DERMATOPATHOLOGY

PANSCLEROTIC MORPHEA

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Background: Morphea or localized scleroderma, is characterized by sclerosis of the skin and subcutaneous tissues. The recent classification individualizes 5 subtypes: plate, linear, generalized, mixed and pansclerotic. It is distinguished from systemic scleroderma by the absence of visceral (lung) and vascular involevement (no Raynaud's syndrome). The disease is generally resistant to usual treatments (general corticosteroid therapy, methotrexate, mycophenolate mofetil and extracorporeal photochemotherapy). It can be the cause of a major functional and aesthetic damage. We report a new observation.

observation: A 76-year-old patient, with no particular pathological history, presented with 6 months history of induration and hyperpigmentation of the right hand, without raynaud phenomenon or other systemic signs. Clinical examination revealed a sclerotic hyperpigmented cup of the hand and the right forearm with amyotrophy and a pudgy aspect of the fingers. The diagnosis of deep morphea was confirmed by skin biopsy. The biological assessment was normal. The patient was put on corticotherapy bolus for 3 days and then oral relay (0.5 mg / kg / day) combined with methotrexate (15 mg / week) with functional rehabilitation sessions. The decline is 2 months with a clear improvement in sclerosis and stiffness.

Key message: This form of scleroderma is poorly known and sometimes confused with systemic scleroderma. It is individualized by its rapidly aggressive evolution and damage to the skin and deep structures causing contractures and trophic disorders complicated by superinfection and squamous cell carcinoma. It is important to report all cases of this rare and very serious form, including cases of therapeutic resistance and fatal outcome, to improve knowledge about the disease.