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INTERESTING MEDICAL IMAGE

Atrophoderma of Pasini and Pierini

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ضمور الجلد بحسب بازيني وبيريني

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Figure 1: Photograph of multiple round, pink-coloured, atrophic plaques on the back of a 25-year-old male patient.

family history of interest attended the outpatient clinic of the Complejo Hospitalario de Granada, Granada, Spain, in 2017 with multiple, non-pruritic, atrophic, mildly indurated, brownish, erythematous patches on his back of one year's duration [Figure 1]. He denied any regular medication use or having had any previous conditions. There was no history of trauma preceding the appearance of the skin lesions. Routine laboratory tests were within normal limits, including a complete blood cell count, urinalysis, thyroid profile, protein electrophoresis and general biochemistry, antinuclear/extractable nuclear antibody and rheumatoid factor tests. Immunoglobulin M and G tests for *Borrelia burgdorferi* were negative.

A histopathological examination showed a normal epidermis with a slight mononuclear perivascular infiltrate in the papillary dermis and focal sclerosis with hyalinisation of the reticular dermis [Figure 2]. Narrowband ultraviolet B phototherapy was proposed as the first line of treatment; however, the patient refused to consent to the therapy.

Comment

Idiopathic atrophoderma of Pasini and Pierini (IAPP) is a rare, exclusively cutaneous condition which occurs most frequently in female patients, with a peak incidence in the second and third decades of life.¹ Overall, fewer than 100 cases have been reported

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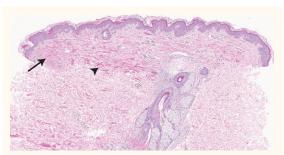


Figure 2: Haematoxylin and eosin stain at x4 magnification showing a normal epidermis with discrete thinning of the upper dermis (arrow) and mild hyalinisation and sclerosis on the reticular dermis (arrowhead).

in the literature.2 The pathophysiology of this entity remains unknown and the role of Borrelia burgdorferi infections remains controversial.2 Some researchers believe that IAPP should be included as a manifestation of the spectrum of localised scleroderma.³ Some cases of familial IAPP have been described; however, a genetic link has not yet been confirmed.2

From a clinical point of view, IAPP lesions may present in various patterns (i.e. rounded, oval or circular) and may be single or multiple.3 The edges of the patches are usually well defined and multiple lesions, such as in the present case, have been described as having a 'swiss cheese' or 'footprints in the snow' appearance. The sites predominantly affected are the lower and upper extremities or the trunk and it is not unusual for the lesions to follow a zosteriform or blaschkoid pattern.3 The condition can cause pain, pruritus or even paresthesia.

The pathological findings of IAPP are variable. In many cases, no alterations can be observed in the epidermis or dermis.^{2,3} The most common finding is a thinning of the affected dermis and no quantitative or qualitative alterations have yet been reported on the appendages.³ Collagen changes can include atrophy, sclerosis, fragmentation and hyalinisation.² Elastic fibre changes may be variable and reduction and fragmentation similar to that of anetoderma cases have been reported.4

The therapeutical approach to IAPP includes the use of different medications such as antibiotics (i.e. penicillin and tetracyclines) and hydroxychloroquine (with good response in cases with associated lupus) as well as topical treatments such as calcineurin inhibitors (although with variable responses).5-7 Treatment with a Q-switched Alexandrite laser has also been reported to result in the clinical improvement of hyperpigmented lesions.8

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