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Generalized Multinucleate Cell Angiohistiocytoma: A Case Report

Greco Daniel Alejandro, Paradeda María Eugenia, Diaz de la Fuente Florencia and Cachau María Victoria*

Departamento de Patología, Hospital de Clínicas José de San Martín, Argentina

Abstract

Multinucleated Cell Angiohistiocytoma (MCAH) is a rare benign fibro-histiocytic and vascular proliferation. It can present in a localized or generalized manner. Localized forms show a predilection for women in the sixth decade of life. Clinically, it is characterized by single, isolated, brown to purple papules and nodules that are asymptomatic, distributed on the face or distal portion of the extremities, with occasional itching. The generalized form is rarer, affecting men and women equally in the fifth decade of life. It presents a centripetal distribution affecting the trunk, neck, and roots of the limbs, and has a tendency for spontaneous resolution. Here, we present a case of this unusual generalized variant.

Keywords: Multinucleated cell angiohistiocytoma; Fibro-histiocytic; Vascular proliferation

Abbreviations

HHV-8: Human Herpes Virus 8; MCAH: Multinucleated Cell Angiohistiocytoma; TBG: Thyroglobulin

Introduction

Multinucleate cell angiohistiocytoma is a rare benign mesenchymal, fibro-histiocytic, and vascular proliferation, described by Smith and Wilson-Jones in 1985. Dermoscopically, it is characterized by reddish-brown to purple papules and nodules located on the distal extremities and face. To date, only 150 cases of this entity have been reported, with a relative male-to-female incidence of 1:3, occurring in middle to older age. Furthermore, the generalized forms are extremely infrequent, with only 12 cases reported in the literature, showing a relative male-to-female incidence of 1:1, and a mean age of 42.75 years. In this report, we describe the clinical, histopathological, and immunohistochemical characteristics of a new case of the generalized type.

Methods

The study involves a detailed examination of the clinical, histopathological, and immunohistochemical characteristics of a 50-year-old male patient with a generalized form of multinucleated cell angiohistiocytoma.

Clinical Case

A 50-year-old male patient presented to our hospital in February 2020, with an 8-year history of multiple disseminated papules on his trunk and extremities, that began in 2012. The papules were asymptomatic, 2 to 3 mm in size, reddish-brown to purple, and firm in consistency (Figures 1 and 2). Physical examination was normal. He did not have any relevant personal or family medical history. The presumptive clinical diagnoses were morphea versus vascular proliferation. In August 2019, biopsies of the abdominal and thoracic skin were taken at another institution and were confusingly diagnosed as chronic dermatitis, psoriasiform and spongiotic, with lymphocytes and eosinophils. In December 2019, another clinic reviewed the samples and diagnosed them as dermal vascular capillary proliferation, with lympho-plasmacytic infiltrate and hemosiderosis. In February 2020, we received the samples, and histopathological study revealed an epidermis with slight acanthosis, irregular elongation of interpapillary ridges, and pigmentation of the basal layer. In the papillary and reticular dermis, a predominantly perivascular lymphocytic inflammatory infiltrate with isolated plasma and mast cells was observed, associated with numerous dilated and congestive vascular structures (Figures 3 and 4). Multinucleated cells were present, some of them emphasized by immunohistochemical staining for CD68 at the dermal level (more numerous in the abdominal

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***Correspondence:**
Greco Daniel Alejandro, Departamento de Patología, Hospital de Clínicas José de San Martín, Avenida Córdoba 2351, Ciudad Autónoma de Buenos Aires, Argentina,

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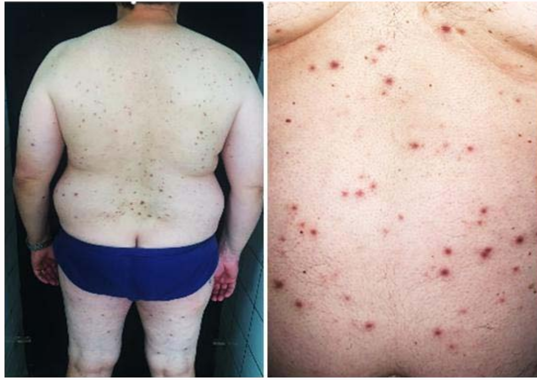

Figure 1 and 2: The papules were asymptomatic, 2 to 3 mm in size, reddish-brown to purple, and firm in consistency.

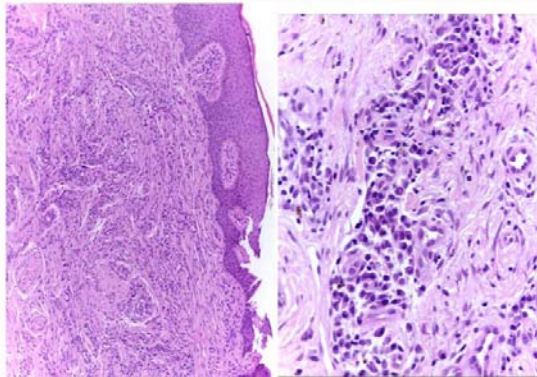

Figure 3 and 4: Predominantly perivascular lymphocytic inflammatory infiltrate with isolated plasma and mast cells was observed, associated with numerous dilated and congestive vascular structures.

skin biopsy), and mild fibrosis (Figures 5 and 6). Considering these findings, in correlation with the immunohistochemical results and clinical features, we concluded the diagnosis of a generalized multinucleate cell angiohistiocytoma.

Results

The histopathological study revealed an epidermis with slight acanthosis, irregular elongation of interpapillary ridges, and pigmentation of the basal layer. In the papillary and reticular dermis, a predominantly perivascular lymphocytic inflammatory infiltrate with isolated plasma and mast cells was observed, associated with numerous dilated and congestive vascular structures. Multinucleated cells were present, some of them emphasized by immunohistochemical staining for CD68 at the dermal level, and mild fibrosis.

Discussion

Multinucleated Cell Angiohistiocytoma (MCAH) is a rare entity, with 150 cases reported to date, occurring in middle-aged or elderly patients. Seventy-nine percent of cases are female, with a relative male-to-female incidence of 1:3. This is justified by some authors due to increased expression of Estrogen Receptor Alpha and Factor XIIIa in the affected cells. Clinically, it is characterized by single or multiple papules or nodules, reddish-brown to violet in color, ranging from 2 mm to15 mm in diameter, grouped into one or more anatomical regions, including the distal extremities, especially the dorsal hands, wrists, thighs, and legs. Many pathologists consider MCAH an inflammatory tumor rather than a true neoplasm because

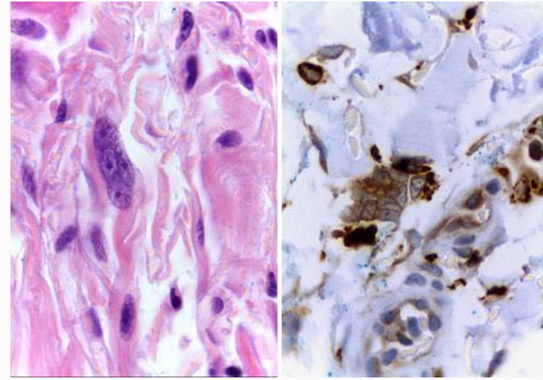

Figure 5 and 6: Multinucleated cells were present, some of them emphasized by immunohistochemical staining for CD68 at the dermal level (more numerous in the abdominal skin biopsy), and mild fibrosis.

the cells have a short half-life when cultured in other tissues, do not demonstrate the presence of HHV-8 (Human Herpes Virus-8), have no extracutaneous locations, do not meet histological criteria for malignancy, and do not undergo malignant transformation. Although some authors consider this entity a variant of dermatofibroma, it is important to differentiate them due to MCAH's tendency for spontaneous resolution, benign nature, and lack of malignancy potential. Cases associated with mycosis fungoides, vitiligo, and intradermal melanocytic nevus have been reported. Occasionally, solitary papules progress to fibrosis or multiple lesions, inversely proportional to CD68 expression, and exceptionally, multiple papules in a generalized distribution have been reported. Generalized MCAH is very rare, with 12 cases reported to date. Unlike its localized counterpart, it has an equal incidence in men and women, and a mean age of 42.75 years (24-64). The lesions are clinically similar to those described in the localized form, with most being asymptomatic; itching was reported in only two cases. They can have a variable duration from 6 months to 20 years and can present spontaneous remission. Of the 12 cases reported in the literature, one case was associated with POEMS syndrome and Castleman's disease; one case with vitiligo and TBG positive antibodies; one case with Anti-RO and ANF antibodies; one case with valve replacement for endocarditis, primary biliary cholangitis, diabetes, and dyslipidemia; and one case with multiple and severe abortions and early menopause. In summary, 50% of the cases presented some association with autoimmune phenomena, but while a probable relation can be inferred, the number of cases reported to date is insufficient to draw definitive conclusions. Based on its histological morphology, it is important to always make a differential diagnosis with angiofibroma, atrophic and vascular dermatofibroma, microvenular hemangioma, and early-stage Kaposi's sarcoma, because unlike these, MCAH tends to spontaneous resolution, is benign in nature, and has no malignancy potential. Although the histopathological findings are not pathognomonic for this entity, a correct and representative analysis of the sample, and a proper correlation with clinical and immunohistochemical findings, are sufficient for an accurate diagnosis [1-5].

Conclusion

Generalized multinucleated cell angiohistiocytoma is a rare variant of MCAH. Proper diagnosis requires thorough histopathological and immunohistochemical examination, in conjunction with clinical correlation.

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