Journal of Pakistan Association of Dermatologists. 2024;**34(2)**:582-586.

Case Report

Deep circumscribed morphea: A case report

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Abstract Idiopathic sclerotic skin illnesses are included under the term "morphea", sometimes known as localized scleroderma. Deep induration of the skin and subcutaneous tissue that extends to the underlying muscle and bone is a feature of the morphea subtype known as "deeply circumscribed morphea. Improving the long-term result requires early diagnosis, accurate evaluation, and efficient therapy. We report a case of a 3-year-old boy with a 2-year history of multiple erythematous to violaceous plaques with subcutaneous atrophy on the buccal, oral, mental, and neck area. He has difficulty swallowing large pieces of food. A skin biopsy confirmed the diagnosis and showed the usual morphology of morphea. The patient was administrated tapered systemic corticosteroid for 20 weeks and oral Methotrexate course for 24 weeks. There was clinical improvement of the lesions clinically with decreased hyperpigmentation of the lesions, regression of the induration and no difficulty of swallowing. The patient is still being monitored to assess progression of the lesions and disease activity. Skin biopsy results and typical clinical symptoms are often used to diagnose morphea. Early diagnosis and treatment are required to reduce harm, such as aesthetic sequelae and functional impairment, that may be caused by uninterrupted exercise. Treatment depends on the depth of lesion involvement and the extent of the disease, focusing primarily on limiting disease activity. Morphea may be self-limited, but frequently has a remitting, relapsing or chronic course, resulting in significant disease burden over time.

Key words
Deep circumscribed morphea; Juvenile localized scleroderma; Morphea.

Introduction

Morphea, sometimes called localized scleroderma, is a connective tissue condition marked by excessive collagen deposition that results in ossification of the dermis, subcutaneous tissue, or both.[1] The progression of morphea comprises an early inflammatory phase marked by skin erythema, fibrosis, sclerosis, and ultimately atrophy. Morphea is often considered a benign, self-limiting illness that only affects the skin and subcutaneous tissues. However, it often affects the underlying bone and muscle, and extracutaneous symptoms may occur.[2] Morphea often leads to permanent functional and cosmetic impairment.[3] According to estimates, 1 to 3 cases of morphea per 100,000 children occur annually in children.[4] The average age of disease onset in children is between 3 to 6 years,[5] but morphea is also acquired in infants and even neonates. The estimated female to male ratio of pediatric morphea is 2:1.[4]

Here we describe a relatively rare case of deep circumscribed morphea in a child. Although the etiology of this disease remains unknown, it can be considered as a disorder of the immune system. Management of this disease is challenging, as there is no definitive treatment for this disease. Treatment in these cases relies

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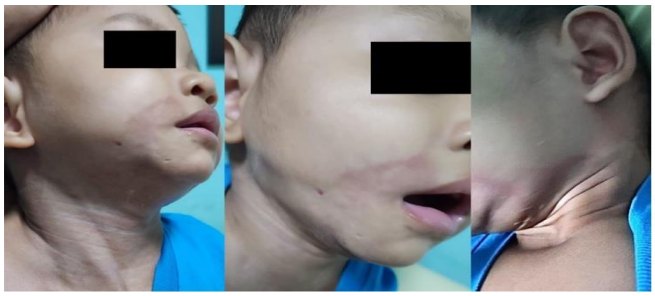

Figure 1 Erythematous to violaceous plaques, 3-8 cm in size with subcutaneous atrophy and induration on some of the plaques.

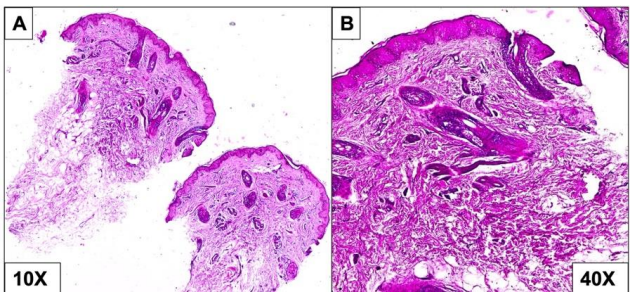

Figure 2 (A) The skin was covered with atrophic epidermis, shortened rete ridges. (B) The dermal layer was fibro-collagenous tissue accompanied by perivascular lymphocytic infiltrates, and the eccrine glands and hair follicles of the sebaceous glands appeared atrophic.

on immunosuppressive therapy such as methotrexate and corticosteroids.

Case report

A 3-year-old boy presented with complaints of purplish-red patches on both lower cheeks and neck for the past 2 years. The patient's mother explained that the patches appeared on both behind both of the patient's ears, disappeared on their own without treatment, and reappeared on the mouth, chin, both lower cheeks, and neck. The patches were slightly harder than the surrounding skin tissue. The patient's mother complained that the patches had been accompanied by complaints of difficulty swallowing food, especially large foods, and difficulty opening the mouth wide. The patient did not appear to scratch or complain of pain at the site. A history of previous trauma to the patch area was denied. No macules were found on other parts of the body. There were no complaints of tightness, difficulty speaking, or difficulty moving either hand or foot. The patient had previously seen a dermatologist and was treated with methisoprinol 250 mg syrup, prednicarbate 0.1% cream, and emollients, but there was no improvement. The patient had no significant medical history, and the patient's mother denied any recent history of illness in the patient.

Physical examination revealed erythematous to violaceous plaques, 3-8 cm in size, with subcutaneous atrophy and induration on some of the plaques **(Figure 1)**. A skin biopsy was obtained from the plaques in the right buccal region. Histopathology showed that the skin was covered with atrophic epidermis, shortened rete ridges. The dermal layer was fibro-collagenous tissue accompanied by perivascular lymphocytic infiltrates, and the eccrine glands and hair follicles of the sebaceous glands appeared atrophic, concluding a morphea **(Figure 2)**. Complete blood count including liver and kidney function, cortisol hormone, ANA serology, anti-DSDNA, and complement C3 and C4 were found to be within normal limits. The patient was also referred to the pediatrics, otolaryngology, and ophthalmology departments and no abnormalities were found in these areas.

Treatment was started with oral prednisone 15 mg/day (dose 1mg/kg/day) for 4 weeks and evaluation was performed. After 4 weeks, the patient's complaints of dysphagia began to decrease, but there was no significant change in the skin lesions, so additional oral therapy was started with methotrexate 10 mg/week and folic acid 3x1 mg/day. Prednisone was tapered and evaluated every 2 weeks. After treatment with tapering oral prednisone for 20 weeks and oral methotrexate 10 mg/day for 24 weeks, no new lesions were found, the color of the plaque faded to light brown in the oral et mental region with softened induration. No lesions were found in

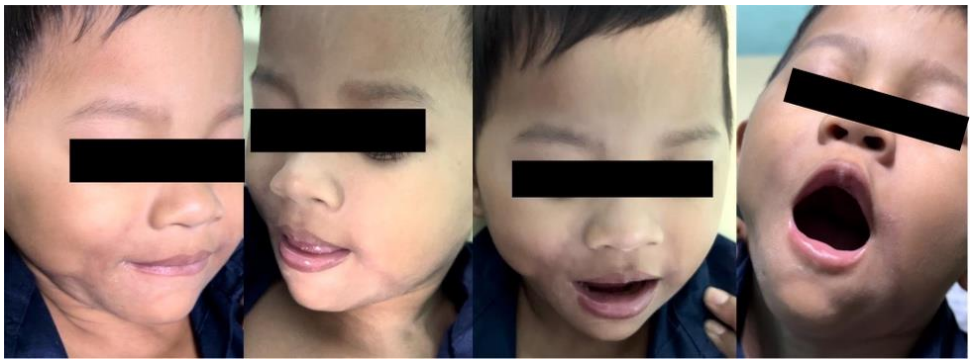

Figure 3 Hyperpigmented plaques with softened induration.

the neck region and swallowing difficulty was resolved **(Figure 3)**. Routine complete blood counts, including liver and kidney function, and cortisol hormone, were performed monthly and found to be within normal limits. The patient continued to undergo routine monitoring every 2 weeks. The patient was then scheduled to taper off methotrexate 2.5 mg/month after 6 months and continue therapy for up to 24 months.

Discussion

Morphea is an inflammatory skin disease caused by fibrosis.[6] The name "morphea", occasionally interchangeably with "localized scleroderma", avoids confusion with scleroderma, a condition with significantly higher morbidity and similar pathogenic hallmarks of immune activation and increased collagen formation. Instead of morphea, sclerodactyly or Raynaud's phenomenon may be regarded as a probable type of systemic scleroderma.[4]

Morphea is a group of disorders characterized by the presence of indurated lesions that vary in extent, distribution, clinical course, and complications.[7] Based on the Padua criteria, morphea is divided into five subgroups: circumscribed, linear (trunk or extremities and head), generalized, mixed, and sclerotic.[8] Circumscribed morphea is divided into two types: superficial and deep. In contrast to the deep version, which also affects the deep dermis, subcutaneous tissue, fascia, and muscle, the superficial variant only affects the epidermis and dermis.[5] The etiology remains unknown, although mechanical trauma is thought to be a precipitating factor in certain cases. Additionally, the etiopathogenesis may be influenced by genetic, immune, hormonal, viral, toxic, neurogenic, or vascular variables.[9] There has recently been speculation that Borrelia burgdorferi infection may be involved in at least some instances; however, this is still debatable.[4]

Morphea is characterized by superficial skin-colored, erythematous, or violaceous plaque-type lesions that progress over weeks to months to dense, hyperpigmented, or ivory-colored plaques, with or without surrounding violaceous inflammation. Signs of active disease include dense borders, warm to the touch, raised borders, and thickening of the dermis.[4] Clinical and histopathologic features are used to make the diagnosis of morphea.[9] Morphea and systemic sclerosis are difficult to distinguish histologically. Early inflammatory skin lesions may be identified from other skin lesions by thickening collagenous tissue in the reticular dermis parallel to the skin surface as well as by dense inflammatory infiltrates within the collagenous tissue and surrounding the blood vessels and sweat glands. Skin lesions become comparatively avascular and less often show signs of continuous inflammation during the late fibrotic phase. Late lesions are frequently extremely eosinophilic and feature thick collagen fibers. There are no sweat glands, or

they are atrophic. The subcutaneous tissue may transition from adipocytes to collagen.[10]

Regular laboratory measurements should include testing for lactate dehydrogenase, creatine kinase, erythrocyte sedimentation rate, and C-reactive protein in whole blood and serum. Although several autoantibodies, including antinuclear, antidNA single chain, anti histon, anti-topoisomerase II alpha, anti-ribonucleoprotein small nuclear, and anti-matrix metalloproteinase antibodies, have been discovered in morphea patients, regular testing for these antibodies is not advised.[10]

Management of this disease is challenging. Treatment usually aims to suppress inflammation and repair collagen. Several therapies include penicillin, antimalarial medications, retinoids, calcitriol, calcipotriol, imiquimod, cyclosporine, and ultraviolet (UV) A irradiation.[2] Methotrexate combined with corticosteroids is the first-line systemic treatment for morphea. In most studies with combination therapy, corticosteroids are used for induction therapy either orally or intravenously (intravenous methylprednisolone 30 mg/kg/day for 3 days every month or prednisolone 1 mg/kg/day) for the first 3 to 4 months. Methotrexate is used as a steroid-sparing agent and is started concurrently (0.6 mg/kg/week or 15mg/m^2) weekly in children given orally or by subcutaneous injection.[1,2,5,10] Methotrexate treatment should last at least 12 months, and dosage decrease should be considered if there are first indications of clinical improvement.[10] Almost 75% of children's diseases improve when receiving therapy for more than two years.[4]

In this case, a combination treatment with Methotrexate and corticosteroids was chosen, after the patient underwent treatment for 6 months, there was improvement in the lesions with no new lesions, thinner lesion color, and softened induration. It is important to understand what constitutes therapeutic success in morphea when treating patients. Successful treatment is determined by resolution of erythema, usually within 2 to 3 months, softening of the lesions, which can last for 12 months or more, end of lesion progression, and no new lesions.[11] Clinical follow-up (at least once a year) should be done in high-risk morphae who may experience recurrence after successful treatment.[10] In this patient, routine controls were still performed to assess lesion progression and recurrence.

Conclusion

In this case, combination therapy with methotrexate and corticosteroids was very beneficial for the patient and well tolerated. This disease relapses frequently with the chronic course of the disease getting more severe over time. As a result, regular check-ups are needed in patients to assess the progression of the disease so as to provide optimal long-term results.

References

1. Eleftheriou D, Shaw L. Morphoea (Localized Scleroderma). In: Hoeger P, Kinsler V, Yan A, Harper J, Oranje A, Bodemer C, *et al*., editors. Harper's Textbook of Pediatric Dermatology. 4th ed. New Jersey, USA: Wiley Blackwell; 2020. p. 1175–1128.
2. Weibel L, Sampaio MC, Visentin MT, Howell KJ, Woo P, Harper JI. Evaluation of methotrexate and corticosteroids for the treatment of localized scleroderma (morphoea) in children. *Br J Dermatol*. 2006;**155(5)**:1013–20.
3. Prasad S, Zhu JL, Schollaert-Fitch K, Torok KS, Jacobe HT. An Evaluation of the Performance of Current Morphea Subtype Classifications. *JAMA Dermatol*. 2021;**157(4)**:399–405.

4. Paller AS, Mancini AJ, editors. Collagen Vascular Disorder. In: Hurwitz Clinical Pediatric Dermatology. 6th ed. Missouri: Elsevier; 2021. p. 612–7.
5. Cyrus N, Jacobe H. Morphea and Lichen Sclerosus. In: Kang S, Amagai M, Bruckner A, Enk A, McMichael A, Orringer J, *et al*., editors. Fitzpatrick's Dermatology. 9th ed. New York: McGraw Hill; 2019. p. 1107–16.
6. Aranegui B, Jiménez-Reyes J. Morphea in Childhood: An Update. *Actas Dermosifiliogr*. 2018;**109(4)**:312–22.
7. Quispe SKJF, Cavaliere A, Weber M, Stramare R, Zuliani M, Quaia E, *et al*. Sarcopenia in juvenile localized scleroderma: new insights on deep involvement. *Eur Radiol*. 2020;**30(7)**:4091–7.
8. Asano Y, Fujimoto M, Ishikawa O, Sato S, Jinnin M, Takehara K, *et al*. Diagnostic criteria, severity classification and guidelines of localized scleroderma. *J Dermatol*. 2018;**45(7)**:755–80.
9. Melani L, Cardinali C, Giomi B, Schincaglia E, Caproni M, Fabbri P. Case study: Periodic Follow-Up Is Necessary in Morphea Profunda to Identify Systemic Evolution. *Skinmed*. 2005;**4(3)**:188–90.
10. Knobler R, Moinzadeh P, Hunzelmann N, Kreuter A, Cozzio A, Mouthon L, *et al*. European Dermatology Forum S1-guideline on the diagnosis and treatment of sclerosing diseases of the skin, Part 1: localized scleroderma, systemic sclerosis and overlap syndromes. *J Eur Acad Dermatol Venereol*. 2017;**31(9)**:1401–24.
11. Browning JC. Pediatric Morphea. *Dermatol Clin*. 2013;**31(2)**:229–37.