

CASE REPORT



Acral microcystic lymphangioma: differential diagnosis in verrucous lesions of the extremities *

Linfangioma microcístico acral: diagnóstico diferencial em lesões verrucosas de extremidades

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Abstract: Lymphangiomas are a malformation caused by the abnormal migration of lymphatic tissue, leading to failures in the communication and drainage of the lymphatic system. They usually present as groups of translucent papules and vesicles in the skin or mucous membranes. Presentation as a verrucous plaque limited to a single toe is unusual and emphasizes the relevance of this case report. Although considered a benign lesion, depending on the site affected by the lymphangioma and its size, it may lead to localized pain or recurrent infections, substantially affecting the patient's quality of life. The present case report describes a patient with lymphangioma of late onset developing in adulthood in the form of a verrucous plaque confined to a single toe. Diagnosis of this unusual presentation could only be confirmed following histopathology. Surgical excision resulted in a satisfactory functional and cosmetic outcome.

Keywords: Lymphangioma; Lymphatic abnormalities; Vascular diseases

Resumo: Linfangioma é uma má-formação originária da migração anormal de tecido linfático, levando a falhas na comunicação e na drenagem da linfa. Apresenta-se mais comumente como vésico-pápulas agrupadas de conteúdo translúcido na pele ou mucosas. A apresentação inicial na forma de placa verrucosa limitada a um único pododáctilo é atípica e reforça a relevância deste relato. Ainda que considerado lesão benigna, o linfangioma, a depender de sua localização e dimensão, pode apresentar dor local ou infecções recorrentes, com interferência substancial na qualidade de vida. Relatamos caso de linfangioma circunscrito a pododáctilo de surgimento tardio na idade adulta, na forma de placa verrucosa – apresentação atípica confirmada apenas após histopatológico. A excisão cirúrgica garantiu resposta estética e funcional satisfatória.

Palavras-chave: Doenças linfáticas; Doenças vasculares; Linfangioma

Received on 20.03.2010.

Approved by the Advisory Board and accepted for publication on 05.05.2010.

* Study conducted at the *Dona Libânia Dermatology Clinic* (CDERM), Fortaleza, Ceará, Brazil.

Conflict of interest: None / *Conflito de interesse: Nenhum*

Financial funding: None / *Suporte financeiro: Nenhum*

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INTRODUCTION

Vascular malformations are present at birth in 60% of cases, becoming apparent by the time the child reaches two years of age in 90% of cases. Regression is not generally spontaneous.

According to the 1996 classification of the International Society for the Study of Vascular Anomalies (ISSVA)/Mulliken, vascular anomalies may be classified as simple (capillary, lymphatic, venous and arterial) or combined.¹ A later classification system subdivided these malformations according to their vascular component and flow characteristics.² Lymphangioma is a malformation that originates from a failure in the normal communication between lymph and the lymphatic system. Lymphangiomas are generally composed of small vesicles containing translucent fluid that form plaques on the skin or mucosae.³

Lymphangioma circumscriptum is generally present at birth or appears within the first few months of life. Involution is rarely spontaneous.⁴ The lesion is characterized by small vesicles-papules clustered together in plaques on the skin or mucosae, with translucent or slightly hemorrhagic contents. The most common sites are the face and neck, the axillae, chest and the extremities; however, it may theoretically affect any region.⁵

Although the vesicular form is the most common, lymphangiomas may also present in the form of verrucous lesions, as in the present case. In this latter form, the presence of small, blackened dots on the surface may be confused with warts when located on the perineum. Clinically, they may be confused with molluscum contagiosum lesions.² Local hypertrichosis is rare.⁷

Initial presentation as a progressively growing

verrucous plaque limited to a single toe reinforces the relevance of this report in view of the unusualness of the present case.

CASE REPORT

A 52-year old, female teacher from the city of Fortaleza reported a lesion on the second toe of her left foot that was progressively increasing in size and associated with pain when walking, becoming worse at the end of the day. She reported no previous trauma at the site and no family history of any similar complaint. At examination, the lesion presented as a verrucous plaque with a soft surface completely encompassing the circumference of the second toe of her left foot (Figure 1). Because of the verrucous nature of the lesion, diagnostic hypotheses included: skin tuberculosis, chromomycosis, leishmaniasis, sporotrichosis and common wart. Laboratory analysis identified no significant abnormalities. Purified protein derivative (PPD) and Montenegro's skin test were nonreactive. Since diagnosis remained undefined, biopsy was performed. At histopathology, the skin showed a proliferation of tortuous lymph vessels, some of which were dilated, situated in the papillary and reticular dermis. Some of these vessels had small intraluminal projections with a papillary appearance beneath the epidermis. The vessels were lined with flat endothelial cells without atypia. A remarkable finding in the epidermis was the associated hyperkeratosis and moderate acanthosis (Figures 2 and 3). In accordance with the clinical and pathological findings, a diagnosis of lymphangioma circumscriptum was made and resection of the lesion was carried out. For local reconstruction, a partial skin graft was performed using thick skin from the calf of the ipsilateral



FIGURE 1: Soft, verrucous tumor on the second toe of the left foot

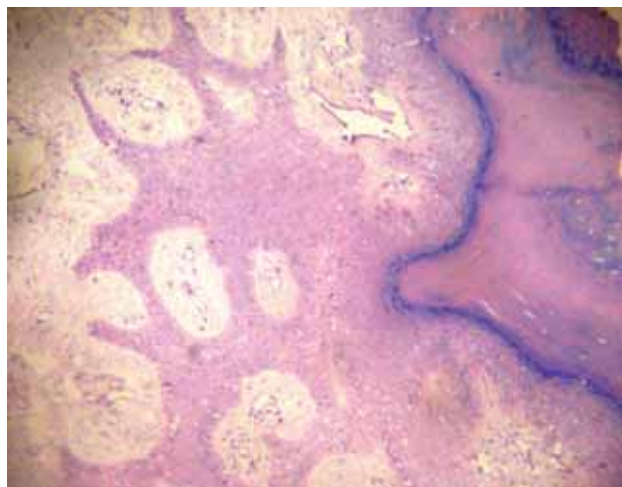


FIGURE 2: (HE 40x). Hyperkeratosis and acanthosis. Proliferation of lymph vessels in the dermis

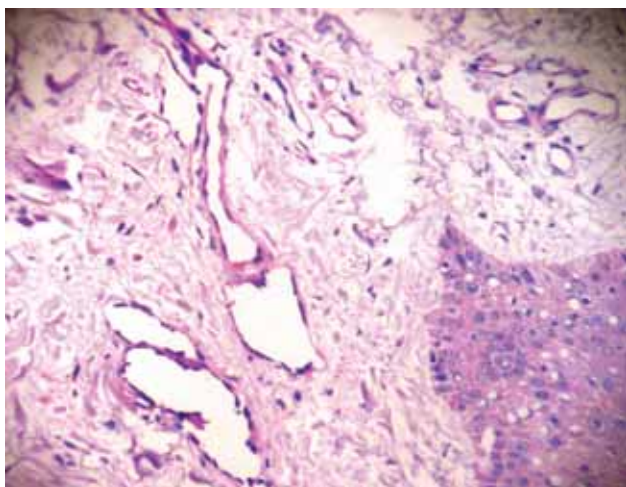


FIGURE 3: (HE 100x). Note the ectatic lymph vessels in the papillary dermis



FIGURE 4: Appearance of the lesion four months after surgery

leg. The cosmetic and functional results were satisfactory. At the present time, four months after surgery, the patient is progressing well and is satisfied with the esthetic appearance and the improvement in pain. Only a small local, residual edema remains. No immediate or delayed significant postoperative complications occurred (Figure 4).

DISCUSSION

Vascular malformations are present at birth in 60% of cases and become apparent by the second year of life in 90% of cases. In general, they do not regress spontaneously.⁸ Localized lymphatic malformations may be macro- or microcystic.

Macrocystic lymphatic malformations, referred to as cystic hygromas, are large, soft lesions covered with a smooth, translucent surface of normal or bluish-colored skin, typically located on the triangle at the back of the neck.⁸

Microcystic lymphatic malformations, also known as lymphangioma circumscriptum, contain cystic spaces of less than two centimeters.² Their etiology is uncertain; however, there are reports of proliferation following trauma.⁹ Although considered a benign lesion, pain, local infection, ulceration and hemorrhage following the slightest trauma are possible complications. The volume of the lesion may increase due to hemorrhages, the accumulation of fluids from poor local drainage and/or exacerbated inflammatory response. Lesions may be single or multifocal, circumscriptive or infiltrative.⁵

There are few reports in the literature of late onset in adults and those reported generally result from local trauma. Cases secondary to inflammatory diseases and ganglion tumors, with or without formation of stenosis and fistulae, have also been described

in association with tuberculosis, extensive Crohn's disease and chronic lymphedemas. Vulvar lesions following surgery and/or radiotherapy for pelvic malignancies have also been described.^{10,11}

At histopathology, hyperkeratosis and acanthosis may be present in variable degrees. In the dermis, lymph vessels with thin walls containing no blood may be surrounded by lymphocytes or not. The presence of blood inside the vessels may be indicative of a recent hemorrhage or a combined malformation, generally venous and lymphatic.¹²

Diagnosis is based on clinical and histopathology findings, the latter being crucial. Supplementary exams such as ultrasonography, computed tomography and magnetic resonance imaging may provide further evidence of the lymphatic origin of the lesion and of its extent, data that are useful when taking decisions regarding therapy and when making a differential diagnosis with soft tissue tumors.

Differential diagnosis should include lymphedema, angioma, angiokeratoma, glomangioma and other vascular tumors. In some cases such as the present one, the clinical manifestation is so atypical (late onset, site restricted to a single toe, onset and progression in the form of a verrucous plaque) that confirmation could only be made following biopsy. Initial presentation as a slowly growing verrucous plaque limited to a single toe reinforces the relevance of this report in view of the unusualness of the case. The fact that the patient denied any history of trauma at the site should be considered with caution, since toes are often subject to micro-trauma, particularly in women, who tend to use tight-fitting, fashionable shoes.

Treatment is generally complicated. In superficial lesions, electrocoagulation, laser and cryotherapy

may be used. Alternative forms of treatment include sclerosing agents such as bleomycin, doxycycline, ethanol, acetic acid and OK-432 (lyophilized incubation mixture of group A *Streptococcus pyogenes* of human origin).^{7,13}

Biomolecular perspectives include studies that are currently being conducted to develop D2-40, a marker that is highly specific for lymphatic endothelial cells and is not found in the healthy endothelium or in venous, arterial or capillary malformations.⁵

Due to the tendency of the lesion to grow, surgical resection is the most effective therapeutic option in the majority of cases, principally when the lesion is affecting function, causing esthetic problems or is prone to infection.⁴ Hypertrophic scars are common postoperative complications due to remaining lymph vessels resulting from incomplete local resection, a factor that also affects prognosis in terms of recurrence.⁷

A recent study carried out in 128 children with lymphangiomas followed up between 1979 and 2005 at a referral center in Japan confirmed that primary surgical excision of the lesion, whenever possible, results in a better response when compared to sclerotherapy with bleomycin and OK-432; however, the potential for complications following the procedure is greater, as previously shown.¹³

In conclusion, we would like to emphasize the importance of including lymphangiomas in the differential diagnosis of verrucous lesions of the extremities. □

ACKNOWLEDGEMENTS

The authors would like to thank Dr. Igor Santos Costa, pathologist, for his collaboration in describing the histopathology results.

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How to cite this article/Como citar este artigo: Jucá NBH, Crisóstomo MGR, de Oliveira LMP, Cavalcante HA, de Sousa ARD. Acral microcystic lymphangioma: differential diagnosis in verrucous lesions of the extremities. *An Bras Dermatol*. 2011;86(2):343-6.