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Morphea: insights into a rare skin condition

Morfia: percepções sobre uma condição de pele rara

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Keypoints

What is known

- Morphea is a rare connective tissue disease, characterized by skin inflammation and fibrosis.
- Genetic predisposition, immune dysregulation, and environmental factors are recognized triggers.
- Treatment can be challenging, especially in moderate to severe cases requiring systemic therapy.

What is added

- Morphea can present with characteristic painful lesions in pediatric patients.
- A multidisciplinary approach, involving dermatology, is essential for accurate diagnosis and management.
- Although diagnosis is clinical, skin biopsies are useful for confirming the diagnosis, especially in complex cases.

The authors describe the case of a previously healthy seven-year-old female with no suggestive family history, who was admitted to the Emergency Department (ED), with hyperpigmented, thickened, longitudinal band-like lesions on the right lateral arm and axillary regions, which had appeared six weeks prior to the ED visit.

The patient reported localized pain, although there was no history of significant medical or trauma.

Upon examination, the lesions measured approximately 15 centimeters on the long axis, had a central waxy area, surrounded by an erythematous margin, were of a brownish color, and appeared to increase in size (Figs. 1A and B). They did not, however, affect joint mobility.

There were no other manifestations, namely arthralgia, loss of mobility, Raynaud's phenomenon, or other cutaneous alterations.

She had been admitted two weeks previously for the same symptoms.

During her first visit to the ED, an ultrasound was performed, which revealed no significant findings and the patient was discharged with flucloxacillin.

Morphea was suspected and the collaboration of the Dermatology Department was then requested.

During the patient's time on the ED, a skin biopsy was performed, showing thickening of the dermis, with lymphocytic and plasma cell infiltrates at the edges, with an area of central sclerosis, confirming the diagnosis of active linear morphea (or juvenile localized scleroderma [JLS]).

There were no other extracutaneous symptoms.

The patient was transferred to Outpatients and was treated with prednisolone (1 mg/kg/day for two months with subsequent gradual tapering), along with topical treatment with calcipotriol, tacrolimus, and betamethasone, with satisfactory resolution of both lesions.

Morphea is an inflammatory fibrotic skin disorder.[1] Although its etiology remains poorly understood, it involves genetic predisposition, immune dysregulation, and environmental factors.[2]

It is more common in girls and typically appears around the age of five to seven.[1]

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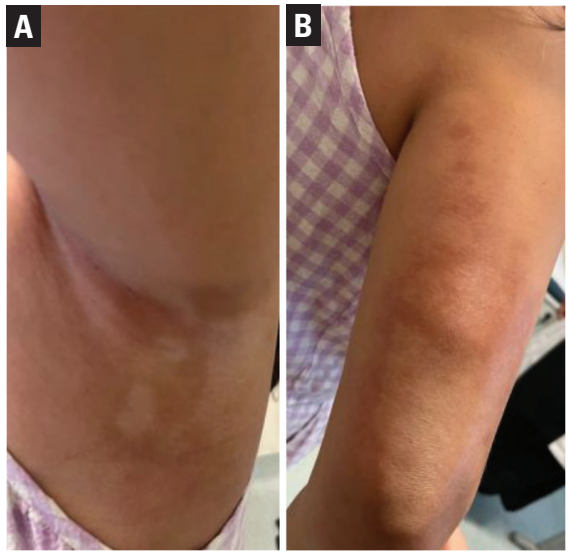

Figures 1. Lesions on **A:** the axillary and **B:** external right arm regions.

According to recent classification systems, morphea is divided into five different types: circumscribed (plaque), linear, generalized, pansclerotic, and mixed.[1]

A diagnosis can usually be made from a clinical history and physical examination alone. However, a skin biopsy may be helpful in establishing a definitive diagnosis, especially in complex or atypical cases.[3]

Certain diagnostic tools, such as the Localized Scleroderma Cutaneous Assessment Tool (LoSCAT) can be used to evaluate disease activity and severity.[4]

Diagnosis and treatment should be prompt so as to prevent the development of any sequelae of the disease.[5]

The differential diagnosis is broad, including conditions such as systemic sclerosis, lichen sclerosus, and keloids.[6]

Treatment aims to arrest disease activity and prevent disfigurement, joint contractures, and restriction to mobility.[7]

Types of therapy depend on the level of disease activity, subtype, and depth of the lesions.[6]

Mild variations of the disease can be managed by topical or intralesional steroids, tacrolimus, calcipotriol, or a combination of calcipotriol and a steroid, imiquimod, and/or phototherapy.[7]

As for the disease-modifying antirheumatic drugs that should be started in combination with corticosteroids, experts recommend methotrexate as first-line treatment.[4]

In moderate to severe morphea, recommendations include methotrexate in combination with systemic steroids as an initial "bridge therapy". In cases of resistance, the options are mycophenolate mofetil, cyclosporine, and biological agents.[7]

Most patients have a good prognosis and achieve remission.[7] However, relapses are common, reported in 15 to 53% of JLS patients, so patients need to be monitored over the long term.[8] Extracutaneous complications are also frequent.[8]

Children with suspected JLS should be referred to specialized pediatric rheumatology centers to ensure accurate assessment and effective management.[4]

Author contributions

C.M. Francisco: conceptualization, data curation, methodoloy, writing – original draft; J. Frade: data curation, methodoloy, validation, writing – review and editing; I. Correia: project administration, validations, writing – review and editing; S. Fonseca: supervision, validation, writing – review and editing.

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Conflicts of interest

None.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from all patients, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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