Sclerodermatomyositis Overlap syndrome: a rare bullous presentation

M.Elomari Alaoui[1], A.Fliti[1], N.Er-rachdy[1] ,L.Benzekri[1], N.Ismaili[1], M.Meziane[1], K.Senouci[1]

Service de dermatologie, Université Mohamed V de Rabat, Hôpital universitaire Ibn Sina, Maroc

INTRODUCTION

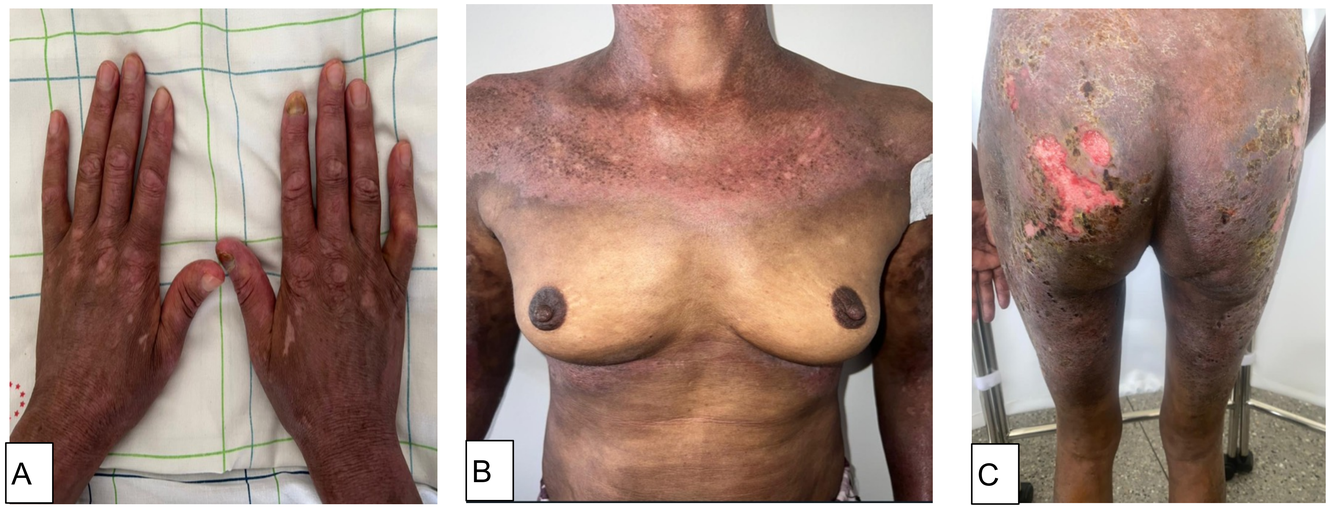
Sclerodermatomyositis overlap syndrome refers to a rare autoimmune condition characterized by features of both scleroderma and dermatomyositis.
Scleroderma involves thickening and hardening of the skin and connective tissues, while dermatomyositis affects the skin and muscles, causing muscle weakness and skin rash. Sclerodermatomyositis has rarely been reported in the literature, and cases of bullous presentation have not been described. We therefore present a case detailing this uncommon manifestation of sclerodermatomyositis.

Case Report

The patient aged 48, with a 4-year history of Raynaud phenomenon, was admitted to the dermatology department because of the appearance of pruritic bullous skin lesions. These lesions are characterized by phases of flare-up and remission.
Clinical examination revealed a lilac-colored erythema on the eyelids, extensive, symmetrical, erythematous and purplish macules on the décolleté, the posterior surface of the neck, the upper part of the shoulders and the roots of the MS , as well as erythematous +/- scaly macules arranged in bands along the extensor tendon sheaths (Gottron sign). Extensive poikilodermal plaques were also noted on the upper trunk, with two sclerotic plaques on the thighs. In addition, several post-bullous erosions were found on the back, upper and lower limbs.
Neuromuscular examination revealed global and segmental muscle weakness, as well as muscle deficits in the shoulder girdle, with difficulty in combing hair and reaching elevated objects, and in the pelvic girdle, with difficulty getting out of bed and a positive stool sign. And the rest of the clinical examination was normal.
Capillaroscopic examination showed mega capillaries with filiform hemorrhage as well as capillary rarefaction.
Laboratory examination revealed increased creatine kinase, aspartate aminotransferase, alanine aminotransferase,slactic dehydrogenase and aldolase, and immunological tests revealed positive anti-nuclear antibodies and PM-Scl antigen.
An electromyogram showed a scattered and multifocal myogenic abnormality of proximal muscularis with absence of motor and sensory nerve conduction abnormalities. A skin biopsy showed hyperkeratosis, vacuolization of the basal keratinocytes, perivascular lymphocytic infiltrate, dermal fibrosis and subepidermal vesiculation.
Finally, the diagnosis of sclerodermatomyositis in its bullous form was retained.
The patient was treated with oral corticosteroids and methotrexate;

DISCUSSION

Sclerodermatomyositis, a rare autoimmune disorder, presents a complex clinical picture due to the combination of symptoms of scleroderma and dermatomyositis. This is what we call overlap syndrome, It clinically manifests with scleroderma-like skin changes, myalgia/myositis, Raynaud's phenomenon . Skin lesions are often associated with itching or burning sensations, following exposure to sunlight or ultraviolet light. Other dermatomyositis-type skin lesions have been described but are rarely observed, such as bullae and post-bullous erosions, as described in our patient case. The bullous form of sclerodermatomyositis is often associated with a high incidence of malignant tumours.In our patient, however, the extension work-up revealed no associated tumours, but clinicians need to be aware of and vigilant for bullous forms of MDS. The association of the clinical signs of dermatomyositis with the signs of scleroderma suggests this diagnosis and the immunological and biological work-up should be completed with a skin and muscle biopsy to confirm the diagnosis.
In terms of biological diagnosis, an antibody directed against the PM-Scl antigen is often found. It is a principal biological marker of sclerodermatomyositis. The Positive ANA may be found in 50-80% of dermatomyositis cases, particularly in patients with overlap syndrome such as sclerodermatomyositis. Electromyography (EMG) may still be useful for confirming myogenic changes. MCTD presents with combined features of Systemic Lupus Erythematosus , systemic sclerosis, dermatomyositis/ polymyositis, and a positive anti-U1-RNP antibody.
Therapeutic management of MDS relies primarily on non-steroidal anti-inflammatory drugs or oral corticosteroids. However, in cases of severe disease, i.e. associated pulmonary or arthritic involvement, aggressive treatment (immunosuppressive therapy, IV immunoglobulins) and rigorous follow-up may be proposed.



A : Gottron sign B : (V-shaped erythema), and poikiloderma. C : several post bullous erosion