Korean J Thorac Cardiovasc Surg 2011;44:373-376 □ Case Report □
ISSN: 2233-601X (Print) ISSN: 2093-6516 (Online) http://dx.doi.org/10.5090/kjtcs.2011.44.5.373

Bilateral Mediastinal Lymphangiohemangiomas Containing Anomalous Venous Components

— A case report —

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Lymphangiohemangiomas of the mediastinum are exceedingly rare and few cases have been published in the English literature. This report may be the only reported case in which lymphangiohemangiomas were found bilaterally. We report a case of a 7-year-old boy with an incidental finding of an abnormal mediastinal shadow on a chest X-ray. The chest CT showed a large mass in the left superior mediastinum and another in the right posterior mediastinum. The left mass had anomalous venous channels connected to the left innominate vein, and the right mass to the left atrium. We performed an excision of the mass in the left side first and then the right side one month later. Anomalous venous channels were dissected carefully and ligated. There were no complications and no signs of recurrence 30 months after the operation.

Key words: 1. Neoplasms
2. Lymphangioma
3. Hemangioma

CASE REPORT

A 7-year-old boy presented with coughing for a week. Breathing sounds were normal and the heart beat was regular without murmur. Laboratory findings including tumor markers were within normal limits. Chest PA showed superior mediastinal widening and bulging of the right paraspinal interface (Fig. 1). Increased opacity of the infrahilar window and thickening of the posterior wall of the bronchus intermedius were seen on the lateral chest X-ray.

CT scan was performed for further characterization of the lesion. An oval-shaped mass measuring 9.7×3.8×6.0 cm was seen in the left superior mediastinum. The lesion showed diffuse low attenuation without enhancement. A bizarre-shaped vascular structure that was connected to left innominate vein was seen in central area of the mass (Fig. 2). The mass showed insinuating appearance without significant compression of adjacent structures. Only the thymus was slightly compressed by the mass. Another similarly natured mass was seen in the right paraspinal area, which measured about 4.2×3.0×6.6 cm. This mass also contained a bizarre-shaped vascular structure that communicated with the left atrium directly through an anomalous vascular structure (Fig. 2). Multiplanar reconstruction (MPR) and 3D reformation showed that the contrast directly entered the vascular components of the masses via an abnormal vein originating from the left in-

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Received: April 14, 2011, Revised: May 30, 2011, Accepted: July 3, 2011
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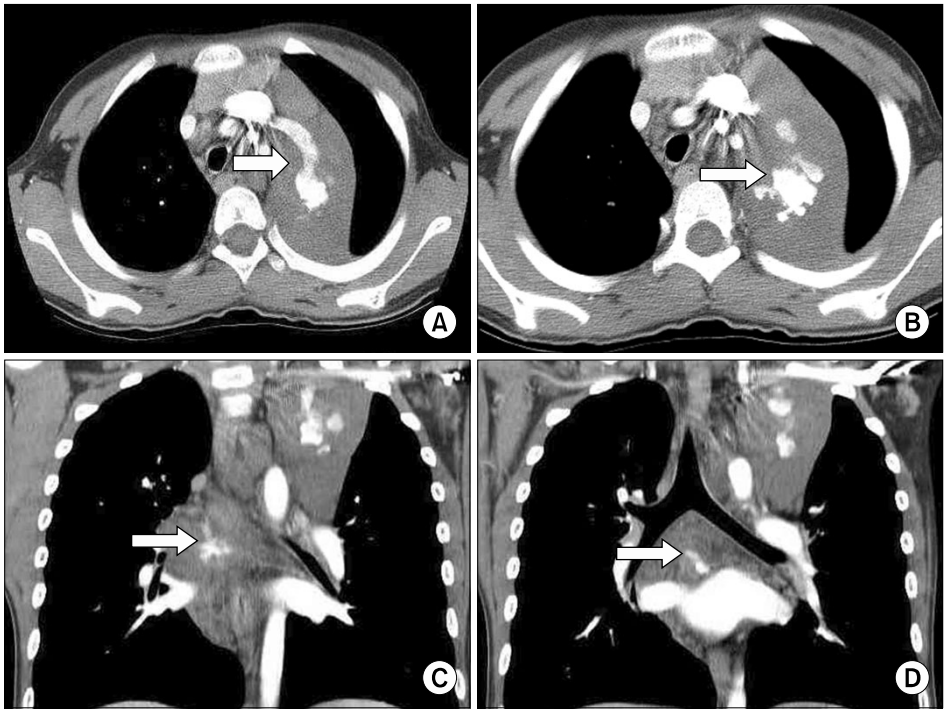

Fig. 2. (A, B) Abnormal venous channel to the left innominate vein in the left mass. (C, D) Abnormal venous channel to the left atrium in the right mass.

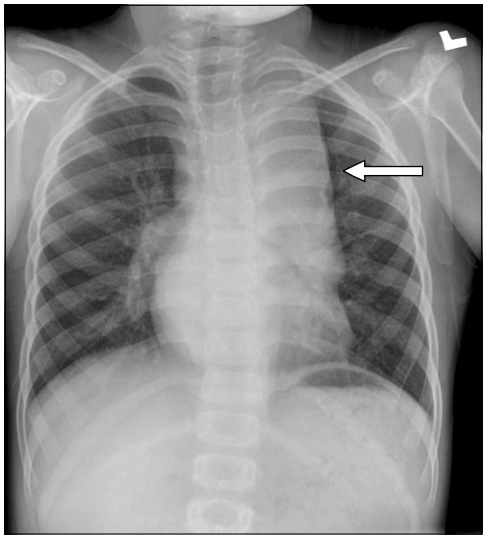

Fig. 1. Preoperative chest PA shows an abnormal left mediastinal shadow.

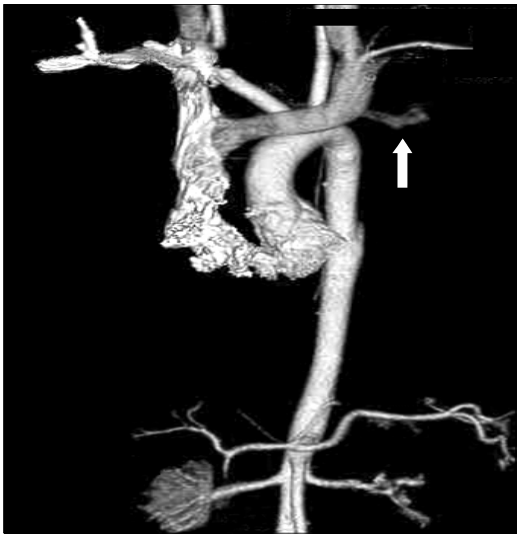

Fig. 3. Multiplanar reconstruction (MPR) and 3D reformation showed that the contrast directly entered the vascular components of the masses via the abnormal vein originating from the left innominate vein.

nominate vein (Fig. 3). The mediastinal window setting of contrast-enhanced CT showed additional multiple small cystic lesions in the middle mediastinum.

A two-stage operation was planned due to the fact that the tumors were located bilaterally. First, for the left side tumor resection, posterolateral thoracotomy was performed and a large cystic mass was found in the anterosuperior mediastinum. The content of the cyst was bloody and a large communicating vein to left innominate vein and few small communicating veins to left intercostal veins were found, which

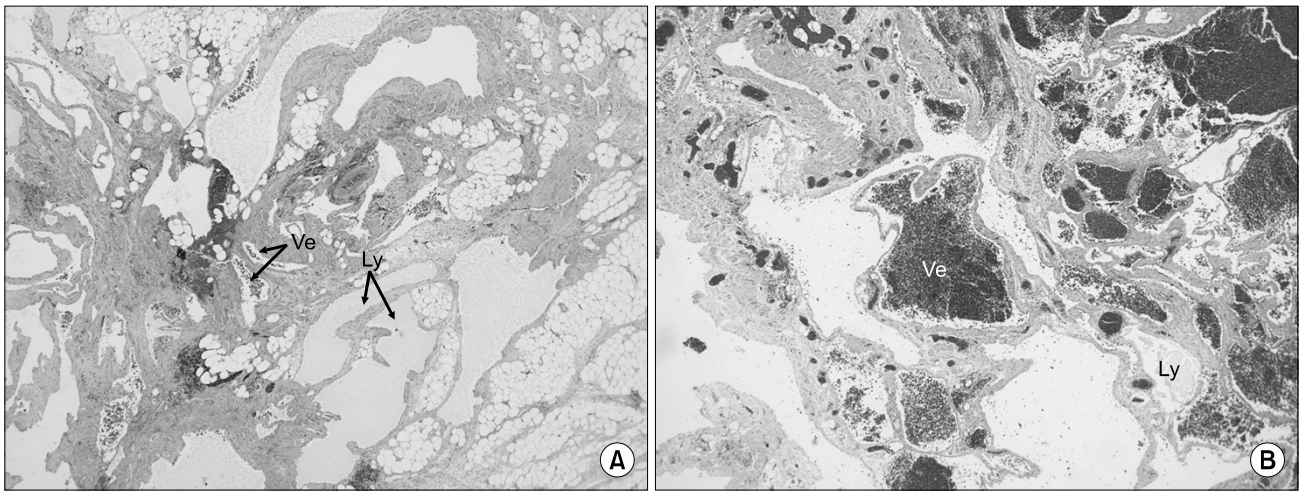

Fig. 5. Microphotograph showing combined proliferation of venous vascular channels (Ve) and lymphatic vessels (Ly) (H&E stain, ×40).

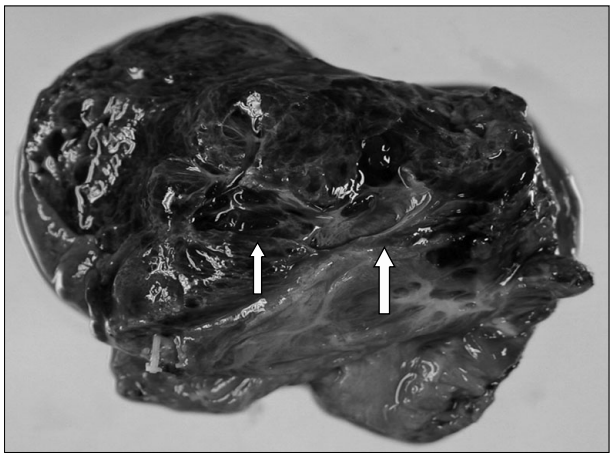

Fig. 4. Cut surface of the left mediastinal mass reveals multiple cystic spaces containing bloody contents.

were then ligated and divided. The left phrenic nerve was passing through the mass and had to be sacrificed. Four other small cysts which contained serous fluid were resected. The second operation was performed for the right mediastinal mass 45 days after the first surgery. The right paraspinal mass was successfully resected and an anomalous communicating vein to left atrium was identified and divided. The patient recovered completely without any complications such as chylothorax or hemothorax.

On gross pathologic examination, the cut surface of the tumor revealed numerous cystic spaces and a cavernous sinus containing bloody contents (Fig. 4). Histology of the mass showed combined proliferation of venous vascular channels (Ve) and lymphatic channels (Ly) that exhibited infiltration into mediastinal fat pad (Fig. 5). The final pathologic diagnosis was made as lymphangiohemangioma for both masses.

DISCUSSION

Mediastinal lymphangiohemangioma is a very rare venolymphatic malformation containing both vascular and lymphatic elements [1,2]. Direct vascular communication between the lesion and the vena cava system have rarely been reported [1,2]. CT images of the mediastinal lymphangiohemangioma are very rarely found in the English literature [1-4]. Moreover, neither a case with multiple masses and more than one vascular communication nor a case directly connected to the heart chamber through an anomalous vessel was found in the literature. We report a case of surgicopathologically confirmed mediastinal lymphangiohemangioma that communicated with the left innominate vein and left atrium directly via anomalous vascular structures (Fig. 2).

The nomenclature and classification of vascular malformations and hemangiomas are confusing to most physicians. Mulliken and Glowacki [5] suggested a categorical scheme for these lesions in 1982. They separated endothelial malformations into two large groups: hemangiomas and vascular malformations. Hemangiomas exhibit cellular proliferation and tend to involute during childhood. In contrast, vascular mal-

formations are comprised of dysplastic vessels and do not regress.

Vascular malformations are subcategorized as lymphatic, capillary, venous, arteriovenous, and mixed malformations on the basis of their histologic nature. They are classified as either low-flow or high-flow lesions. High-flow lesions have arterial components and low-flow lesions do not. Therefore, our case can be categorized as low-flow vascular malformations with mixed lymphatic and venous components. MRI may play a role in differentiating the types. Slow-flowing blood appears as high signal intensity on T2-weighted images and as intermediate signal intensity on T1-weighted images [6].

Most lesions present early in childhood and the most common locations for lymphatic malformations are the neck and axilla. A less common location is the mediastinum, and the anterior mediastinum is the most commonly found mediastinal site.

The diagnosis can be made by characteristic CT and pathologic findings. Pathologic examination confirmed the lymphatic and venous components (Fig. 5). On noncontrast CT scan, the mass showed diffuse low attenuation without calcification, although previous reports showed scattered phleboliths in the mass, which represent calcified emboli in the venous channels [1,2]. The anomalous vascular structures were enhanced at the same intensity as the systemic venous system and was connected directly to the left innominate vein and left atrium. The peripheral portion of the lesion was not enhanced on all phases of dynamic CT scan, which may be related to the lymphatic components or fibroconnective tissues of the mass. A previous report postulated that the anomalous vein acts not only as a draining vessel but also as a feeding vessel [1]. In our case, only the proximal part of the vascular structure was enhanced on the pulmonary arterial phase, whereas all of the large bizarre-shaped vascular spaces were enhanced on the systemic venous phase (Fig. 2). A delayed CT scan may show delayed clearance of contrast within the anomalous venous channels because of the lack of smooth muscle in these channels [1]. Lymphangiohemangiomas are not considered an congenital anomaly but are frequently associated with other vascular malformations, most commonly in the left superior vena cava [2]. In our case, the anomalous vessels connected to the left innominate vein may be considered a left superior vena cava remnant.

A lymphangiohemangioma should be included in the list of differential diagnoses of an incidentally found low-attenuating mediastinal mass, especially in young patients. Enhancing venous channels on the venous phase are characteristic CT features of a lymphangiohemangioma.

The primary treatment is surgical excision in infancy [6]. Another therapeutic option is percutaneous sclerotherapy using agents such as absolute ethanol, bleomycin, cyclophosphamide, doxycycline, alcohol solution of zein, or OK-432 [6]. Recognizing that the lesion is a low-flow vascular malformation is more important than determining whether the lesion is predominantly venous or lymphatic when making treatment decisions [6].

We planned surgical excision of the lesions and successfully excised them without any complications such as hemothorax or chylothorax. For successful surgery, the anomalous vascular pedicle of the mass communicating with the systemic vein or heart chamber should be carefully identified and divided to prevent massive hemothorax.

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