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Morphoea: A disfiguring disease

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Introduction: Deep morphoea is a subtype of localized scleroderma, a benign and often self-limiting condition of the skin in which inflammation and scarring occurs in the dermis, panniculus, fascia or superficial muscle. We illustrate a case of an adult with childhood-onset deep morphoea which had led to significant disfigurement.

Result: A 29-year-old woman was referred to the Rheumatology Clinic for the diagnosis of localized scleroderma. She initially presented at the age of three years old with a skin rash on her right leg and foot, which had progressively spread over her body with skin tightening and contractures leading to limb deformities. Tissue biopsy showed abnormal collagen deposition seen at the subcutis with homogenization, consistent with deep morphea. She was treated with systemic steroids in the past and underwent multiple corrective limb surgeries between ages of 4 and 25, including five osteotomies and Ilizarov surgery. She had an underdeveloped right breast with a patch of tightened skin over her abdomen for which she had undergone a recent abdominoplasty. There were no other lesions subsequently after puberty. Physical examination revealed generalized tightened and depressed skin plaques over the abdomen and her lower limbs. Other system examination and laboratory tests were unremarkable. She is currently not on any medication. She ambulates without any aid, and wears shoes with special insoles. She had just completed her studies.

Discussion/ Conclusion: Childhood-onset morphoea, also known as localized juvenile scleroderma, is more common in girls, typically around the age of 5 to 7 years. It primarily affects the skin with no visceral involvement. It is treated with phototherapy, topical corticosteroids, tacrolimus, or calcitriol. Systemic corticosteroids and immunomodulators such as methotrexate, mycophenolate mofetil and tacrolimus have been utilized. The disease tends to run a chronic or intermittent-recurrent course and frequently causes cosmetic sequelae in the majority and functional limitations.



Focal morphoea at abdomen



Linear morphoea at right breast



Multiple underdeveloped digits at lower limbs well healed linear morphoea at left thigh down to left foot

Biography

Janaardhan Tinakaran is a medical officer currently working in the internal medicine department in Hospital Putrajaya, Malaysia since October 2021. He is graduated from I.M. Sechenov First Moscow state Medical University in June 2014 (1st class honours). As a medical officer, he served in the Neurosurgical Department in Hospital Ipoh, Perak, Malaysia from April 2017 – September 2019. He was then transferred to the Oncology department in National Cancer Institute and served there from October 2019 – September 2021. Now, currently he is doing his MRCP examinations (PACES) to further pursue a career in dermatology with a keen interest in dermatosurgery and dermato-oncology to help increase the awareness of patients suffering from morphea.

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