Diagnostic imaging of lymphatic malformations

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Learning objectives

- To describe the characteristic features of lymphatic malformations (LM) in both common and uncommon locations.
- To illustrate the imaging findings in LM and their differential diagnosis, with special focus on computed tomography (CT) and ultrasonography.
Background

LM are benign congenital lesions which consist of abnormally developed dilated lymphatic vessels caused by blockage of lymphatic flow. Histopathologically, LM are characterized by thin-walled unilocular or multilocular cysts that are lined by endothelial cells and contain clear or milky fluid. The wall contains varying amounts of connective tissue, small lymphatic spaces and smooth muscle fibers [1,2]. In 50%-60% of patients, these malformations are present at birth and they have no predilection for either sex or race. Frequently, LM clinically manifests as an asymptomatic soft mass. Although these lesions tend to surround and sometimes invade adjacent anatomic structures, they have no malignant potential. However, surgical excision is the treatment of choice. [3]

Also termed as lymphangiomas, these lesions can be divided into microcystic LM (lymphangioma circumsctriptum, cavernous lymphangioma), macrocystic LM (cystic hygromas) and combined forms. The 3 different histologic subtypes are determined by the size of the lymphatic channels.

**Lymphangioma circumsctriptum** is the common form of cutaneous lymphangioma and is characterized by clusters of multiple small cysts that usually contain clear fluid.

**Cavernous lymphangiomas** contain larger lymphatic channels and lymphoid stroma and are connected to normal adjacent lymphatic tissue; they are most commonly found in the skin or the subcutaneous tissue and can infiltrate surrounding tissue [4].

**Cystic lymphangiomas**, also referred to as hygroma, are composed of cysts of various sizes and can be uni- or multilocular; they have no connection with the adjacent lymphatic tissue.

There is also a subtype, the **hemangiolymphangioma** that represents lymphangioma with a vascular component.

**Retroperitoneal lymphatic aneurysmal dilatation** represents a very large cisterna chyli, with variable appearance, which receives the right and left lumbar lymphatic trunks. The lymphatic aneurysmal dilatation appears as fluid-filled cystic spaces usually homogeneous, with possible sedimentation caused by hemorrhagic debris. It is essential to demonstrate the communication with retroperitoneal lymphatic vessels to prove the lymphatic origin of the unilocular or multilocular aneurysmal dilatation.[5]
LM may occur at any anatomical location, but are generally found in the cervicofacial region and axilla, followed by proximal extremities and the trunk. LM of the head and neck, approximately 75% of all LM, most commonly occur in the posterior cervical triangle followed by the axilla (20%) and the tongue musculature [6]. Uncommon sites include the skin, mediastinum, retroperitoneum, mesentery, intra-abdominal organs, pelvis, groin and bone.
Findings and procedure details

**Ultrasonography** in LM characteristically reveals thin-walled, anechoic or hypoechoic cysts, with internal hyperechoic septations of variable thickness and little evidence of flow at Colour Doppler. Microcystic LM consist of multiple tiny cysts within an avascular echogenic solid component. Calcifications can be detected if they cause acoustic shadowing. Hemorrhagic, high-protein content or infected cysts are more echogenic. [7,8] (Fig.1-7)

At **CT scan** LM appear as multiseptated cystic masses and are typically homogeneous, well-circumscribed, thin-walled and low-attenuating, usually without contrast enhancement. They may contain mural calcifications and high-attenuating fluid depicting high protein content, hemorrhage or infection [4]. (Fig.8-10, Fig.12)

At **MRI**, uncomplicated LM usually appears as a multiloculated heterogeneous mass, hypointense on T1-weighted images and hyperintense on T2-weighted images, indicating a cystic lesion, with possible rim or septal enhancement following intravenous gadolinium (Fig.11). Focal heterogeneity within the lesions appears as low-intensity linear structures representing fibrous septa.[9] They uniformly have T2 hyperintensity unless they have acute bleeding. Fluid-fluid levels may be present secondary to hemorrhage. Sometimes, there are some flow phenomena with signal heterogeneity on T2-weighted images in very large lesions. MRI cannot accurately establish the presence of mural calcifications. [5,8] Currently noncontrast MR-lymphography is considered the best technique for non-invasive evaluation of normal anatomy and abnormalities of the lymphatic systems, using heavily T2-weighted sequences to visualize static fluid with suppressed background signal. [10]

The **differential diagnosis** of LM is made, depending on its location, with other cystic-like tumors or cystic lesions, vascular malformations, hematomas and fluid collections. Neoplastic epithelium-lined cysts need to be separated histologically, as clinical presentation and ultrasound, CT and MRI imaging characteristics may be similar. Invasion of adjacent organs or the presence of metastasis could suggest a differential diagnosis of lymphangioma. True cysts, with true epithelial lining, and false cysts, such as old hematomas, usually show no wall enhancement. Mesechymal tumors, such as hemangiomas, may show similar clinical and radiographic appearance. [7,11]
Fig. 1: Microcystic lymphatic malformation (cavernous lymphangioma) of the right arm region. Grey-scale ultrasound images show multiple thin-walled, fluid-filed, irregular cysts located in the subcutaneous tissue (arrows).

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Fig. 2: Grey-scale ultrasound images of a hemangiolympangioma of the neck region showing both high-protein lymphatic (green arrows) and vascular (yellow arrow) component.

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**Fig. 3:** Microcystic lymphatic malformation (cavernous lymphangioma) of the face region. Grey-scale ultrasound (a) and Colour Doppler (b) images show a septated anechoic cystic lesion (green arrow) with few septal vascular structures (yellow arrow) located in the subcutaneous tissue.

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**Fig. 4:** Macrocystic lymphatic malformation (cystic lymphangioma) of the parotid gland. Grey-scale ultrasound (a) and Colour Doppler (b) images show large anechoic fluid-filled cysts within the parotid gland (asterisk), with few septal vessels at Colour Doppler.

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Fig. 5: Microcystic lymphatic malformation (cavernous lymphangioma) of the tongue. Grey-scale ultrasound image shows multiple thin-walled, anechoic, irregular cysts (arrows).

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Fig. 6: Macrocystic lymphatic malformation (cystic lymphangioma) of the cervical region, which appears as a large anechoic uniloculated cyst on grey-scale ultrasound images.

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Fig. 7: Macrocystic lymphatic malformation (cystic lymphangioma) of the submandibular gland. Grey-scale ultrasound image shows an anechoic fluid-filled cystic mass (green arrow) within the submandibular gland (yellow arrow).

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**Fig. 8:** 11-month old infant: Axial non-enhanced CT scan (a) and coronal contrast-enhanced reformat (b) show a macrocystic lymphatic malformation (arrows) in the right cervical region, extending inferiorly through the retroclavicular space to the axilla. The mass appears as non-enhancing, thin-walled, multiloculated, with near fluid attenuation.

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**Fig. 9:** Axial contrast-enhanced cervical CT scan (a) and sagittal reformat (b) of a 33-year old man with microcystic lymphatic malformation (cavernous lymphangioma) of the left cervical region infiltrating the sternocleidomastoid muscle (arrows).

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**Fig. 10:** A case of retroperitoneal lymphatic aneurysmal dilatation: Axial contrast-enhanced CT scan (a) and coronal reformat (b) show a homogeneous, fluid-attenuating, well-demarcated cystic lesion (arrows) occupying the left para-aortic retroperitoneal space.

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**Fig. 11:** Axial (a) and coronal (b) MRI steady-state FIESTA acquisition, of the same patient (Fig. 10) confirmed the left retroperitoneal lesion (arrows) with homogeneous fluid-like hyperintensity, communicating with the retroperitoneal lymphatic system. Axial fat-saturated gadolinium-enhanced T1-weighted MR image (c) shows no enhancement of the lesion (arrow).

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Fig. 12: Sagittal (a) and coronal (b) contrast-enhanced CT reconstructions of a patient with a left ovarian cystic tumor (asterisk), incidentally shows a left para-aortic retroperitoneal tubular structure with fatty-content (arrows), which may correspond to a dilated cisterna chyli.

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Conclusion

LM are benign lesions which have several radiological features that can be suggestive of the diagnosis, imaging allowing for a detailed preoperative evaluation of their morphology and location.
References


