JAMA Dermatology Clinicopathological Challenge

Bilateral Round Scar-like Lesions on the Face of a Young Man

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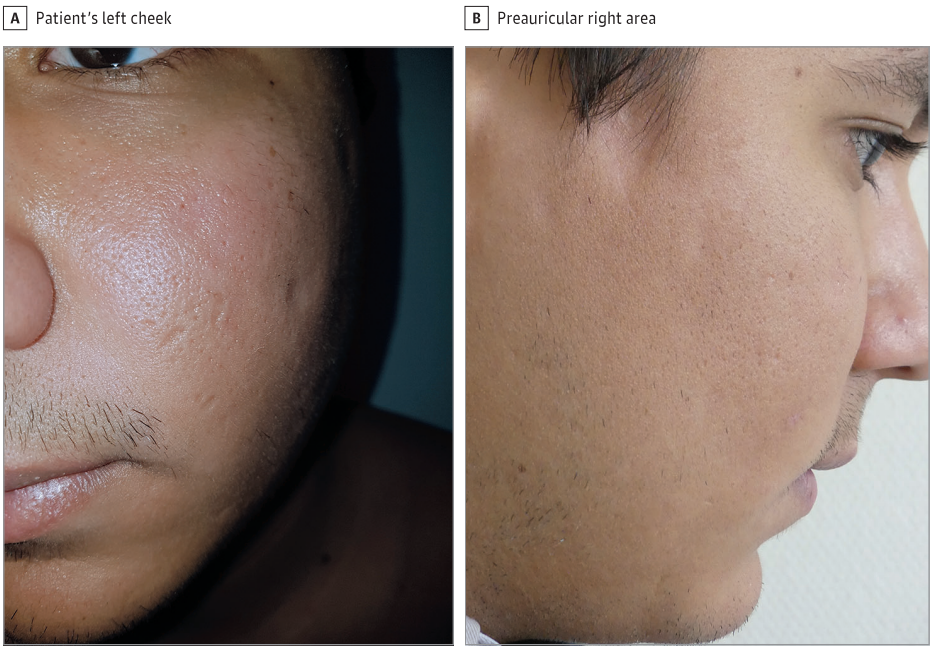

Figure 1. Atrophic lesions on the patient's left cheek (A) and with a cicatricial appearance on the preauricular right area (B).

A 26-year-old man presented with a several-year history of persistent round lesions with a scar-like appearance on his face. Before evaluation, his condition had been diagnosed as lupus and lipoatrophy and he tried several treatments with no success.

The results of his physical examination showed several round atrophic lesions on his face, symmetric and bilateral, that were located especially on the malar and preauricular areas, with a diameter of 5 to 10 mm. His skin lesions were not indurated and displayed no epidermal changes (**Figure 1**). The patient denied experiencing previous inflammation. He had no history of chicken pox or acne or traumatic lesions in those areas. New punch biopsy specimens were obtained for further evaluation.

WHAT IS YOUR DIAGNOSIS?

A. Primary anetoderma

B. Atrophoderma vermiculata

C. Atrophia maculosa varioliformis cutis

D. Facial morphea

Diagnosis

C. Atrophia maculosa varioliformis cutis

Discussion

Atrophia maculosa varioliformis cutis (AMVC) was first described in 1918 by Heidingsdfield and since then approximately 20 cases have been reported.[1] It is considered a rare disease that appears spontaneously and is not preceded by other inflammatory or traumatic events. From a clinical point of view, this entity is characterized by the progressive appearance of sharp depressions of various shapes, often round or oval, with regular margins and clear-cut edges, which are all asymptomatic. Its etiology remains unclear, although its family clustering suggests that it could be a genetic disorder.[2] The histological findings are variable and nonspecific, including a thinning of the horny layer or the entire epidermis, decreased or fragmented elastic fiber, and a lymphocytic perivascular infiltrate.[1-3]

The differential diagnosis includes scars that are related to acne, chicken pox varicella, dermatitis artifacta, atrophoderma vermiculata, facial morphea, or primary anetoderma. Atrophoderma vermiculata is a rare condition that is considered to belong to the keratosis pilaris group, characterized by smaller lesions on the cheeks, which is presumably a late reaction to inflammation around the horny

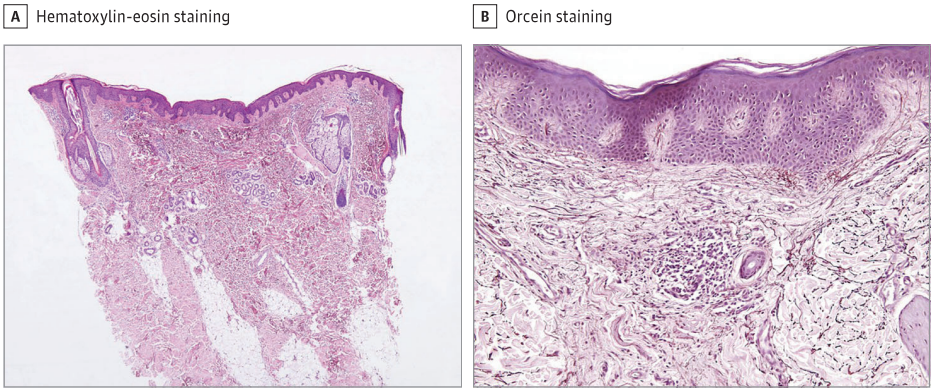

Figure 2. A, Panoramic histological view shows a slight depression of the epidermis (hematoxylin-eosin; original magnification, ×2). B, Orcein staining allows a demonstration of the reduction in the elastic fibers beside the lymphocytic perivascular infiltrate compared with the surrounding and healthy area (orcein; original magnification, ×10).

plugs. The honeycomb-like atrophy on the cheeks of atrophoderma vermiculata helped to exclude this entity in the patient. Facial morphea usually shows an active erythematous or violaceus inflammatory border. Primary anetoderma is a rare idiopathic dermatosis that is characterized by areas of flaccid skin located in extremities and trunk due to the loss of elastic tissue in the absence of a secondary cause.

There is no standardized treatment for patients with AMVC, although filler injections, dermabrasion, or laser resurfacing may theoretically be helpful in improving the facial scarring.[3,4] Although there are no follow-up data reported in most previous cases, it is important to explain to patients with AMVC that it is a benign and nonprogressive dermatosis.

Microscopic Findings and Clinical Course

The histopathologic examination results showed a significant epidermal dell and a superficial dermal lymphocytic infiltrate that was associated with dermal fibrosis. An orcein stain demonstrated a reduction of elastic fibers in the dermis beneath the epidermal dell (**Figure 2**). The patient's skin lesions remain stable after 1 year of follow-up.

ARTICLE INFORMATION

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