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CASE FOR DIAGNOSIS

Long-Standing Macular Lesions on the Face

Lesiones maculosas de larga evolución en la cara

Medical History

The patient was a 69-year-old man who consulted for asymptomatic lesions on the left cheek that had been present for 10 years and that had gradually increased in size. The lesions occurred on the site of an electrical burn received 30 years earlier.

Physical Examination

Physical examination showed a group of slightly elevated, infiltrated grayish-brown lesions on the left cheek and temple (Figure 1). The lesions were irregular and had poorly defined borders.

Histopathology

Histopathology showed a proliferation of small diameter vessels in the superficial and middle dermis with rounded or long lumina and lined by endothelial cells with prominent nuclei. The surrounding dermis presented marked edema and a mild superficial perivascular infiltrate of lymphocytes, histiocytes, and plasma cells (Figure 2A). There was a noticeable presence of multinucleated giant cells of varied morphology and angular outline between the collagen bundles (Figure 2B). The epidermis was normal. Alcian blue stain revealed no fibrous tissue proliferation or mucin deposits. Immunohistochemistry was positive for vimentin in the multinucleated giant cells. There was intense staining for CD68 and factor XIIIa in the dermal mononuclear cells (Figure 3) and staining for CD31 in the endothelium of the vessels.

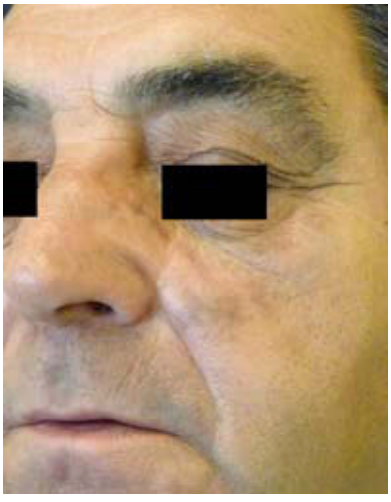

Figure 1

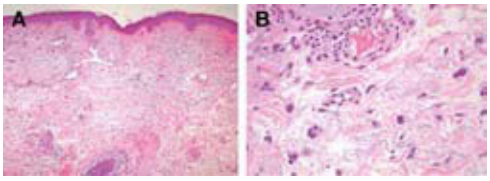

Figure 2 A, Hematoxylin-eosin, original magnification ×250. B, Hematoxylin-eosin, original magnification ×400.

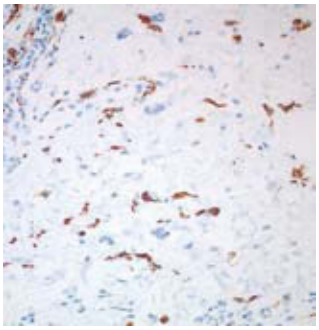

Figure 3 Vimentin, original magnification ×400.

What Is Your Diagnosis?

Diagnosis

Multinucleate cell angiohistiocytoma (MCA).

Clinical Course and Treatment

No treatment was prescribed as the lesions were benign and asymptomatic.

Comment

MCA is a rare clinical and anatomic entity that tends to occur in middle-aged women, preferentially in exposed areas of skin such as the dorsum of the hands, the wrists, or thighs.[1-3] It has also been reported at other sites including the face, thorax, upper lip, and oral mucosa.[1] It presents in the form of raised erythematous-violaceous lesions that are typically grouped in a single anatomic region. Bilateral involvement is rare[4] and only 1 case has been reported of generalized lesions. MCA is usually asymptomatic and shows slow progression over the years.

Some authors class MCA among the dermatofibroma-like proliferations of dermal dendritic cells as it is difficult to determine whether these are reactive disorders or true neoplasms. Most authors conclude MCA is a reactive disorder as it tends to occur as multiple eruptive lesions located in areas exposed to trauma, and there have been some reports of spontaneous resolution.[3] Lesions histologically identical to MCA have recently been identified in continuity with other neoplastic or reactive disorders, and Calderaro et al[5] suggested the lesions may be induced by scarring and chronic inflammatory conditions.

The diagnosis is based on histopathology. The superficial and middle dermis show a proliferation of capillaries and venules with prominent endothelial cells that are positive for CD31, CD34, and factor VIII. The surrounding dermis contains a lymphohistiocytic infiltrate formed of plasma cells and irregular multinucleated giant cells with angular cytoplasm. The mononuclear cells express factor XIIIa, CD68, lysozyme, α1-antitrypsin and vimentin. The multinucleated cells usually express only vimentin but may also stain positive for serum markers of monocyte/macrophage activation.[3] An increased presence of mast cells has been described close to the multinucleated giant cells, with speculation that these may be implicated in the pathogenesis of the disorder by stimulating vascular hyperplasia and a proliferation of fibrohistiocytic cells.

The clinical differential diagnosis basically includes Kaposi and pseudokaposi sarcomas. The vascular nature of the lesions is not always evident clinically and, in cases like ours, brownish coloring may predominate. In these cases, the differential diagnosis must include lichen planus, sarcoidosis, lymphocytoma, granuloma annulare, and lupus erythematosus.[3]

The benign nature of the lesions means no treatment is necessary although good outcomes have been reported with surgical excision, cryotherapy, argon laser, carbon-dioxide laser, and intense pulsed-light therapy.[6]

Conflict of Interest

The authors declare that they have no conflict of interest.

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