Pediatric lymphatic malformations: the same pattern in different sites

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Learning objectives

1. To review lymphatic malformations epidemiology, pathophysiology and treatment;
2. To discuss the main findings in different imaging modalities, including ultrasound, CT and MRI;
3. Demonstrate that the image pattern is characteristic regardless of the site of involvement.
Background

Lymphatic malformations are uncommon lesions due to failures in the normal development of the lymphatic system and usually present as cystic masses (macro and/or microcystic), multiloculated, in childhood. They have characteristic image pattern, independent of its site of involvement.

They can be found in the most diverse age groups, but are usually diagnosed before two years of age.

Given the universal presence of the lymphatic system in various organs and systems, these lesions can be found in almost every part of the body (1).
Findings and procedure details

Epidemiology

The incidence of lymphatic malformations is about 1 in 6000 live births. They can be found in the most diverse age groups, but are usually diagnosed before two years of age and about half of cases are already present at birth. They do not have a predilection for sex. About 75% of cases occur in the cervical region and 15% in the axilla. Intra-abdominal involvement accounts for about 10% of cases (less than 1% of them are found in the retroperitoneum). They are associated with Down syndrome (1).

Pathogenesis

Lymphatic malformations are rare congenital lesions that result from failure in the development of normal lymphatic tissues, which no longer communicate with the central lymphatic system. Isolated lymphatic channels undergo a process of dilation until they form multiloculated cystic masses (2). It is also believed that they may originate from acquired factors such as trauma, fibrosis and inflammation (3).

Given the universal presence of lymphatic system in various organs and systems, these lesions can be found in almost every part of the body. They may present some complications like anemia, infection and hemorrhage, as well as diverse symptomatology due to expansive effect on adjacent structures (4).

Despite the denomination "lymphangiomas", these lesions do not present neoplastic origin, representing a congenital malformation. They are usually subdivided by the size of their cysts in microcystic (< 1cm), macrocystic (> 1cm) and mixed and may have mixed venous components (5, 6).

Treatment

Despite representing benign lesions, spontaneous regression is rare, occurring in less than 15% of cases. For asymptomatic patients, follow-up may be a viable option, although surgery with total excision of the lesion is considered the treatment of choice. Partial excision is related to recurrence, which reaches up to 10% of cases, but may be the only option due to the close relation of the lesion with adjacent organs (7).
One controversial approach for extra-abdominal injuries is intra-lesional injection of bleomycin (7). Some authors have also demonstrated reduction of the lesion with use of propranolol, which could act by inhibiting vascular endothelial growth factor (VEGF) (8).

Imaging appearance

**Computed tomography (CT):** usually shows well-defined cystic mass with attenuation similar to that of water or fat and rare cases of hemorrhagic component. The interior of the lesions does not show enhancement after contrast administration, although its internal septa and walls may be enhanced. They may also present small parietal calcifications. CT is better to determine the content of the lesion and to evaluate the relations with major vessels and surrounding structures.

**Magnetic Resonance (MRI):** depicts cystic lesions usually multiloculated, circumscribed, with low signal in T1 (some lesions may present a higher signal in this sequence when they present fatty, hemorrhagic or hyperproteic content) and high in T2, being able to form liquid levels. After administration of the paramagnetic contrast, parietal enhancement is usually present. MRI is an excellent modality to assess lesion extent in terms of tissue planes, airway compression, mediastinal extension, and potential solid organ and bone involvement (9, 10).

**Ultrasonography:** is used as the initial method of investigation of lymphangiomas. They usually appear as well delineated cystic formations with multiple septa delimiting several spaces and with posterior acoustic reinforcement. Their content may present various aspects from totally anechogetic to very heterogeneous with spaces of hyperechogenic content and debris. Color Doppler study may demonstrate the presence of flow in some septa. Ultrasound is also used to monitor the size of the lesion and determine extent preceding excision (11, 12).

We used some illustrative cases from our Pediatric Radiology group to demonstrate lymphatic malformations in different sites of the body but with similar ultrasonographic findings as previously described:

1. **Supraclavicular lymphatic malformation** (lymphangioma / cystic hygroma) - (Fig. 1)
2. **Retroperitoneal lymphatic malformation** (Fig. 2)
3. **Mesenteric lymphatic malformation** (mesenteric cyst) - (Fig. 3)
4. **Parotid lymphatic malformation** (Fig. 4)
5. **Periumbilical lymphatic malformation** (Fig. 5)
6. **Lymphatic malformation of the thigh** (Fig. 6)
Fig. 1: Supraclavicular lymphatic vascular malformation (lymphangioma or cystic hagroma). A, B, C, D and E: ultrasonographic images show cystic formation in the supraclavicular region, with multiple septations delimiting several spaces, some with hyperechoic content and others with thin debris. The color Doppler study (B and E) shows the presence of flow in some septa.

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Fig. 2: Retroperitoneal lymphatic vascular malformation. A: Ultrasonography demonstrates multiloculated (C) retroperitoneal cystic formation, located on the left of the stomach (EST), anterior to the left adrenal gland (SRE) and medial to the left kidney (RE). The same findings in B.
Fig. 3: Mesenteric lymphatic vascular malformation (mesenteric cyst). Ultrasound depicted an intra-abdominal cystic formation (C) adjacent to intestinal loops (arrows), occupying the right hypochondrium, with multiloculated appearance and fine septa. Some spaces had thick content and fine internal debris (stars). (VB: gallbladder and BEX: bladder).

Fig. 4: Left parotid gland lymphatic vascular malformation. Ultrasound images depict multiloculated cystic formation in the left parotid gland (L) with micro and macrocystic components. Right normal parotid gland (R).
**Fig. 5:** Lymphatic vascular malformation of the right paraumbilical abdominal wall. A, B, C and D: ultrasonography shows multiloculated cystic formation with some spaces with a thicker and hyperechogenic content, which may represent hematic contents. Some septal vessels are characterized by color Doppler study (C and D). (GB - gallbladder).
Fig. 6: Lymphatic vascular malformation of the posterior aspect of the right thigh. A, B, C and D: ultrasonographic images show a well delimited, multiloculated cystic formation located in the myofascial plane of the posterior region of the right thigh. It presents multiple hypervascularized septa in color Doppler study (B and D). C and D: one week later ultrasonographic images show a thicker and hyperechogenic content, representing intralesional bleeding.

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Conclusion

Lymphatic malformations can affect multiple organs and systems, participating in a vast differential diagnosis depending on their site of involvement. This makes the role of the radiologist in the recognition of his typical patterns of imaging paramount, allowing a more assertive diagnosis.
References


