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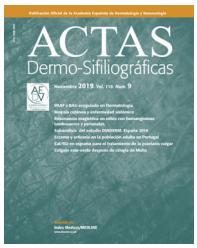

RESIDENT'S FORUM

RF - Generalized Morphea: Definition and Associations☆

FR - Morfea generalizada: definición y asociaciones

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Morphea, also known as localized scleroderma, encompasses a set of cutaneous sclerotic disorders of unknown etiology, with a wide range of manifestations and symptoms that range from mild local discomfort to severe complications.

There are many classifications of morphea, although one of the most widely used is that of Laxer and Zulian, which differentiates between 5 subtypes: circumscribed (the most common of the 5), linear, generalized, pan-sclerotic, and mixed.[1] Other classification systems include less frequent subtypes such as guttate and bullous morphea.

Generalized morphea is characterized by ≥ 4 plaques measuring at least 3 cm that coalesce and affect 2 or more anatomical regions. This subtype must be distinguished from systemic sclerosis, mainly through clinical characteristics. The absence of Raynaud phenomenon, sclerodactyly, facial involvement, abnormal nailfold capillaroscopy findings, visceral involvement, and specific autoantibodies all point toward a diagnosis of generalized morphea.[1,2]

Teske et al.[3] recently mapped lesions in patients with generalized morphea in order to attempt to define the different patterns of presentation. The authors reported 2 clinically relevant subtypes: the isomorphic subtype, with lesions at areas affected by friction; and the symmetric subtype, where involvement was similar on the trunk and extremities at both sides of the midline. The latter pattern was predominant in males and more frequently affected the deep planes of the dermis, the subcutaneous cellular tissue, and the fascia. The authors excluded unilateral generalized morphea from the classification. In this condition, the lesions affect only 1 side of the body and are considered linear morphea in most publications.[4]

The literature contains several cases of patients with generalized morphea and various types of malignancy, such as lung cancer and breast cancer,[5] although while systemic sclerosis has been reported to be a paraneoplastic phenomenon, the association between morphea and cancer is not so consolidated. Nevertheless, given the potential

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association, it seems advisable to take a targeted clinical history and appropriate work-up to rule out malignancy, especially in older patients with acute onset morphea and extensive involvement. Morphea has also been reported to be induced by various medications, with the most commonly reported in the literature being tumor necrosis factor α drugs[6] (although, paradoxically, infliximab has led to a good response in some cases of morphea[2]). Given the recent report of a case of morphea induced by nivolumab,[7] this possibility should be taken into account when there is a temporal relationship with the introduction of a new drug.

As for treatment of morphea, topical drugs are the approach of choice in localized and superficial forms. Cream formulations of corticosteroids, tacrolimus, calcipotriol, and even imiquimod 5% have been shown to improve symptoms in several series. In the case of generalized or deep forms, immunosuppressants and phototherapy are more useful. The most widely reported and successful immunosuppressant is methotrexate, with mycophenolate mofetil being a good alternative.[1] As new molecular pathways involved in the pathogenesis are discovered, new therapeutic targets appear. In this sense, favorable responses have been reported with abatacept, imatinib, tocilizumab, and apremilast (although data for apremilast are based on animal models).[8] In the future, other drugs aimed at specific interleukins could prove very useful.

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