GIANT CELL ANGIOHISTIOCYTOMA

ALEXANDRU OANȚĂ*, AMBROS MIRCEA**, TIBERIU TEBEICĂ***, OANA TIUCĂ*,**, ****, SILVIU HORIA MORARIU**,****, SMARANDA ȚĂREAN*

Summary

Introduction: *Multinucleate cell angiohistiocytoma (MCAH) is a rare benign vascular tumor.*

Clinical case: *A 32-year-old patient is examined for multiple asymptomatic brownish papules, ranging in size from 2 to 4 mm, located on the forehead. Histopathological examination revelead a normal epidermis, with a rich dermis vascularization and with the presence of mononucleate histiocytes and rare multinucleate giant cells. Immunohistochemistry was positive for CD31, CD34, factor VIII, CD68, factor XIIIa and vimentin. A diagnosis of MCAH was established.*

Discussions: *MCAH represents a tumor due to a benign vascular proliferation. It affects mostly women and it presents as multiple papules located mainly on the dorsal side of the hands. The etiopathogenesis of MCAH remains unknown, suggesting the fibriblastic origin of this tumor. The slow spread and localised evolution of this tumor makes the treatment unnecessary.*

Conclusion: *The diagnosis of MCAH is mainly useful in excluding other disorders, particularly Kaposi sarcoma.*

Keywords: *angiohistiocytoma, histyocite, multi-nucleate cells.*

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Introduction

Multinucleate cell angiohistiocytoma (MCAH) is a rare benign vascular tumor, first described by Smith and Wilson-Jones in 1985 [1].

Clinical case

We present the case of a 32-year-old male patient who was examined for cutaneous lesions located on the forehead. Anamnesis did not reveal the use of any drugs. Clinical examination identified slightly elevated multiple asymptomatic brownish papules, ranging in size from 2 to 4 mm. Apart from that, skin and mucosal examination, as well as clinical examination were normal.

Histopathological examination revelead a normal epidermis. The dermis presented capillary hyperplasia, associated with lymphohistiocytic infiltrate and rare dispersed multinucleate giant cells, with angular aspect and clout-shaped nuclei. Immunohistochemistry was positive for CD31, CD34, factor VIIIa, CD68, factor XIIIa and vimentin.

The clinical and histopathological aspects established the diagnosis of MCAH. The patients refused, for the time being, treatment with cryotherapy.

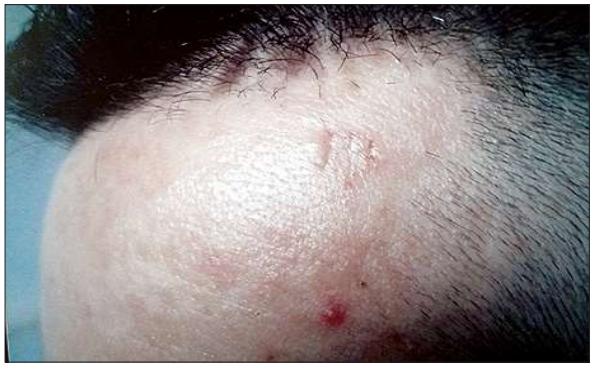


Figure 1. Clinical appearance of MCAH. Multiple brown papules located on the forehead.

* SC Dermamed SRL, Brașov
** Clinical Hospital Country Mureș, Dermatovenerology Clinical
*** Dr. Leventer Center, Bucharest
**** UMFST "G.E.Palade" Târgu-Mureș, Dermatology Discipline

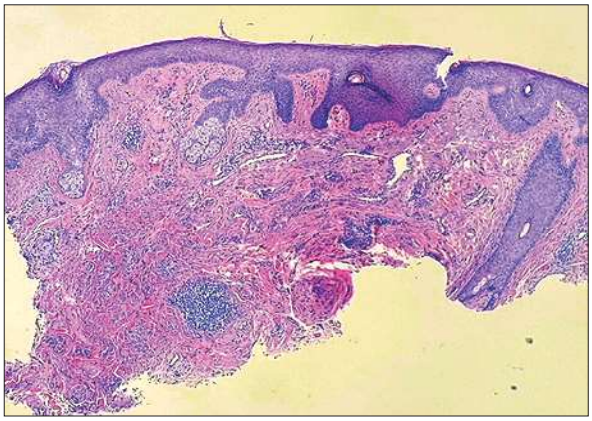
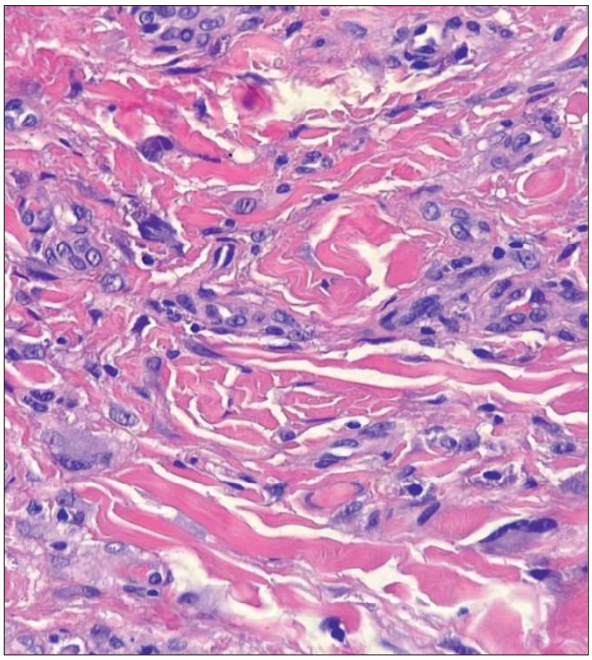
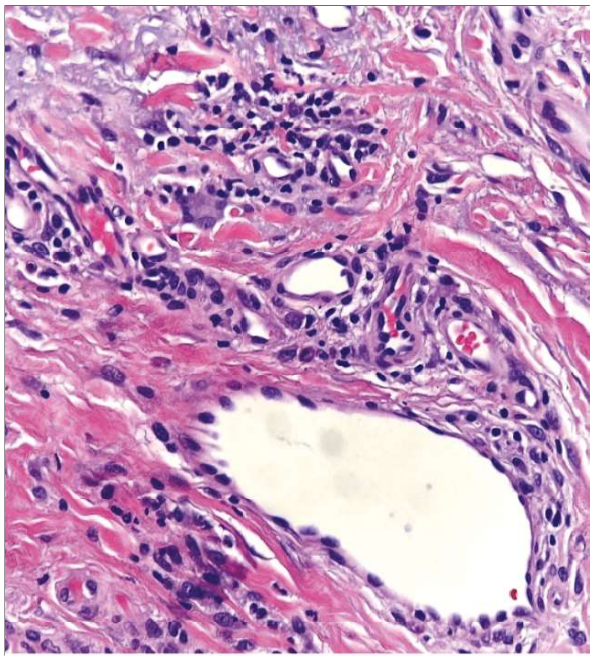


Figure 2. Histopathological aspect. Normal epidermis. In the dermis can be observed a vascular hyperplasia and the presence of a rare lymphohistiocytic infiltrate. Hematoxylin eosin stain, 10x magnification.



Figures 4. Histopathological aspects. In the dermis, capillary hyperplasia associated with lymphohistiocytic infiltrate and rare dispersed multinucleate giant cells. Hematoxylin eosin stain, 40x magnification.



Figures 3. Histopathological aspects. In the dermis, capillary hyperplasia associated with lymphohistiocytic infiltrate and rare dispersed multinucleate giant cells. Hematoxylin eosin stain, 40x magnification.

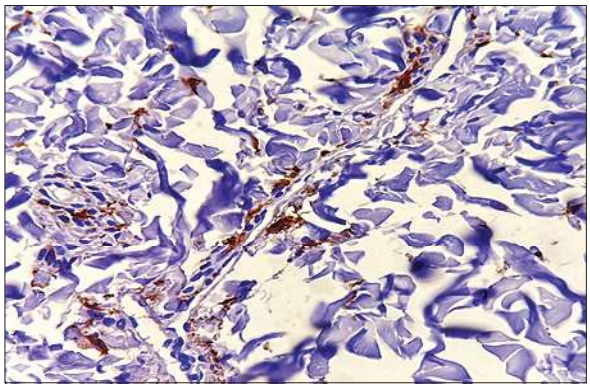


Figure 5. Immunohistochimistry. Positivity for factor XIIIa.

Discussions

MCAH affects mainly middle-aged women, with male/female sex ratio of 1/3. Clinically, the lesions present as asymptomatic and slightly elevated erythematous, brown, or purplish papules, with smooth and shiny surface, most often encountered on the limbs, especially acral, but also on the dorsal side of the hands, face, trunk, and mucous membranes. They are usually unilateral, rarely bilateral or generalized. The lesions progressively evolve through weeks or months and can reach a medium size from 2 to 15mm and can rarely regress spontaneously. Clinical differential diagnosis should be performed with Kaposi sarcoma (for those cases with big, grouped papules), annular granuloma, angiofibromas, dermatofibromas, microvenular hemangioma, lichen planus, lymphocytoma and insect bites. [2]

The diagnosis is confirmed by histopathological examination, which reveals a small vessels proliferation associated with discrete lymphohistiocytic infiltrate and the presence of multinucleate cells [1,3,4]. Histopathological differential diagnosis should be made with Kaposi sarcoma, dermatofibroma, angiofibroma, cutaneous lymphocytoma, lichen planus and angiolymphoid hyperplasia with eosinophilia. Immunohistochemistry is positive for factor VIII, CD31 and CD 34, endothelial cells expression, and for factor XIIIa, MAC-387, CD68, macrophages and histiocytes expression, as well as for vimentin. Several authors consider that MCAH belongs to the spectrum of dendrocyte proliferation disorders, which express factor XIIIa, the same as angiofibromas and dermatofibromas [3,4].

The etiopathogenesis of MCAH remains unclear. Most authors suggest rather an inflammatory origin than a tumoral one for this disorder [5,6,7]. Hormonal influence should also be considered, considering the identification of α estrogen receptors in female patients. [5,6]. Frew [8] considered that MCAH has an inflammatory and vascular debut, followed by fibrosis and atrophy. The slow and localized evolution of the lesions make the treatment often unnecessary. Treatment is performed mainly due to esthetic reasons. Intralesional corticosteroid injection, surgical excision, cryotherapy, argon, pulsed dye, and CO2 lasers can be considered [5,6].

Conclusion

The diagnosis of MCAH is difficult, this disorder being underestimated. The importance of a proper diagnosis resides in excluding other diseases, particularly Kaposi sarcoma.

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Conflict of interest
NONE DECLARED

Correspondance address: Oana Tiucă
Clinical Hospital Country Mureş, Dermatovenerology Clinical
12 Gheorghe Doja street, Târgu-Mureş
e-mail: oanaa.marginean@yahoo.com