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CASE FOR DIAGNOSIS

Long-Standing Widespread Purpura

Púrpura extensa de largo tiempo de evolución

Clinical History

A 47-year-old man reported slightly pruritic lesions that had developed 8 years earlier. The lesions had commenced in both axillas, had subsequently spread to the abdomen and thighs, and in the last few months had appeared on the dorsum of the feet (Figures 1 and 2). They had been biopsied on 3 occasions and diagnosed successively as nonspecific purpuric dermatosis, lichen aureus, and chronic pigmented purpuric lichenoid dermatosis. Several courses of treatment with topical corticosteroids and tacrolimus had been prescribed, with only partial improvement during use and subsequent progression once treatment had been suspended.

Physical Examination

The incipient lesions on the dorsum of the feet were small purpuric petechiae that did not blanch under pressure and tended to become confluent. The fully established lesions of the abdomen were well-defined brown spots that did not affect the orifices of the hair follicles and showed no superficial desquamation.

Additional Tests

The blood test showed no abnormalities in coagulation parameters or in the complete blood count.

Histology showed a moderate lichenoid lymphocytic infiltrate in the papillary dermis that was positive for immunohistochemical markers CD4 and CD8 and negative for CD20, with red blood cell extravasation (Figure 3).

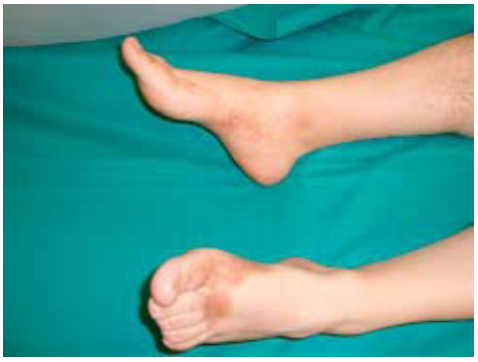

Figure 2

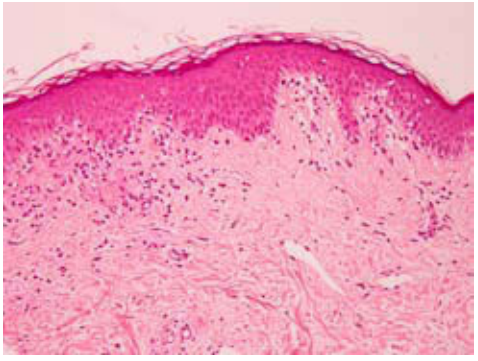

Figure 3 Hematoxylin-eosin (original magnification ×40).

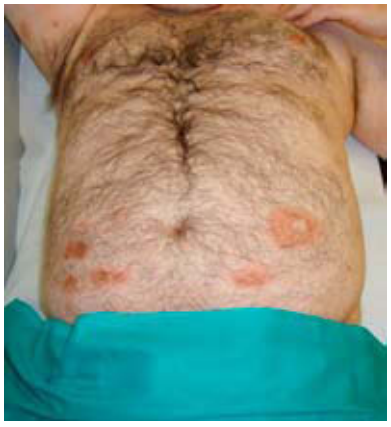

Figure 1

What is your diagnosis?

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Diagnosis

Pigmented purpura-like variant of mycosis fungoides

Treatment

Psoralen-UV-A

Comments

Pigmented purpura-like mycosis fungoides is a rare variant of mycosis fungoides that was first described by Bazin in 1876. It shares many clinical, histological, and genotypic features with chronic pigmentary purpura (CPP).[1]

The diagnosis of CPP encompasses several conditions which, like pigmented purpura-like mycosis fungoides, are characterized clinically by petechiae and bronze discoloration of the skin beginning on the lower limbs and histologically by a lymphocytic infiltrate with red blood cell extravasation and hemosiderin deposits.

The relationship between pigmented purpura-like mycosis fungoides and CPP is not entirely clear, and there are many reports of cases of CPP that have eventually progressed to mycosis fungoides.[2-4]

Due to these difficulties in distinguishing the 2 disorders, several authors have looked for the presence of a series of clinical, histological, and genotypic criteria that could help to differentiate between them, or at least to indicate which patients should be followed more closely.

Clinically, Lipsker et al [3] suggested that patients with extensive, long-standing (more than a year) CPP with a reticular morphology, and in which patch tests to rule out other causes are negative, should be followed up closely due to an increased risk of progression to mycosis fungoides.

Histologically, Toro et al,[5] after studying a series of 56 patients diagnosed with CPP, described 3 patterns —psoriasiform lichenoid, psoriasiform spongiotic, and atrophic lichenoid—that were common to both entities. They also observed that edema in the papillary dermis and greater red blood cell extravasation was more characteristic of CPP and that an increased presence of atypical lymphocytes and epidermotropism was associated with mycosis fungoides.

From a molecular point of view, it has been suggested that monoclonal cases are more likely to be mycosis fungoides and with CPP showing extensive cutaneous involvement and a predominantly CD4-positive atypical lymphocyte infiltrate. Polyclonal cases, on the other hand, are not usually associated with mycosis fungoides and show a predominantly CD8-positive lymphocyte infiltrate.[6]

In the case we describe, the patient presented none of the risk factors associated with the onset of CPP, such as the use of certain drugs or venous stasis; the lesions were more extensive than those described for this condition and the infiltrate, which was consistent with mycosis fungoides, showed a monoclonal rearrangement.

In conclusion, long-standing widespread pigmentary purpura should be closely followed, as it may be the initial manifestation of mycosis fungoides.

Conflict of Interest

The authors declare that they have no conflict of interest.

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