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Livedoid vasculopathy in a patient with bullous pemphigoid and primary Sjögren's syndrome☆

Vasculopatía livedoide en una paciente con penfigoide ampolloso y síndrome de Sjögren primario

Dear Editor,

Livedoid vasculopathy is a chronic, recurrent and painful skin disease that usually affects the lower limbs. It was first described by Bard and Winkelmann in 1967,[1] as a vaso-occlusive disorder affecting the small vessels of the dermis.

We present the case of an 82-year-old woman under dermatological follow-up for bullous pemphigoid, which was stable on low doses of prednisone. Of interest in her personal history was a prior diagnosis of primary Sjögren's syndrome with positive anti-Ro antibodies, meeting the diagnostic criteria set established by Vitali et al.[2]

During a check-up visit, coinciding with more xerophthalmia and xerostomia than usual, reticulated erythematous macules of livedoid appearance together with painful ulcerated nodules and other areas of atrophie blanche were observed on both lower limbs bilaterally and symmetrically (Fig. 1).

Due to a clinical suspicion of livedoid vasculopathy, a diagnostic biopsy was undertaken showing thickening and hyalinisation of the vessel walls, with no inflammatory component.

A complete blood test highlighted: anaemia with a haemoglobin of 10.4 g/dl, positive ANA 1/1280 with speckled cytoplasmic patterns, positive anti-SSA/Ro antibodies (>240.0 U/ml), reduced C3 and C4 (43 and 3 mg/dl, respectively). The remaining parameters showed no anomalies.

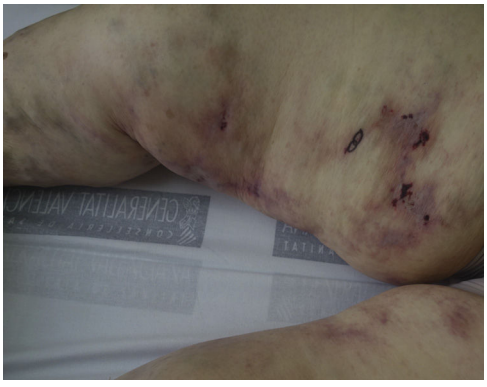

Fig. 1. Clinical image of the skin lesions.

Therefore, given the characteristic clinical skin symptoms and compatible histological findings, the diagnosis of livedoid vasculopathy was confirmed, and possible haematological alterations that would explain a prothrombotic condition were ruled out.

Treatment was started with pentoxifylline 400 mg every 8 h, with 100 mg of aspirin daily, achieving a good response and gradual resolution of the lesions.

Livedoid vasculopathy is a rare, chronic and painful disease, characterised by the presence of macules or purpuric papules and plaques with a tendency to form irregular ulcers that develop into star-shaped atrophic scars and peripheral hyperpigmentation, described as atrophie blanche.[3] It usually affects the lower limbs, with a bilateral, symmetrical distribution. It is characteristically, although not always, associated with livedo reticularis.

It can manifest at any time of life, and is more frequent in young women, with a 3:1 ratio over males.[3] However, our patient was older than the average.

The most frequent histopathological finding is hyalinising vascular changes of the inner layers of the dermal vessels, generally with little inflammation, together with thrombosis inside the blood vessels.[4] These signs enable the diagnosis to be confirmed, and other processes that present with similar skin symptoms to be ruled out.[5]

The condition's aetiopathogenesis remains unknown. However, the presence of thrombophilic alterations is considered increasingly relevant, and complementary tests are needed to rule out prothrombotic conditions.

In turn, it has been related to systemic diseases, such as scleroderma, systemic lupus erythematosus, rheumatoid arthritis, cryoglobulinaemia, and mixed connective tissue disease.[6,7] However, only one case relating to Sjögren's syndrome[8] has been published, and its simultaneous coexistence with two autoimmune conditions has not been recorded to date.[9]

To conclude, we present a case of livedoid vasculopathy in a patient with bullous pemphigoid and primary Sjögren's syndrome with positive anti-Ro antibodies and complement consumption. The co-existence of this disease with both autoimmune conditions reinforces the hypothesis that there is an association with systemic diseases that can be aggressive to the endothelium, triggering the onset of this vaso-occlusive disease. However, studies with a larger number of patients are necessary to obtain definitive results.

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