Cystic Lymphangioma of the Chest Wall: A Case Report

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Key Words
Cystic lymphangioma · Chest wall · Surgical resection

Abstract
Cystic lymphangioma is a congenital cystic malformation that occurs mostly in children. While it is mainly found in the cervix, cases in the chest wall are very rare. We report a case of cystic lymphangioma found in the chest wall of a 2-month-old girl. The patient was noted to have a tumor in the left chest wall at birth. Since it gradually increased in size, the patient was referred to our department. Transillumination and ultrasonography showed a cystic lesion in the left chest. Surgical resection was performed in one step. Histopathological examination showed a cystic lymphangioma.

Introduction
Cystic lymphangioma is a congenital malformation that results from failure of a primary lymphatic sac to establish drainage into the venous system. Isolated cystic lymphangioma of the chest wall is a rare condition. Cystic lymphangiomas grow very slowly, usually located to one organ. Transillumination can help differentiate cystic lymphangioma from solid tumors. Ultrasonography shows a multilobular cystic mass that contains a septum. Several treatments of cystic lymphangioma have been reported. We present a rare patient with lymphangioma in the chest wall, who underwent surgical resection.
Case Report

A 2-month-old girl was noted to have a tumor in the left chest at birth. The tumor gradually increased in size and the patient was referred to our hospital. No physical abnormalities had been detected at birth. At the time of hospitalization the patient’s height was 57.5 cm and weight was 5.8 kg. Blood pressure, breath sounds, heart sounds and blood examinations were normal. Macroscopically, the tumor measured 4 × 5 × 1 cm. On palpation, it consisted of two soft tumors. Chest ultrasonography revealed three well-demarcated tumors with an internal hypoechoic pattern. Doppler echo showed no blood flow (fig. 1).

At surgery, a skin incision was made to the surface of the tumor and a skin flap was made. We excised part of the muscle and extracted the tumor (fig. 2), as part of the undersurface of the tumor had adhered to the serratus anterior muscle. The tumor contained lymphatic fluid.

Histopathological examination revealed that the tumor was covered by flat endothelial cells followed by multiple cystic spaces. There were muscle and irregular elastic fibers in the wall, and they were irregular compared with the structure of vessels (fig. 3). The content of the tumor was pellucid and colorless, and there were lymphoid cells. It was diagnosed as a cystic lymphangioma of the chest wall.

The patient’s postoperative condition was good, without complications. On follow-up, she revealed no evidence of recurrence and had normal growth and development.

Discussion

Cystic lymphangioma is a rare disease considered as a congenital malformation of lymphoid tissue. Cystic formation is induced when primary lymphatic spaces fail to join the central system. The lack of communication between a lymphatic sac and the venous system produces a cystic lymphangioma [1]. The incidence of the disease is approximately 1 in 12,000 births and accounts for 5–6% of all pediatric neoplasms [2]. Approximately, 65% of cystic lymphangiomas are apparent at birth, and 90% appear at the end of the second year [1, 3]. About 75% of cases occur in the neck, 20% in the axilla and 5% in various parts of the body including the mediastinum, retroperitoneum, pelvis and groin [4, 5]. Chest wall lymphangioma is a rare condition [1, 5]. Hancock et al. [6] reported the distribution of lymphangiomas to be cervical (31.4%), craniofacial (18.9%), truncal (9.2%) and cervico-axillothoracic (4.9%). Furthermore, Suzuki et al. [7] and Ardenghy et al. [8] reported that the incidence of cystic lymphangioma of the chest wall was low. The reason for this very rare localization can be explained embryologically. Pokorny [9] reported that the final position of the cystic lymphangioma depends on the structure to which the sequestrated sac is attached.

The diagnosis of cystic lymphangioma is made by physical examination, transillumination and ultrasonography. Clinically, cystic lymphangioma occurs as a large, soft, cystic mass. Transillumination can help differentiate cystic lymphangiomas from solid tumors. Ultrasonography shows a characteristic multilobular cystic mass that contains a septum of variable thickness. Doppler ultrasonography should be done to detect any vascular component of the cystic lymphangioma. In our case the diagnosis was confirmed by histopathological examination.

Surgical resection is recommended principally. In general, since in many cases there is lymphatic tissue showing cavernous structure in the surroundings of the cystic lymphangioma, it is important to perform complete resection including the circumferential tissues in order to prevent recurrence. Ward et al. [10] reported that recurrence after excision oc-
curred when the surgical resection was performed incompletely. Flanagan and Helwig [11] reported that recurrence often occurred within 3 months after excision. Since an exceptional recurrence appeared 7 years after excision [11], long-term follow-up is necessary.

Other types of adjuvant treatment have been proposed, such as radiotherapy, injection of sclerosing agents via 100% ethanol or OKT-432, but they are controversial [12, 13]. Systemic chemotherapy and interferon-α have been tried with limited success for patients with extensive inoperable lesions [14, 15]. Reports of malignant alteration are unfounded.

When the lesion has been completely excised, it is thought that the prognosis is good. We presented a rare case of cystic lymphangioma in the chest wall that was treated with surgical resection.

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**Fig. 1.** Chest ultrasonography showing three tumors with an internal hypoechoic pattern.

**Fig. 2.** Specimen of cystic lymphangioma showing a mass with a clear septum (arrowheads). The tumor content was lymphatic fluid.
Fig. 3. Histopathological examination showed the tumor to be lined by a single epithelium with irregular elastic fibers in the wall. H&E, original magnification ×50.