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Linear and Bilateral Multinucleated Cell Angiohistiocytoma (MCAH)

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Abstract

Background: Multinucleated Cell Angiohistiocytoma (MCAH) is a rare disease, first described by Smith and Wilson Jones in 1985. Since then, less than 100 cases have been reported in the literature. Clinically it is characterized by papules or plaques arising from a specific anatomical area such as lower extremities, dorsum of the hands and face. Some generalized cases have been reported.

Main observations: We report a case of 77-year-old woman who presented with multiple itching. reddish to violaceous, flat to domed-shaped plaques on the lower legs with symmetrical and bilateral distribution along the saphena veins. On dermoscopy examination only a red-violaceous homogeneous area was visible.
Histology showed remarkable proliferation of dilated small vessels in the upper and mid dermis and bizarre-shaped multinucleate giant cells with scalloped cytoplasm that were intermingled with numerous mononucleated spindle cells. Many mast cells containing the characteristic granules were also detected, often adjacent to the multinucleate cells. Based on the clinico-pathologic findings the diagnosis of MCAH was established.

Conclusions: To our knowledge, this is the first documented case of MCAH with a bilateral and linear pattern disposed on the lower limbs, following the saphena veins. In this patient chronic trauma induced by ambulation might have contributed to development of the lesions. (*J Dermatol Case Rep*. 2016; 10(4): 58-61)

Introduction

Multinucleated Cell Angiohistiocytoma (MCAH) is a rare disease first described by Smith and Wilson Jones in 1985[1] and since then, less than 100 cases have been reported in the literature.[2-10]

MCAH is more frequent in elderly women than in men, suggesting that hormones can have a role in the pathogenesis of this entity. The typical clinical features consist of a cluster of red to violaceous papules or plaques, ranging in size from few millimetres to 1.5 cm, located on a single anatomical area as lower extremities, dorsum of the hands, wrists, fingers or face. The lesions can be arranged in an annular configuration or grouped in a cluster and are usually asymptomatic but may be rarely itchy. The clinical differential diagnoses may include dermatofibroma, angiofibroma, Kaposi sarcoma, pseudo-Kaposi sarcoma, granuloma anulare, lichen planus, pyogenic granuloma, giant-cell fibroblastoma, sarcoidosis and insect bite.

Case Report

We report a 77-year-old woman who presented with a 5-year history of multiple papules on the lower extremities. The patient reported that the first lesion appeared after a holiday and it was initially diagnosed as a mosquito bite. With time the lesion did not regress and new similar lesions occurred at the same site. Treatment with argon laser, performed by another institution, was unsuccessful. Physical examination showed multiple itching reddish to violaceous, flat to domed-shaped plaques on the lower legs with an unusual

symmetrical and bilateral distribution along the saphena veins (Fig. 1). The overall lesions count was 38 and the size varied from 6 to 13 mm. Clinical diagnoses included Kaposi sarcoma, dermatofibroma and angiofibroma.

On dermoscopy examination we observed no specific clues for the diagnosis. Only a red-violaceous homogeneous area is visible (Fig. 2).

Histological examination of three biopsy specimens showed a remarkable proliferation of dilated small vessels lined by prominent endothelial cells in the upper and mid dermis (Fig. 3 A-B). All around were scattered abundant large and basophilic, bizarre-shaped multinucleate giant cells with scalloped cytoplasm (Fig. 3C), that were intermingled with numerous mononucleated spindle cells. There was a moderate degree of dermal fibrosis, a few collagen bundles being almost completely hyalinized (Fig. 3C). Many mast cells containing the characteristic granules were also detected, often adjacent to the multinucleate cells (Fig. 3D). Discrete foci of interstitial mucin and a sparse lymphohistiocytic infiltrate were found as well.

Immunohistochemistry showed that both multinucleate cells and mononuclear spindle cells stained positively for CD68, while factor XIIIa was found only in the mononuclear cells.

Based on the clinico-pathologic findings the diagnosis of MCAH was made.

The therapeutic options were discussed with the patient but finally the decision to wait and see was taken based on the possible spontaneous regression of cutaneous lesions. Treatment with HI antihistamine receptors was prescribed for few months for the pruritus with good result. The patient came back after 6 months and the lesions were still present, but without any new lesions.

Discussion

The pathogenesis of MCAH is still unknown and it is still controversial if MCAH s a genuine neoplasm or a reactive condition. Nevertheless its benign course, its tendency to present as multiple lesions in areas exposed to trauma and arthropod bites or stings, the absence of clonality, and cases of spontaneous regression would all support a reactive process.[11]

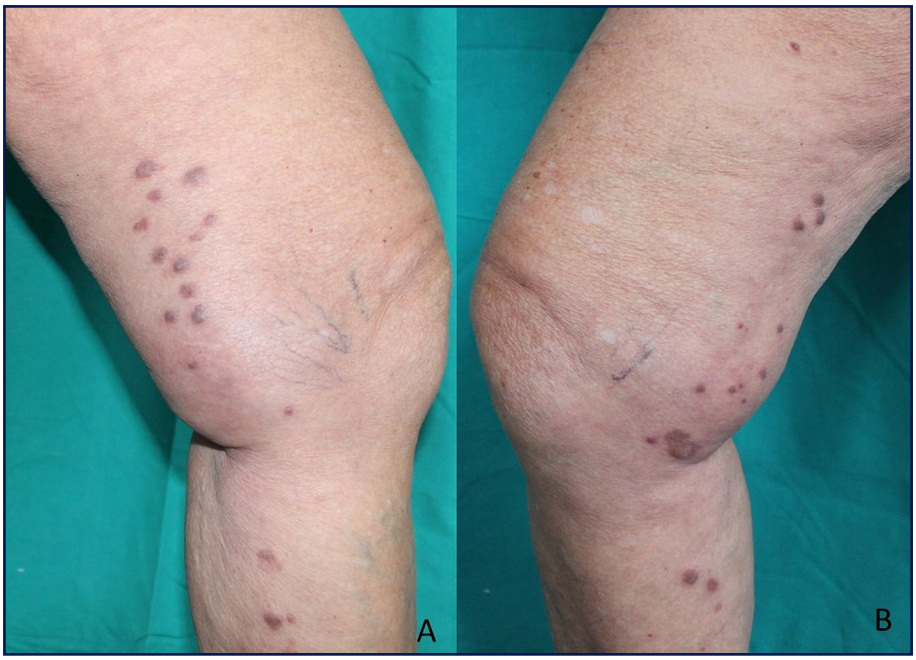

Figure 1

(A-B): Bilateral and simmetrical distribution of red to violaceous papules of MCAH, localized along the flow of saphena veins in a 77-years-old lady.

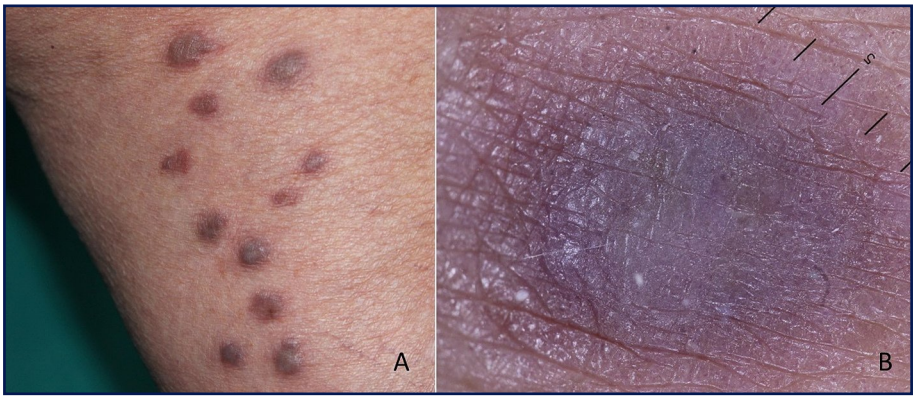

Figure 2

(A) Magnification of a clinical appearance of red papules of MCAH. (B) Dermoscopic imagine of MCAH.

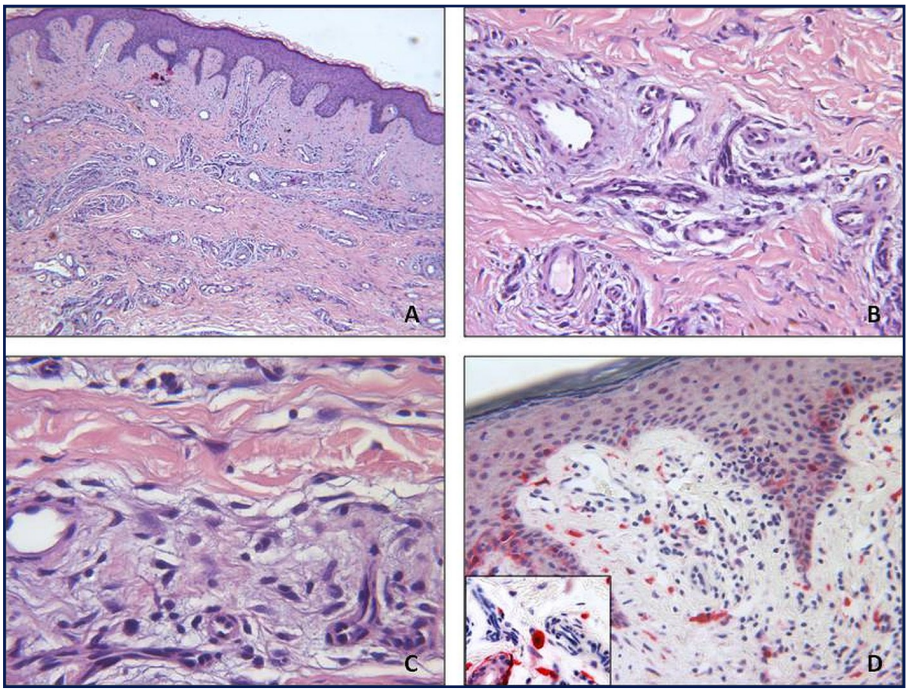

Figure 3
(A-B) Histologically, there was an increased number of small ectatic vessels in the superficial and mid dermis.
(C) High power view of a few multinucleate giant cells finely interspersed among thick collagen bundles.
(D) Strong cytoplasmic positivity for mast cell tryptase.

The characteristic giant multinucleated cells are derived from fibroblasts or macrophages, which in conditions of chronic stimulation, lose their ability to divide after mitosis and so become inactive.[12]

Some Authors consider MCAH to be a variant of dermatofibroma with a prominent vascular component and bizarre multinucleated cells, but for many authors the combination of clinical and histologic features indicates that it is a discrete entity.[13]

The estrogen receptor alpha is overexpressed in MCAH. According to the authors these findings underline the pathogenetic difference between MCAH and dermatofibromas and also corroborate the hypothesis of a role played by hormones in the development of this lesion.[14] Nevertheless the immunochemistry for ERalpha was negative in our case

A significantly increased number of mast cells was evident in our case, in which mast cells were characteristically found immediately adjacent to multinucleate cells. A specific role played by mast cells in the morphogenesis of multinucleated cells through mechanisms of degranulation and interaction with fibroblasts has been postulated by many authors. Puig *et al*.[7] consider that the dermal angiogenesis and multinucleated giant cells formation processes are secondary to the release of cytokines by mononucleated fibrohistiocytic cells and mast cells present in the lesions. In particular they propose that the mast cells degranulation could be implicated in the upregulation of factor XIIIa+ fibrohistiocytic cells which are the main proliferating cell population in MCAH. From an histological point of view we find many similarities between MCAH and cutaneous mastocytosis.

The MCAH lesions are usually located in a single anatomical area although, more rarely, they can be generalized.[11,15] The generalized variant is extremely rare, with only four cases recorded in the literature. Our patient present a localized form, with almost 40 lesions distributed exclusively on the lower limbs.

In our case the papules were unusually arranged following the saphena veins in a symmetrical linear pattern (Fig. 1-2) supporting the hypothesis that chronic trauma induced by ambulation might have been the primum movens for the origin of the lesions. The cytokines released from the endothelium in response to a physical stimulus or in a chronic inflammatory condition such as chronic venous insufficiency could have induced the fibroblasts to proliferation.

Treatment include argon laser, surgical excision or cryosurgery although it is not mandatory since the lesions may spontaneously regress.[16] As mentioned before in our case we not did any physical treatment considering that the patient had multiple lesions and that in the past years she was treated with argon laser without any benefit; we only gave her some HI antihistamine receptors for the itch with good result.

Conclusions

To our knowledge, this is the first documented case of MCAH with a linear pattern disposed bilaterally on the lower limbs. We think it might be interesting to further understanding the clinical presentation of the disease.

In the future further investigations would be needed to better understand the pathogenesis of this entity and also to develop new treatment.

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