CASE REPORT

Acréal pseudolymphomatous angiokeratoma: case report and literature review*

Angioqueratoma pseudolinfomatoso acral: relato de caso e revisão da literatura

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DOI: http://dx.doi.org/10.1590/abd1806-4841.20132413

Abstract: The authors describe a case of a female patient with Acréal Pseudolymphomatous Angiokeratoma of Children, known as APACHE. It is a rare benign cutaneous disease, of unknown etiology, characterized by multiple, asymptomatic erythematous-violaceous papules and nodules, usually located unilaterally with acral distribution. Today, this denomination is questionable, since there are published reports of this disease in adults and in different locations. Clinically, it is similar to an angiokeratoma, whereas histologically, it corresponds to a distinct type of pseudolymphoma. The immunohistochemical study is required to distinguish APACHE from cutaneous lymphoma.

Keywords: Child; Extremities; Hemangioma; Pseudolymphoma

Resumo: Relata-se o caso de uma paciente com diagnóstico de angioqueratoma pseudolinfomatoso acral, conhecido por sua sigla em inglês APACHE - Acréal Pseudolymphomatous Angiokeratoma of Children. É uma doença cutânea benigna, rara, de etiologia desconhecida, caracterizada por múltiplas pápulas e nódulos eritemato-violetas assintomáticas, de localização geralmente unilateral e acral. Atualmente, questiona-se esta denominação, já que há relatos na literatura do quadro em adultos e em outras localizações. Clinicamente, é similar a um angioqueratoma, porém, histologicamente, corresponde a um tipo distinto de pseudolinfoma. O estudo imunohistoquímico é necessário para diferenciação dos linfomas cutâneos.

Palavras-chave: Criança; Extremidades; Hemangioma; Pseudolinfoma

INTRODUCTION

Acréal pseudolymphomatous angiokeratoma is a rare benign, cutaneous disease, mostly affecting children aged between 2 and 13. It is characterized by multiple asymptomatic erythematous-violaceous papules and nodules, usually located unilaterally with acral distribution. Initially considered a vascular malformation, today it is classed as a distinct type of pseudolymphoma. It is known by the acronym APACHE (Acréal Pseudolymphomatous Angiokeratoma of Children) and was described for the first time by Ramsay in 1988. Currently, this denomination is under debate, as there are reported cases in the literature of adults with the condition, located in other parts of the integument. Its etiology remains unknown. Histopathological exams reveal preserved epidermis and dense lymphocytic infiltrate in the dermis, permeating structures of the conjunctive tissue, affecting (or not) the cutaneous annexes. The immunohisto-
A chemical study is necessary in order to distinguish the cutaneous lymphomas. The elective therapeutic choice is total excision of lesions but intralesional corticotherapy, cryotherapy and radiotherapy are also described, though they entail recurrence.

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White, female patient, aged 11, complaining of “small lumps on the left wrist since the age of two”. The dermatological exam revealed erythematous-violaceous papules on the left wrist, which appeared fainter during the vitropression test (Figure 1). The condition had been evolving for 9 years, without any changes in clinical characteristics, local symptomatology or previous trauma. The histopathological exam revealed an integral epidermis without atypia; and, in the dermis, inflammatory infiltrate of superficial and deep, perivascular location, around the pilo-erector muscles, hair follicles, eccrine glands and nerves (Figures 2 and 3A). The infiltrate was made up of lymphocytes, some histiocytes, a moderate number of plasmocytes and small basophilic granulations in the cytoplasmas of the histiocytic cells. Infiltration was noticed in the hypodermal lobules. The search for alcohol-acid resistant bacilli using the Ziehl-Neelsen staining procedure culminated in a negative result. In regard to the inflammatory infiltrate, abundant in plasmocytes, a clinical and laboratory investigation was conducted to rule out leishmaniasis and syphilis, with the following findings: non-reactive VDRL and FTA-Abs results (Figure 3B). Surveys for micobacteria, treponema and leishmania, by immunohistochemistry, produced negative results. In addition: hemogram, hepatic and renal function, ESR, dosage of immunoglobulins and protein electrophoresis, without changes.

Thus, total excision of the lesions was initiated, to diagnostic and therapeutic ends. The histopathological exam revealed the same histopathological pattern as before. The immunohistochemical study of the biopsy material revealed histological characteristics of dermal lymphoid proliferation of small, perivascular and adnexal, T and B lymphocytes, with a low proliferative index (Figure 4 and Chart 1).

The patient is undergoing outpatient follow-up, with no signs of recurrence at present.
Chemical and physical changes are often observed in the affected skin. The most common symptoms include erythema, pruritus, and pain. The lesions are typically located on the extremities, particularly on the hands and feet. The lesions may be single or multiple, and they can vary in size and shape. The histological examination reveals a dense inflammatory infiltrate composed of lymphocytes, histiocytes, and eosinophils. Immunohistochemical staining is performed to identify the presence of specific cell markers and gene receptors, such as CD4, CD8, and CD20. These markers help to differentiate between different types of cutaneous lymphomas. The presence of high levels of CD4 and CD8-positive T lymphocytes suggests a pseudolymphomatous process, while the absence of B lymphocytes indicates a T-cell lymphoma. In rare cases, an immunohistochemical panel is used to rule out cutaneous lymphomas. The presence of B lymphocytes in significant numbers suggests the diagnosis of a cutaneous lymphoma. The clinical course of the disease is usually self-limited, and the lesions resolve spontaneously over time. In some cases, however, the disease may persist for several years. The prognosis is generally good, and there are no documented cases of metastasis. Therefore, the treatment is usually conservative, with close monitoring and follow-up. Reference: An Bras Dermatol. 2013;88(6 Suppl 1):S39-43.
Studies with polymerase chain reaction (PCR) showed the polyclonality of the inflammatory infiltrate, thus suggesting the reactional process of this entity.\textsuperscript{4,5,6}\n
Tokuda et al., through an immunostaining study of the lesion's vessels, obtained positive results for specific markers of the endothelium of the lymphatic vessels (podoplanin) and blood vessels (CD 34).\n
Linking these findings to the epidemiology of the disease, they again considered the hypothesis that the histogenesis of APACHE could be a type of vascular malformation or hemangioma.\textsuperscript{7}\n
The elective therapeutic choice is total excision of the lesion, but intralesional corticotherapy, cryotherapy and radiotherapy are also described, though they entail recurrence.\textsuperscript{2}
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How to cite this article: Lessa PP, Ferreira Jorge JC, Ferreira FR, Alvarenga Lira ML, Mandelbaum SH. Acral pseudolymphomatous angiokeratoma: case report and literature review. An Bras Dermatol. 2013;88(6 Suppl 1):S39-43.