

Cutaneous lymphangioma circumscriptum: The relevance of clinical, dermoscopic, radiological, and histological assessments

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Key Clinical Message

Cutaneous lymphangioma circumscriptum (CLC) is a rare congenital malformation of the superficial cutaneous lymphatic ducts. Case report: A 7-year-old boy presented plaque with grouped vesicles resembling a “frog spawn” on the upper left thigh, since 4 months old. The clinical, dermoscopic and histopathological correlation is of great importance.

KEYWORDS

cystic tumor, dermoscopy, lymphangioma, vascular malformations

1 | INTRODUCTION

Cutaneous lymphangioma circumscriptum (CLC) is a rare congenital malformation of the superficial cutaneous lymphatic ducts, which manifests isolated or associated with deep lymphangioma or angiodysplastic syndromes (Proteus, Cobb, or Maffucci). This lymphangioma is characterized by changes in the lymphatic structures with the formation of subcutaneous cisterns and ectopic vessels interconnected through dilated ducts that project into the skin in the form of superficial vesicles with thin walls and variable color. In this article, we report a rare case of cutaneous lymphangioma circumscriptum with emphasis on the relevance of analyzing dermoscopic patterns for a proper clinical diagnosis. We also report its correlation with radiological and histopathological findings.

2 | CASE PRESENTATION

A 7-year-old boy, whose background presented painless, nonitchy grouped injuries, with the same tone as the skin,

clear or reddish secretion drainage on the upper left thigh since he was 4 months old.

The dermatological examination showed presence of well-defined, irregular, brownish plaque of 15 × 4 cm, surmounted by small grouped translucent vesicles with a serohematic content and satellite lesions with the same characteristics, resembling a “frog spawn” (Figure 1).

The examination did not present palpable lymphadenomegalies.

Dermoscopy examination showed lacunes of brown-orange coloration in the upper portion and red-violet coloration in the lower portion, characterizing the hypopio-like, wrapped by pale septa, and a diffuse pink lacunar area with fine straight-line vessels (Figure 2).

The magnetic resonance imaging (MRI) revealed serpiginous-like structures in the subcutaneous tissue on the anterolateral face of the thigh root on the left, which are isointense in relation to musculature in T1 and hyperintense in T2 (Figure 3).

Histopathological examination showed lymphatic ducts with thin walls in the papillary dermis, finely granular hyaline material, and erythrocytes in the lumen of the lymphatic vessels, which, as they were arranged, appeared to have an intra-epidermal location.

The CLC diagnosis was suggested by the clinical practice and dermoscopy examination, mapping the extent and

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FIGURE 1 Cutaneous lymphangioma circumscriptum. Well-defined, irregular, brownish plaque of 15 × 4 cm, surmounted by small grouped translucent vesicles with a serohematic content and satellite lesions with the same characteristics on the upper left thigh



FIGURE 2 Cutaneous lymphangioma circumscriptum. Dermoscopy examination: Brown-orange lacunes in the upper portion and red-violet in the lower portion (hypopio-like), wrapped by pale septa

location with the MRI and confirming it with a histopathology examination.

All the guidelines regarding the disease were given to the family, as well as its evolution and therapies. The family chose the conservative treatment.

3 | DISCUSSION

Lymphangiomas are uncommon malformations of the lymphatic ducts and may compromise any cutaneous or mucous surface.¹⁻³ They may be localized or generalized malformations and have a congenital or acquired origin.^{2,3}

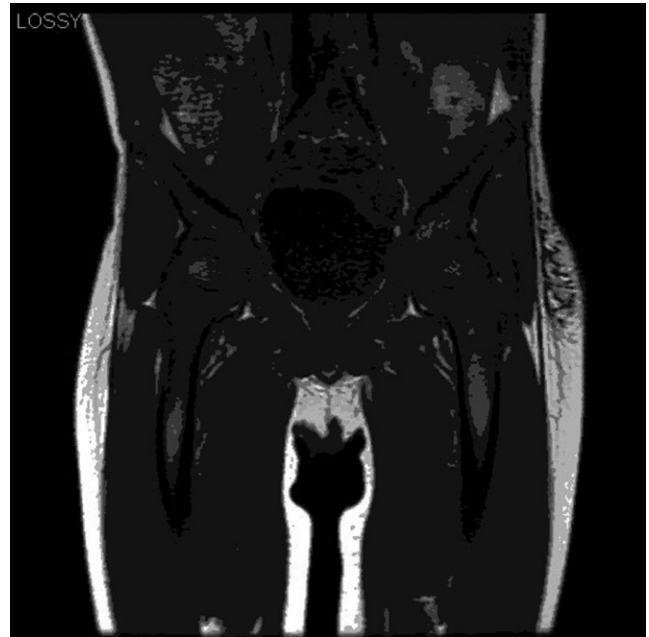


FIGURE 3 Cutaneous lymphangioma circumscriptum. MRI of the left thigh: Serpiginous-like structures in the subcutaneous tissue on the anterolateral face of the thigh root on the left, which are isointense in relation to musculature in T1

Lymphangiomas were classified as simple, cavernous, and cystic,⁴ named simple cutaneous lymphangioma as lymphangiectodes,⁵ and renamed circumscribed lymphangioma.⁶ Later, they were grouped according to the depth and size of the abnormal lymphatic vessels into two categories: superficial, represented by cutaneous lymphangioma circumscriptum (CLC), and deep, with cavernous lymphangioma and cystic hygroma.² Lymphangiomas represent 4% of the vascular tumors and around 25% of benign vascular tumors in childhood.^{2,3,7,8} This congenital malformation is apparent at birth in approximately 50% of cases.^{1,2} In 90% of cases, it arises until two years of age and it is not a gender-related disease.^{2,7}

Cutaneous lymphangioma circumscriptum, currently named as superficial lymphatic malformation (SLM), is a rare benign tumor and the most common lymphangioma.^{1,3} It affects skin and mucous surfaces, especially head and neck, proximal extremities, buttocks, chest, armpits, and oral cavity.^{1,3} They are common cutaneous findings: a group of transparent vesicles of 2-4 mm, similar to frog spawn, with varied colors as a result of hemoglobin degradation.^{2,3,7} They may be under hyperkeratotic and/or verrucous plaques, with areas that range from small and well delimited to large, diffuse areas with imprecise limits.⁷ The case report corroborated the literature regarding the presentation, location, and most common clinical findings of the superficial lymphangioma on the buttocks with appearance of “frog spawn.”

Evolution is stable, with rare episodes of lymphorrhea, intracystic hemorrhage, and infection. The most relevant

TABLE 1 Correlation between clinical, dermoscopic, histological, and radiological findings

Clinical manifestations	Dermoscopic examination	Histopathological examination	Imaging test (MRI)
Small grouped translucent vesicles with a serohematic content as a “frog spawn”	Light brown to violet lacunes, surrounded by pale septa Hypopio-like: transition from a light color on the upper portion to red-violet in the lower portion	Dilated lymphatic vessels with lumen filled with finely granular hyaline material and red blood cells from shunts, in the papillary dermis, with reticular capillary and subcutaneous tissue Hypopio-like represents the sedimentation of blood in serum and cellular components	serpiginous-like structures in the subcutaneous tissue, isointense in relation to musculature in T1 and hyperintense in T2
Serous-content vesicles	Areas varying from yellowish white to diffuse rose with fine linear vessels	Dilated lymphatic vessels with lumen filled with finely granular hyaline material and intermediate blood vessels	

diagnoses were as follows: herpes zoster, molluscum contagiosum, viral warts, angiosarcoma, hemangioma, epidermal nevus, angiokeratoma, and leiomyoma.^{2,3,7,8}

Dermoscopy presents a lacunar/saccular pattern, because of the presence of lymphatic fluid varying from light brown to violet color, a result of the volume of blood wrapped by pale septa, which shows the hypopyon-like, characterized by blood sedimentation.^{3,8,9} Another dermoscopic pattern consists of colored areas varying from yellowish white to diffuse rose with linear and sharp vessels.^{1,3,9} In the case reported, despite the suggestive clinical examination, the evidence of dermoscopic patterns, associated with the presence of hypopyon-like, reinforces the reliability of this method for clinical diagnosis, allowing to rule out the most important differential diagnoses.

Imaging tests determine the extent of the lesion, the involvement of internal organs, or compression of adjacent structures.¹⁰ These tests also help planning the surgical excision. MRI is the examination of choice, in which the diagnosis is referred to by the evidence of serpiginous-like structures in the subcutaneous tissue.¹⁰ We observed the low signal intensity in the weighted images of T1 and the signal hyperintensity in the weighted images of T2. The intensity of the signal may vary according to the composition of the liquid within the vesicles. Histopathological examination is considered gold standard in the diagnosis of CLC,^{1-3,9} in which vesicles represent dilated lymphatic vessels in the papillary dermis, with the possibility to extend up to the reticular dermis and subcutaneous tissue, with or without hyperkeratosis and irregular acanthosis.^{2,7} The lymphatic lumen is filled with finely granular hyaline material and red blood cells, probably from small capillary shunts.¹

The correlation of the clinical, dermoscopic, radiological, and histopathological findings of the lymphatic system was well demonstrated in this case (Table 1).

Surgical excision is the approach of choice, with 75% of cure and low rates of recurrence.^{1,2,10} Other therapeutic modalities described are sclerotherapy, cauterization, cryotherapy, radiotherapy, CO2 laser, and thermoablation.¹⁰

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CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTION

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