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Pigmentation disorders as a guide sign of autoimmune diseases

Trastornos de la pigmentación como signo guía de enfermedades autoinmunitarias

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Reynolds syndrome is an autoimmune disease characterised by the association of primary biliary cirrhosis (PBC) with limited systemic sclerosis. Multiple studies have shown that the cutaneous form of limited systemic sclerosis is more common in patients with PBC than the diffuse subtype (up to 93% of patients with Reynolds syndrome have the limited systemic sclerosis subtype).[1–3]

A 40-year-old woman diagnosed with PBC three years ago was referred to us with onset of generalised hypopigmented lesions. Physical examination revealed leukomelanoderma with morphology of reticulated hypomelanosis with follicular hyperpigmentation. There was also microstomia and peribuccal telangiectasias, and capillaroscopy was compatible with an active sclerodermiform pattern. Anti-centromere antibodies were requested in order to rule out limited systemic sclerosis, which were positive. Based on these findings, the patient was diagnosed with Reynolds syndrome.

The combination of CREST syndrome and PBC has been given its own name (Reynolds syndrome) and according to some authors is a distinct subgroup of patients with distinct clinical features and a better prognosis.

Pigmentation disorders, such as the salt-and-pepper pattern described in systemic sclerosis, can be of great help in the early diagnosis of this disease. This sign is described as a vitiligo-like dyschromia due to depigmentation with characteristically marked perifollicular accentuation. The capillary network of the hair follicles preserves melanogenesis and maintains pigmentation. This finding seems to correspond to perifollicular fibrosis, and is associated with the presence of systemic damage due to pulmonary involvement[4] (Figs. 1 and 2).

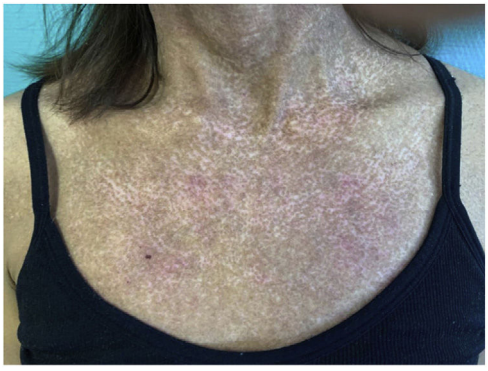

Figure 1. Widespread hypopigmented lesions. Dyschromia with salt and pepper pattern.

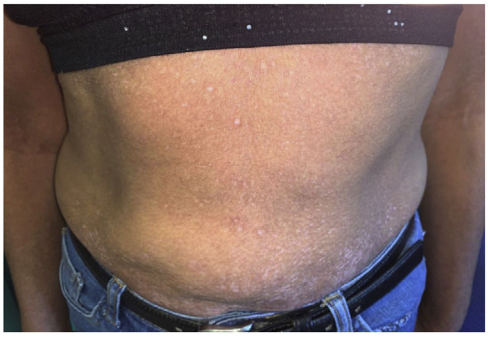

Figure 2. Physical examination revealed leukomelanoderma with morphology of reticulate hypomelanosis.

It is recommended that patients with a diagnosis of PBC should be routinely screened for symptoms related to systemic sclerosis. Skin pathology may act as a diagnostic guide sign in this disease.

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Conflict of interests

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