Persistent swelling at the ankle joint: diagnosis and discussion

Wim Verelst · Johan Huygh · Eric Van Marck ·
Philip Van Hoenacker · Jan Gielen ·
Arthur De Schepper

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Diagnosis

The histopathological examination showed multiple thin lymphatic channels, lined with flattened endothelium, and some small lymphoid aggregates. Few red blood cells could be demonstrated. There was no immunoreactivity for CD31, CD34, EMA or S-100. Therefore, the diagnosis of multiple lymphangioma of the capillary subtype (lymphangioma simplex) was made.

Discussion

Lymphangiomas are regarded as congenital sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system [1, 2]. Histologically, lymphangiomas

consist of dilated thin-walled lymphatic channels and are divided into three groups: the lymphangioma simplex (composed of thin-walled lymphatics), the cavernous lymphangioma (composed of larger lymphatic channels), and the cystic lymphangioma (or ‘cystic hygroma’, consisting of large macroscopic lymphatic spaces with collagen and smooth muscle tissue) [2–4].

Although it is probable that some lesions are acquired following surgery, irradiation or infection, lymphangiomas are mainly congenital malformations present at birth in more than 50% of the cases. As many as 90% are revealed before the end of the second year of life [1, 5, 6]. Although lymphangiomas typically develop in areas where the primitive lymph sacs occur (i.e. head, neck, and axilla, accounting for about three-fourths of all lymphangiomas), lymphangiomas can affect any part of the body served by lymph vessels. Other reported locations [1, 2, 5, 7] include skin, mediastinum, abdomen, retroperitoneum, and the limbs.

Location at the ankle/foot, as described in this case, is extremely rare: case reports of ankle/foot lymphangiomas are very scarce [8, 9].

Symptoms caused by lymphangiomas are mainly attributed to mass effect (in which size and location are critical), torsion, rupture, intratumoral bleeding, and the proclivity to become secondary infected.

Only rare cases of spontaneous regression have been reported [8]. Most lesions require surgical treatment, as they have a tendency towards gradual growth. Frequently, postsurgical recurrences are seen, especially after incomplete resection.

Because of the high percentage of recurrence after partial excision, total primary surgical resection is of utmost importance. In assessing the local extent of the lesions, MR imaging is indispensable [10] due to its superiority to other techniques for defining soft tissues.

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Wim Verelst and Johan Huygh are 6th year medical students.

W. Verelst · J. Huygh
University of Antwerp,
Antwerp, Belgium

J. Huygh (✉)
Drabstraat 81B,
2640, Mortsels, Belgium
e-mail: johanhuigh@hotmail.com

E. Van Marck
Department of Pathology, University Hospital of Antwerp,
University of Antwerp,
Antwerp, Belgium

P. Van Hoenacker · J. Gielen · A. De Schepper
Department of Radiology, University Hospital of Antwerp,
University of Antwerp,
Antwerp, Belgium

We compared our own findings to the general characteristics described in the radiologic literature:

Although lymphangiomas mostly appear as single lesions, we present a lymphangioma consisting of four different lesions.

On T1-weighted images, most reported lymphangiomas are of low SI, similar to that of muscle. In our case, the lesions have a slightly higher SI than muscle. Very often, lymphangiomas contain focal heterogeneities. The latter could not be demonstrated in the current case.

On T2-weighted images, signal intensities of lymphangiomas are higher than signal intensities of fat, reflecting the preponderance of fluid-filled spaces. Frequently, focal intralesional inhomogeneities of lower SI can be demonstrated. Pathologically, these inhomogeneities correspond to fibrous septa. These features were also found in this case.

In lymphatic malformations, enhancement is rarely seen, except in the mixed lymphatic venous lesions. However, in the presented case of a capillary lymphangioma, there was a marked non-homogeneous enhancement.

A striking feature in our case is the erosion of the tibial cortex caused by a slowly growing paracortical lesion, a feature that was not previously reported. This is probably attributable to the fact that the tumor was growing in a space confined by the superior extensor retinaculum.

The radiological differential diagnosis in this type of multinodular lymphangioma should include tumors such as neurofibroma in neurofibromatosis type I, schwannoma in schwannomatosis and multiple hemangiomas, as in Klippel–Trenaunay–Weber syndrome.

On MR imaging, this differential diagnosis is often straight forward, because of different contrast behavior and enhancement pattern. The differential diagnosis in this case was difficult due to the mild hyperintensity on T1-weighted images and marked non-homogeneous enhancement after

contrast injection, features also present in MR images of neurofibroma, schwannoma, and hemangioma. The atypical location of the lesions was also not specific for lymphangiomatosis. Features that were contributory to the diagnosis included the reticular pattern of infiltration suggestive for lymphangitis and the clinical investigation.

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