed. However, the lesions may enlarge and hemothorax may appear again. The clinical course should be carefully followed up.

The diagnosis of PCHs is not usually considered because of its low frequency and the lack of specific symptoms. Most PCHs are identified on routine chest roentgenography. Contrast-enhanced CT of the chest is helpful for diagnosis, demonstrating single or multiple pulmonary nodules without prominent infiltration, and some cases may reveal draining veins of the lesions. Spread lesion calcifications may be found, and they are attributed to multiple phleboliths inside the dilated vascular spaces. These radiographic features are nonspecific but may arouse suspicion of PCH. More extensive investigation should be done, confirming the diagnosis and assessing the extent of the lesions throughout the body.

The definitive diagnosis of PCH can be made by histopathologic examination. Cavernous hemangiomas are characterized by large, dilated vascular spaces interposed with various stromal elements such as fat, myxoid fibroblastic proliferation, and fibrous tissue. The abnormal vessel agglomeration is sharply defined but not encapsulated; it may be accompanied by intravascular thrombosis or dystrophic calcification [3]. When cavernous hemangioma is suspected, it is necessary to determine whether the lesion is associated with endothelium, which can be achieved by immunohistologic staining of the cells lining the cavernous lumen.

The treatment for cavernous hemangiomas include pharmacologic therapy, operation, sclerotherapy, laser therapy, cryotherapy, electrocoagulation, and treatment with copper needles, but most of them are not applied in PCH. The management of PCH varies depending on the severity of symptoms and the location and extent of the lesions. A solitary PCH should be treated with surgical excision; the impossibility of ruling out malignancy is the more common criteria for surgical resection. For multiple PCHs, local resection is rarely used; surgical procedure usually is just for pathologic diagnosis. Endoscopic treatment is suitable for airway PCH. Glade and colleagues [5] reported endoscopic management of venous malformations using a neodymium-doped yttrium aluminum garnet laser, which appeared to be both effective and relatively safe. Embolization may be a good method for some vascular malformations, just as with pulmonary arteriovenous fistula, although PCHs are a variant of slow-flow pulmonary venous malformations and it is difficult to locate the target vessels.

However, there are some reports about PCH with durable results without intervention. A 29-year-old woman had an abnormal shadow on chest roentgenogram for 20 years before the operation revealed a PCH [6]. In a case of multiple PCHs, a 61-year-old woman with multiple shadows on chest radiographs for 10 years before her death of congestive heart failure [4]. Xavier and Emil [7] presented 2 patients whose vascular malformations
spontaneously resolved. These experiences highlight that radiographic follow-up may play a significant role in asymptomatic and stable PCH.

In summary, PCHs are extremely rare. They can be asymptomatic or can present with life-threatening symptoms. The clinical features are not well revealed. For a solitary PCH, surgical excision yields excellent outcomes, but for multiple PCHs, surgical intervention or close radiographic follow-up remains the treatment of choice.

References

Successful Lung Transplantation After Donor Lung Reconditioning With Urokinase in Ex Vivo Lung Perfusion System
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Acute pulmonary embolism is considered a contraindication to lung donation for transplantation as it might result in graft dysfunction. Ex vivo lung perfusion (EVLP) is a novel method to assess and recondition a questionable donor graft before transplantation. In this report we present a case of successful bilateral lung transplant after donor lung assessment and treatment with a fibrinolytic agent, urokinase, during EVLP.


Table 1. Parameters During Ex Vivo Lung Perfusion

<table>
<thead>
<tr>
<th>Time (hour)</th>
<th>PVR</th>
<th>C-Dyn</th>
<th>Delta P O2</th>
<th>PiP</th>
<th>Lactate</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4.9</td>
<td>72</td>
<td>278</td>
<td>14</td>
<td>1.4</td>
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<tr>
<td>2</td>
<td>4.9</td>
<td>73</td>
<td>322</td>
<td>15</td>
<td>2.5</td>
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<tr>
<td>3</td>
<td>3.7</td>
<td>74</td>
<td>280</td>
<td>15</td>
<td>3.1</td>
</tr>
</tbody>
</table>

C-Dyn = dynamic lung compliance (mL/cm H2O); PiP = peak inspiratory pressure (cm H2O); PO2 = partial pressure of oxygen; PVR = pulmonary vascular resistance (Wood Units).

Despite improvements in both donor management and organ preservation, only a limited number of potential donor lungs are considered acceptable for transplantation. This leads to high waiting list mortality for lung transplant recipients. Ex vivo lung perfusion (EVLP) is a novel technique proposed for the assessment, resuscitation, and repair of extended donor lungs [1, 2]. Similar early outcomes have been reported in recipients who underwent transplantation after EVLP to those with conventionally selected and transplanted lungs [1–3]. We report successful bilateral lung transplantation after donor lung treatment and assessment in the EVLP system with a fibrinolytic agent, urokinase.

Recipient was a 27-year-old male suffering with end-stage cystic fibrosis (176 cm, 75 kg, body mass index 24 kg/m2), diagnosed in childhood due to recurrent respiratory symptoms and found to have a delta F508 homozygous cystic fibrosis genotype. Forced expiratory volume in 1 second (FEV1) and forced vital capacity were 1.7 l, 40% predicted and 3.38 l, 64% predicted, respectively. Informed consent was obtained from the patient.

The donor was a 45-year-old female (180 cm, 98 kg) who was hospitalized for spine surgery. At postoperative day 4 she had suffered diffuse arterial and venous emboli. She underwent femoral arterial embolectomy and received intravenous lytic treatment for pulmonary emboli. Thereafter she developed intracranial bleeding resulting in brain death. She was declared brain dead after developing intracranial bleeding. Her arterial oxygen partial pressure at the time of retrieval was 360 mm Hg. Chest X-ray revealed bilateral infiltrations. Thorax computed tomography (CT) scan could not rule out persistent segmental arterial lung emboli. The lung was accepted for EVLP and reconditioning during EVLP with urokinase. Acellular normothermic EVLP [1] was performed.

At 37°C 100,000 IU urokinase was added into the reservoir. During EVLP for the functional assessment, tidal volume was set at 10 mL per kilogram of donor body weight and 10 breaths per minute, with fraction of inspired oxygen at 1.0. Lung function was evaluated hourly during EVLP according to the following calculations: Delta PO2 (partial pressure of oxygen) = [left atrial PO2 – pulmonary-artery PO2 (in mm Hg)], and pulmonary vascular resistance = [(pulmonary-artery pressure – left atrial pressure)] ÷ pulmonary-artery flow (in Wood Units), dynamic compliance (in mL/cm H2O), and peak inspiratory pressure (in cm H2O). The lung was functionally evaluated for 3 hours and then cooled down to...