Macrocystic lymphatic malformation of the neck – spontaneous haemorrhage in a patient with bleeding diathesis

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Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Arteriovenous malformations Congenital
Haemorrhage Case Type: Clinical Cases
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Patient: 5 years, male

Clinical History:

A 5-year-old boy presented to the emergency due to a sudden asymmetric increase in neck volume that morning. Physical examination revealed a slightly painful soft-tissue mass in the left cervical region, non-compressible, non-pulsatile and without inflammatory signs. There was no relevant personal medical history; his sister has von Willebrand disease.

Imaging Findings:

Emergency ultrasound showed a large lobulated multiseptated cystic lesion with well-defined walls and some echogenic content, without intralesional Doppler signal. It was in direct continuity with the left submaxillary gland.

Magnetic resonance imaging (MRI) performed 3 days later depicted a macrocystic transpatial lesion centred in the left parapharyngeal space with high signal intensity on T1 and T1 with fat saturation and multiple fluid-fluid levels due to the presence of haemorrhagic content, without contrast-enhancement, compatible with a lymphatic malformation.

Discussion:

Lymphatic malformations are congenital lesions resulting from maldevelopment of the lymphatic system, with arrest and sequestration of channels that fail to communicate with the rest of the draining system [1-3].

The neck is the most common location, comprising around 70-80% of cases [2].

Histologically, they are cavernous lymphatic spaces lined by endothelium, lacking muscularised walls. The term lymphangioma is discouraged as they are not neoplastic lesions.

The revised classification of the International Society for the Study of Vascular Anomalies divides these common lymphatic malformations into three subtypes – macrocystic (with cysts > 1-2cm; previously known as ‘cystic hygroma’), microcystic (can appear solid), and mixed [3].
Being present at birth, they grow with the child and do not spontaneously regress [2]. Most are diagnosed before age 2.

On physical examination they show up as a soft-tissue mass of rubbery consistency, non-compressible and non-pulsatile [2]. Occasionally, they can present as a sudden cervical mass due to intralesional haemorrhage or infection, with dramatic enlargement, as was the case with our patient.

These lesions can vary significantly in size, and very young patients can present with pain, dyspnoea or even respiratory compromise.

MRI is the best imaging tool for conclusively diagnosing lymphatic malformations, enabling precise anatomic extension of the lesion and cervical spaces involved. By nature, they are infiltrative lesions that do not respect fascial planes and can be bilateral [1-3]. On T2 the cystic spaces have predominantly high signal and well-defined contours. There are no ‘flow voids’ or phleboliths – if present, a mixed venous-lymphatic malformation needs to be considered [1]. On T1 they have predominantly low signal, unless there was haemorrhage, in which case hyperintensity and fluid-fluid levels due to layering blood products are characteristic [1-3].

Surgical treatment is usually employed. Larger lesions, due to their infiltrative nature, can be very difficult to excise and sclerotherapy can be used [1]. There are also some reported good responses to medical therapy with sirolimus [1, 3].

Our patient tested positive for von Willebrand disease with <10% of factor VIII. Due to the size of the lesion, he was still proposed for surgery.

TEACHING POINTS

Lymphatic malformations are congenital non-neoplastic lesions most frequent in the neck.

They can have a sudden presentation as a soft-tissue mass, mimicking cervical adenopathies and other cystic lesions of the neck, like branchial cleft cysts.

MRI is the modality of choice for diagnostic purposes and presurgical anatomic mapping.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Macrocystic lymphatic malformation, Branchial cleft cyst, Lymphadenomegaly, Adenophlegmon, Vascular malformation, Haematoma

**Final Diagnosis:** Macrocystic lymphatic malformation

**References:**


**Figure 1**

**a**

Description: Ultrasound of the left cervical region shows a hypoechoic lesion with regular margins and multiple internal septa, with over 5 cm. **Origin:** © Department of Radiology, Hospital Beatriz Ângelo, Loures, Portugal 2019

**b**

Description: The lesion shows no internal colour Doppler signal. Notice the fluid-debris level due to the presence of blood products (arrow). **Origin:** © Department of Radiology, Hospital Beatriz Ângelo, Loures, Portugal 2019
**Figure 2**

**a**

**Description:** MRI of the neck, axial T2. There is a large cystic lesion centred in the parapharyngeal space with multiple septa and fluid-fluid levels (arrow). **Origin:** © Department of Radiology, Hospital Beatriz Ângelo, Loures, Portugal 2019

**b**

**Description:** MRI of the neck, axial T1. The lesion is hyperintense on T1 and T1 FS, due to the recent haemorrhage. **Origin:** © Department of Radiology, Hospital Beatriz Ângelo, Loures, Portugal 2019
**Description:** MRI of the neck, axial T1 FS after gadolinium administration. The lesion is hyperintense on T1 and T1 FS, due to the recent haemorrhage. There is minimal peripheral enhancement. **Origin:** © Department of Radiology, Hospital Beatriz Ângelo, Loures, Portugal 2019