CASES FOR DIAGNOSIS

Erythematous Papules on the Upper Chest Wall

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Clinical History

The patient was a 51-year-old woman with no past medical or surgical history of interest. She was seen in our outpatient clinic for asymptomatic lesions that had appeared on her chest 1 year earlier and that had remained stable in recent months. She denied any history of trauma in the area and there were no other symptoms on systems review.

Physical Examination

Physical examination revealed multiple, slightly infiltrated, erythematous-violaceous papules with a diameter of 2 to 7 mm, grouped in the left anterosuperior region of the chest wall (Figure 1).

Additional Tests

Routine blood tests were performed, which showed no abnormalities of interest. Biopsy was taken from one of the lesions for pathologic study.

Histopathology

The pathologic study showed a proliferation of vascular structures in the dermis, mostly capillaries with prominent endothelia, together with marked stromal cell hyperplasia and the presence of multinucleated giant cells, frequent mast cells, lymphocytes, and plasma cells (Figures 2 and 3). On immunohistochemical analysis, the vascular structures were positive for CD34 and the stromal cells for factor XIIIa and CD68.

What Was the Diagnosis?

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Figure 1.



Figure 2. Hematoxylineosin, ×20.



Figure 3. Hematoxylineosin, ×60.

Diagnosis

Multinucleate cell angiohistiocytoma.

Clinical Course and Treatment

Treatment with carbon dioxide laser was proposed but was rejected by the patient. It was therefore decided to keep her under observation, and the lesions remained stable in number and size over 2 years of follow-up.

Discussion

Multinucleate cell angiohistiocytoma is a rare condition first described in 1985 by Smith and Wilson-Jones.¹ It is most common in middle-aged women and preferentially affects the limbs, particularly the dorsum of the hands, the wrists, and the thighs, although also it has been reported on other sites such as the face and chest.²

Clinically it is characterized by multiple, asymptomatic, slow-growing, erythematous-violaceous papules or plaques of 2 to 15 mm in diameter and of firm consistency. They are usually grouped in a single anatomic region. Bilateral lesions are rare and there has only been 1 case report of a patient in whom the lesions were generalized.³

Histology reveals a dermal proliferation of capillaries and venules with prominent endothelial cells, associated with a lymphohistiocytic infiltrate and geometric multinucleated giant cells with angulated cytoplasm.²⁻⁷ The interstitial mononuclear cells express factor XIIIa, CD68, lysozyme, α 1-antitrypsin, and vimentin.³⁻⁵ The multinucleated cells are positive for vimentin and the vascular component stains for factor VIII and CD34, among others.³

The clinical differential diagnosis includes various conditions such as Kaposi sarcoma,^{2,6} lichen planus, lymphocytoma, insect bites, and granuloma annulare. The main conditions included in the histological differential diagnosis are dermatofibroma and, particularly, atrophic vascular histiocytoma. Other conditions in which multinucleated cells can appear, such as fibrous papules on the face, must also be considered.⁴

Multinucleate cell angiohistiocytoma is a benign vascular proliferation.¹⁻⁷ It is unclear whether it is a reactive disorder or a true neoplasm; however, due to the multiple and eruptive nature of the lesions, their tendency to develop in exposed areas, and the absence of clonality and mutations, the majority of authors consider it to be a reactive condition.² A possible hormonal influence has also been suggested due to its predominance in women.³ Another controversy about multinucleate cell angiohistiocytoma concerns the histogenetic origin of this type of cell, as immunohistochemical and ultrastructural analyses give varying results, suggesting a fibroblastic origin in some cases and a histiocytic origin in others.^{3,6}

Multinucleate cell angiohistiocytoma has a benign course, with a tendency to persist or to show slow progression in the majority of cases; treatment is therefore usually unnecessary except for cosmetic reasons. In such cases, good results have been reported with cryotherapy,³ argon laser, and carbon dioxide laser.⁷

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