Case Report

A Case of Mediastinal Cystic Lymphangioma

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A 35-year-old Japanese man’s routine chest radiography revealed an abnormal opacity. Chest computed tomography and magnetic resonance imaging showed a 5.5 cm in dia. cystic tumor located at the left anterior mediastinum. The tumor was suspected to be an asymptomatic thymic cyst, and we chose observation for the tumor. At the 3-year follow up, the cystic tumor had gradually enlarged to 7.5 cm in dia. and we thus performed a surgical resection via left video-assisted thoracic surgery. An immunohistochemical analysis showed that the cystic tumor was not a thymic cyst but rather a mediastinal cystic lymphangioma. Mediastinal cystic lymphangiomas are very rare, and they are difficult to diagnose preoperatively. Complete surgical resection is proposed for the treatment of such tumors.

Key words: mediastinal tumor, mediastinal cystic lymphangioma, thymic cyst

Lymphangiomas are rare benign congenital malformations derived from the lymphatic system. Most appear in the neck or axillae, and approximately 1% extend into the mediastinum [1]; these mediastinal cystic lymphangiomas account for 0.7−4.5% of all mediastinal tumors [2]. We encountered a rare case of mediastinal cystic lymphangioma that was diagnosed postoperatively.

Case

A 35-year-old Japanese man underwent chest radiography as part of a routine medical examination, and the radiography revealed an abnormal opacity. A further examination was undertaken at the local hospital. Chest computed tomography (CT) revealed a left anterior mediastinal tumor, and the patient was consequently referred to our hospital. He was on medication for diabetes mellitus.

Chest CT showed a cystic tumor, 5.5 cm in dia., with a smooth surface, round shape, and low attenuation area (Fig. 1). The cystic nature was confirmed upon magnetic resonance imaging (MRI), which showed a high-intensity area on T2-weighted images (Fig. 2). We highly suspected an asymptomatic thymic cyst, and we thus chose observation for the cystic tumor. After 3 years of follow-up, the cystic tumor had gradually enlarged to 7.5 cm in dia., at which time we performed a surgical resection. With the patient under general anesthesia in the right hemi-lateral decubitus position, the tumor resection was performed via left video-assisted thoracic surgery (VATS) using one access window plus two ports.

The resected tumor was soft and had serous effusion that was aspirated during the operation to allow for easy dissection. No obvious communication was found between the tumor and thoracic duct. The cystic tumor was not adhered to the left lung and could be separated from the pericardium by blunt dissection.

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(Fig. 3). The left phrenic nerve was preserved. The duration of the operation was 105 min, and the intra-operative bleeding was 30 mL. No major complications were encountered, and the patient was discharged on postoperative day 5.

The histopathological examination showed that the cystic lesion consisted of several separate parts, and neither Hassall bodies nor thymic epithelial cells were observed. No lining epithelium was observed, whereas fibrous tissue and smooth muscle proliferation were noted in the cyst walls (Fig. 4A). An immunohistochemical analysis showed consistently positive staining for D2-40 (Fig. 4B) and negative staining for

Fig. 3 Intra-operative photo with macroscopic findings of the mediastinal cystic lymphangioma.

Fig. 4 Histopathological examination showed fibrous tissue and smooth muscle proliferation in the cyst walls. Neither Hassall bodies nor thymic epithelial cells were observed. The immunohistochemical analysis showed consistently positive staining for D2-40.
cytokeratin AE1/3 in the endothelium of the lymph vessels. The histological diagnosis was thus mediastinal cystic lymphangioma.

At the latest follow-up (6 months after the surgery), the patient was doing well.

Discussion

Histologically, Lymphangiomas can be subdivided into 3 types depending on the size of the lymphatic channels: cystic (macro-cystic), simple (super micro-cystic), and cavernous (micro-cystic) types [3]. The classification system for vascular anomalies was expanded at the 2014 International Society for the Study of Vascular Anomalies (ISSVA) workshop in Melbourne [4]. Our patient’s case would be defined as macrocystic lymphatic malformation in the ISSVA classification.

The cystic type of lymphangioma is the most common, and the other 2 types are very rare [5, 6]. These tumors are usually found incidentally, and they are sometimes associated with dysphagia, dyspnea, cough, or chest pain.

Both CT and MRI are useful for the detection of mediastinal lymphangiomas. The most common CT appearance is a smoothly margined cystic mass. Unusual features include calcification, speculated margins, and homogeneous soft-tissue attenuation. MRI shows cystic components with heterogeneous signal intensity on T1-weighted images and high signal intensity on T2-weighted images [7]. However, the diagnosis cannot be made solely on the basis of radiologic studies, and mediastinal lymphangiomas are usually diagnosed after the histological examination of surgically resected specimens.

Mediastinal lymphangioma are usually multi-cystic lesions, and thymic cysts are usually a simple cystic lesion. In our patient’s case, the chest CT and MRI showed a cystic lesion, and before his surgery the diagnosis of mediastinal lymphangioma seemed unlikely. Surgical resection is traditionally performed via a thoracotomy, although, recently, several reports have advocated VATS as was used in our case [8, 9]. Based on the present case, we suggest performing VATS if possible when mediastinal cystic tumors are suspected. In terms of the treatment, complete surgical resection is generally recommended, as incomplete resection can result in recurrence of the tumor [10].

The histological appearance along with immunocytochemical staining for CD31 has been reported to confirm the diagnosis of mediastinal cystic tumors [8]. In our patient’s case, the definitive diagnosis was made on the basis of hematoxylin and eosin staining, together with positive D2–40 staining of the endothelium of the lymph vessels, and positive cytokeratin AE1/3 staining of the epithelium.

In conclusion, we encountered a rare case of mediastinal cystic lymphangioma in an adult man. The tumor was difficult to diagnose before the surgery, and complete surgical resection is proposed for therapeutic purposes.

References