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## Case report

# Macrocystic lymphatic malformation of the chest wall and axilla: A case report in a 45-year-old man ☆☆☆

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## ABSTRACT

Lymphatic malformations (LMs) are rare, with more than 90% occurring during childhood. Most LMs are located in the head, neck and axilla. LMs in chest wall are extremely rare, particularly in adults. This report describes a 45-year-old man with a large macrocystic LM in the right anterior chest wall. Computed tomography showed a ~15 cm sized, well-defined, homogeneous and hypoattenuated mass without enhancement in the right anterior chest wall. On ultrasonography (US), the mass was circumscribed and anechoic, with internal septations and posterior acoustic enhancement. Following surgical excision, the mass was diagnosed as a macrocystic LM.

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## Introduction

Lymphatic malformations (LMs), formally called lymphangiomas, are uncommon congenital anomalies of the lymphatic vessels [1]. LMs occur mainly in children, usually between birth and age 2 years. There are three major subtypes of cystic LM, macrocystic, microcystic, and mixed cystic LMs. Up to 95% of LMs are located in the cervical region and axilla, but can also occur elsewhere in the body, including the mediastinum, retroperitoneum, and groin, but not in the brain. The chest wall is an extremely rare location of LMs and is especially rare among adults [2]. To our knowledge, there have

been only two reported cases of chest wall LMs in adults [3,4]. The present report described a rare case of large macrocystic LM in a 45-year-old man, including imaging and pathological findings.

## Case report

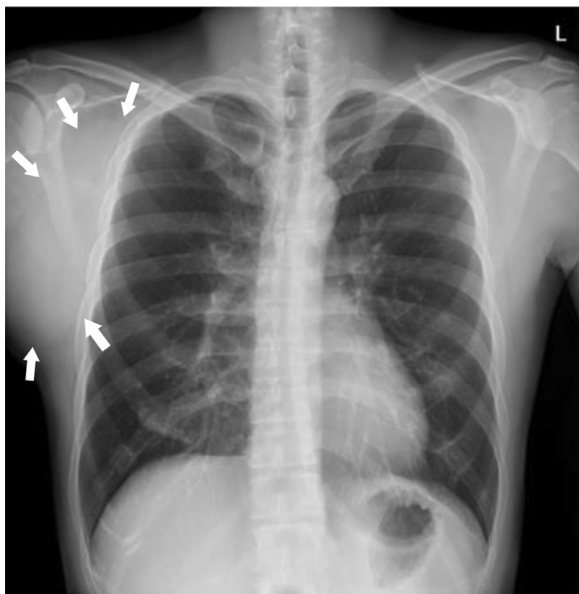
A 45-year-old man presented with a growing palpable mass in the right chest wall of 5 months' duration. The patient was not taking any medication and had no history of congenital anomaly, trauma, or previous surgery. Physical examination

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**Fig. 1 – Chest radiograph of a 45-y-old man with a growing palpable mass in the right anterior chest wall, shown to be a macrocystic LM. The white arrows indicate the large mass-like opacity in the right chest wall.**

revealed a large, soft and nontender mass in the right anterior chest wall. Laboratory findings, including complete blood count, were normal.

A chest radiograph revealed an obvious mass-like opacity in the right chest wall (Fig. 1). Chest wall ultrasound (US; IU22 unit; Philips Medical System, Bothell, WA) using a linear transducer with a frequency of 8–15 MHz detected a huge, well-circumscribed and anechoic mass with thin septation and posterior acoustic enhancement in the right anterior chest wall. Color Doppler US showed no internal vascularity within the mass (Fig. 2). Chest computed tomography (CT; Somatome Sensation 64, Siemens, Forchheim, Germany) was performed

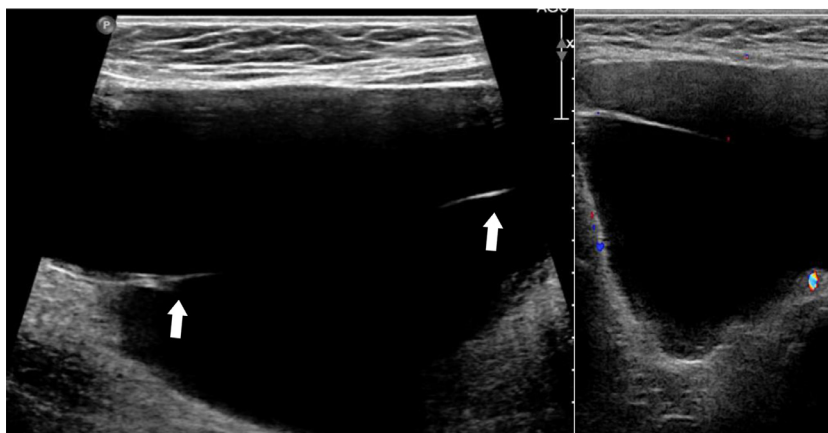
to evaluate the relationship between the mass and adjacent structures. Axial and coronal CT images revealed a 15 cm x 11 cm sized, thin-walled, homogeneously low-attenuated mass lesion without definite enhancing portion in the right anterior chest wall, extending from beneath the pectoralis minor and latissimus dorsi muscles to the apex of the axilla (Fig. 3). The lesion extended to the ribs of the right lateral chest wall but there was no intra-thoracic or intra-abdominal extension.

Because the mass was huge and had grown larger over several months, it was surgically excised without trying other treatment options such as aspiration or sclerotherapy. During the surgery, a lump containing the mass was removed from the left pectoralis major muscle while conserving the left axillary vein and the thoracodorsal and long thoracic nerves. Histopathological examination showed a large cyst lined with a thin endothelium layer and surrounding vascular channels, consistent with an LM (Fig. 4).

## Discussion

LMs are benign tumors of the lymphatic system, with the formation of cystic LM induced when primary lymphatic spaces fail to join the central system [1]. Although LMs have also been described using several other terms, including cystic hygroma, cavernous lymphangioma, cystic lymphangioma, and lymphangioma circumscriptum, they are currently grouped as LM according to the 2018 ISSVA classification.

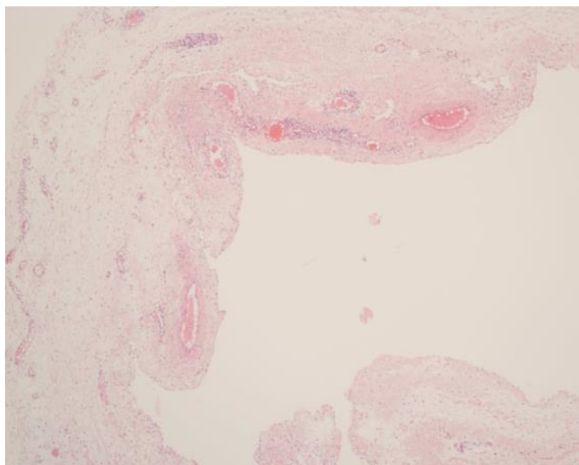
The reported incidence of LM is approximately 2.8–5 of 100,000 people [5]. LMs are usually evident prior to age 2 years and are very rare in adults. About 75% of LMs occur in the neck and 20% in the axilla, with less frequent occurrences in the shoulder, breast, mediastinum, intra-abdominal region, retroperitoneum, and pelvis [6]. The chest wall is an exceptionally rare location, with only two adults with LM in the chest wall previously reported [3,4]. These two patients were men aged 64 and 35 years, whereas the present patient was a 45-year-old man.



**Fig. 2 – Transverse (A) and color Doppler (B) ultrasonography showing a huge, circumscribed and anechoic mass with thin septation (white arrows in A) in the right anterior chest wall. Posterior acoustic enhancement without internal vascularity was observed.**



**Fig. 3 – Axial (A) and coronal (B) chest CT scans, showing a well-defined, homogeneous and low-attenuated mass without enhancement in the right anterior chest wall.**



**Fig. 4 – Hematoxylin and eosin staining of the resected specimen, showing LM lined by flat endothelial cells and composed of vascular channels of varying sizes interspersed with fibroadipose tissue (magnification, 40x).**

The exact nature of LM remains unclear, although pediatric LMs are thought to be caused by chromosomal abnormalities, including trisomy 21 and Turner syndrome [7]. LMs have been shown to be positive for an activating mutation in the PIK3CA gene, a somatic (non-inherited) mutation in lymphatic endothelial cells [8]. Predisposing factors in adults include trauma, infection, and tumor growth [6], but none of these factors was present in our patient. The cause of LM in our patient is unclear, as in the two other adults with LM in the chest wall.

Cystic LMs can be classified into three subtypes. The first, or macrocystic type (previously called cystic hygromas), is characterized by cysts larger than 2 cm with clear boundaries; the second, or microcystic type, is characterized by cysts smaller than 2 cm without clear boundaries; the third, or mixed cystic type, consists of a combination of macro- and microcystic types. The lesion in our patient consisted of a large, thin-walled cyst with thin septation on multiplanar images,

with pathologic examination confirming its identity as an LM, most likely the macrocystic type. A study of 40 children with cervicofacial LM showed a nearly equivalent incidence of each subtype [5]. However, the prevalence of each subtype in adults is unknown due to its rarity.

The clinical presentation of LM is variable and is dependent on many factors, including subtype, location, size, relationship to surrounding structures, and the presence of infection. Adults with thoracic LM can present with chest pain, cough or dyspnea [9]. By contrast, all three adults with chest wall LM, including our patient, presented with an enlarging painless lump. On physical examination, LMs are usually transilluminated and have an intact overlying skin without pulsatility or bruit.

Image findings contribute to the diagnosis of chest wall LM. On US, LMs usually appear as well-defined hypo- or anechoic trans-spatial masses and can be unilocular, multilocular, or septated masses. LMs may also possess internal echogenic components, indicative of lymphatic channels or internal hemorrhage [10]. Findings of LM on CT images parallel those on US, with CT showing a unilocular or multilocular low-attenuated cystic mass without enhancement. These masses may also show fluid-fluid level or calcification and may include cystic components mixed with other tissue types [9]. MRI can be the most informative imaging modality, as LMs generally show low intensity on T1 and high intensity on T2 images. Additionally, MRI often shows enhancement of the septa, with the mass often appearing multiseptated [2]. The LM in our patient appeared typical on both US and CT, presenting as a large unilocular cystic mass with thin walls and thin septation without an enhancing portion.

CT may be required to determine the possibility of intrathoracic extension of the LM and its affinity to adjacent anatomic structures. In particular, CT may be crucial for determining a surgical plan, as it can highlight the affinity of the LM with adjacent structures [4,9]. CT in our patient was helpful in evaluating the anatomical structures adjacent to the large chest wall LM.

The diagnosis of chest wall LM may be obvious in patients with typical findings, such as a large, smooth and cystic nontender mass in the chest wall. Due to their rarity, however, LMs

in the chest wall may be misdiagnosed. LMs vary widely in appearance; for example, they may not contain detectable cystic components or may have spiculated margin or internal calcifications. Other cystic masses should be considered in differential diagnosis, including necrotic tumors, hematomas, and infectious fluid collections [9].

Although the most effective treatment of LM is resection, incomplete resection has been associated with recurrence [4]. Other therapeutic options include aspiration and drainage of the cysts for emergency decompression; injection of sclerosing agents, such as bleomycin or OK-432; and low-dose radiation therapy [3].

In conclusion, the patient described in this report is of particular interest due to the low incidence of LM in adults, the rarity of LMs in the chest wall, and the large size of the lesion. Although rare, it is important to consider LM in the differential diagnosis of patients with chest wall tumors. LM can be easily distinguished from other tumor types by imaging methods, such as US and CT. In addition to their use in diagnosis, multiplanar imaging methods can help plan the optimal surgical approach.

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### Patient consent

Institutional Review Board approval was obtained for this retrospective study, and the requirement for informed consent was waived.

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