

AUTOIMMUNE CONNECTIVE TISSUE DISEASES

BULLOUS SCLERODERMA: AN ENTITY THAT CAN NOT BE FORGOTTEN

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Background: Bullous scleroderma is a rare type of localized scleroderma whose pathogenesis is still poorly understood. It is believed that lymphatic obstruction is caused by cutaneous fibrosis. It is most common in legs and lower abdomen. As a result of severe sclerosis the formation of ulcers is common and establishes a gateway for infections by increasing the morbidity of the disease. In this way, the early recognition it is fundamental for appropriate treatment and prevention of complications.

Observation: A 73-year-old woman, referred to the dermatology clinic for ulcers in lower limbs for 8 months. A patient reported hypocromic lesions with texture alterations (hardened) appeared in the whole body that were diagnosed as scleroderma in plaques by histopathology. She reports that shortly after, bubbles appeared on lesions. The lesions present themselves as bright, hardened, yellowish, erythematous plaques with vesicles and bubbles on the surface, as well as ulcerations. The patient was diabetic making us think of bullosis diabeticorum or necrobiosis lipoidica. In skin biopsy it was possible to observe the subepidermal bubble with important fibrosis and chronic inflammatory perivascular infiltrate favoring a bullous scleroderma hypothesis. Methotrexate was started with partial improvement of the lesions.

Key message: Bullous scleroderma is still a diagnostic and therapeutic challenge. The higher frequency in lower limbs leads to the erroneous diagnosis of stasis ulcers due to peripheral vascular insufficiency, especially when there is no evidence of scleroderma plaques in other places. Thus, this disease must always be remembered as a differential diagnosis of ulcers in the lower limbs preceded by blisters and with difficult response to the usual treatments.





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