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Coral reef lymphatic malformation: A new clinical phenotype

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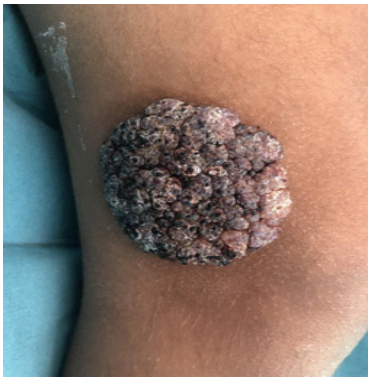
Abstract

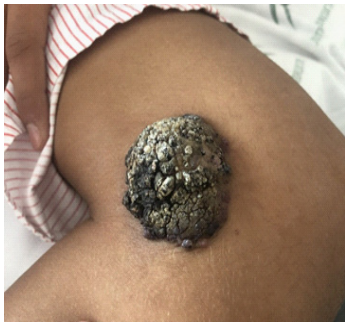
We present a new clinical phenotype of lymphatic malformation with a characteristic morphology in two young patients. Strikingly, in both patients the lesion had the same anatomic location..

Keywords: Microcystic; lymphatic malformation; D2-40; vascular malformation

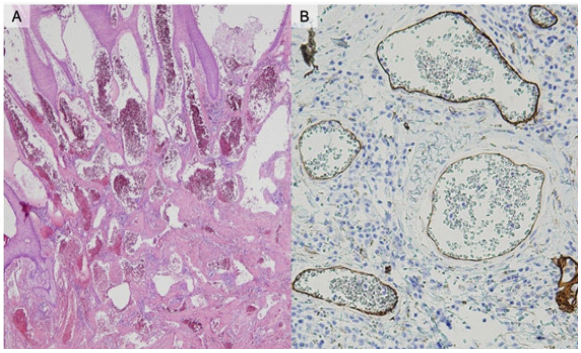
Presentation of the Cases

A 15-year-old girl of gypsy ethnicity presented with a slow-growing exophytic lesion present since birth on the lateral aspect of the right thigh. It caused mild local discomfort. Physical examination revealed a 6 cm brown multilobulated mass (**Figure 1**). An 11-year-old girl of Moroccan origin presented with a lumpy congenital lesion on the lateral aspect of the right thigh, intermittently draining a foul-smelling exudate. Physical examination revealed a 5.5 cm dark brown multilobulated mass (**Figure 2**). In both cases the lesion was surgically excised.





Histological study in both cases (Figure 3A) revealed small-medium sized channels lined by flat lymphothelium and irregular smooth muscle. The endothelial cells were immunohistochemically positive for D2-40 (**Figure 3B**). The diagnosis was microcystic lymphatic malformation (LM).



LMs are benign vascular anomalies secondary to embryologic alterations in the development of the lymphatic system [1]. As vascular malformations, they are present from birth and can cause both anatomical deformation and functional Deficits [1]. Microcystic LMs present as clear, tiny vesicles permeating the subcutaneous tissue and muscles. Patients usually present with serous or bloody exudate, crusting, swell-

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ing or even infection in the affected region. Histologically, cystic LMs are composed of vascular spaces filled with eosinophilic, protein-rich fluid [2]. Antibodies against lymphatic endothelial cell markers help to identify LMs [2].

Depending on the clinical presentation, surgery may be offered to some patients. For microcystic LMs, sclerotherapy has been less successful. The utility of systemic rapamycin in the treatment of complex LMs has been demonstrated in several studies [3], and compelling evidence for the use of topical rapamycin has also emerged in recent years [4].

A multidisciplinary approach allows the optimal treatment plan for each patient. LM can be challenging for the clinician. We present this case because of its originality. To the best of our knowledge, these are the first two published cases with this curious phenotype.

Competing interest Disclosures: The authors have no competing interests to declare.

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