Panscleroticmorphea: A male child with hemiatrophy of lower limb

Malay K. Dasgupta, Chaitali Patra, Shatanik Sarkar, Sabyasachi Das

Department of Pediatric Medicine, R. G. Kar Medical College and Hospital, Kolkata, India

ABSTRACT

Morphea is a variant of localized scleroderma in which lesions are usually limited to the skin and subcutaneous tissue. Panscleroticmorphea is a rare atrophying and sclerosing type of morphea. It can follow a comparatively benign course with spontaneous resolution of symptoms, or sometimes can lead to a variety of complications resulting in progressive disability. We report a case of Panscleroticmorphea in an 8-year-old male child involving one lower extremity with extension to the lower trunk. It was associated with deformity and hemiatrophy of that limb, leading to restriction of normal day-to-day activity. The case is being reported in view of its rare occurrence in conjunction with other rarer features.

Key words: Panscleroticmorphea, contracture, hemiatrophy

Access this article online

Website: www.idoj.in

DOI: 10.4103/2229-5178.131092

Quick Response Code:

Address for correspondence:
Dr. Shatanik Sarkar,
Flat No. 304,
Block-B, DB-24,
Deshbandhunagar,
Baguiati - 700 059,
Kolkata, India.
E-mail: baselinegame@gmail.com

INTRODUCTION

Morphea is a variant of localized scleroderma in which lesions are usually limited to the skin and subcutaneous tissue.[1] According to the clinical presentation and depth of tissue involvement, morphea is of 5 types - guttate, plaque, linear, bullous and deep type.[2] Panscleroticmorphea is a rare variety of deep subcutaneous type, involving not only the skin and subcutaneous tissue, but extending upto the muscle and bone. Here, we report a case of Panscleroticmorphea presenting with unilateral atrophy of right lower limb and lower right part of the trunk associated with contracture of the ankle joint.

CASE REPORT

An 8-year-old male child presented with gradually progressive atrophy of right lower limb along with development of hyperpigmented lesions over right lower limb extending to right lower part of the trunk for last 3 years. There was a history of limping gait since a year and there was no history of fever, trauma or any exposure to drugs, chemicals or radiation. His family history was not contributory.

On examination, there were multiple thick, indurated, hyperpigmented, hypoaesthetic, anhidrotic and depilated lesions over right lower extremity extending to the right lower quadrant of trunk. There was muscle wasting and true shortening of right lower limb (measuring about 5 cm less than the left one) with fixed flexion deformity of the right ankle joint [Figure 1]. Skin was fixed to the underlying bone and there was presence of limping gait. There was no perioral or periorbital skin fixation, esophageal dysmotility, arthralgia, myalgia, Raynaud's phenomenon, respiratory or gastrointestinal abnormality. Motor power was grade IV and deep tendon reflexes were absent on the right lower limb.

On blood examination, there was eosinophilia (Absolute Eosinophil Count-1220/cmm), positive antinuclear antibody (ANA) and positive anti scl-70 antibody. Rheumatoid factor was negative. Radiological examination of affected limb showed soft tissue atrophy and decreased diameter of the long bones compared to the normal side. Skin biopsy from a hyper pigmented area over the leg showed atrophy of epidermis with loss of appendageal structures, collagenization of dermis and diminished subcutaneous tissue without any eosinophilic infiltrate [Figure 2]. According to the morphology and histopathology, he was diagnosed as a case of unilateral Panscleroticmorphea. He was treated with steroid, diphenylhydantoin and vitamin D3 and advised for physiotherapy, but no improvement was noted even after 3 months.

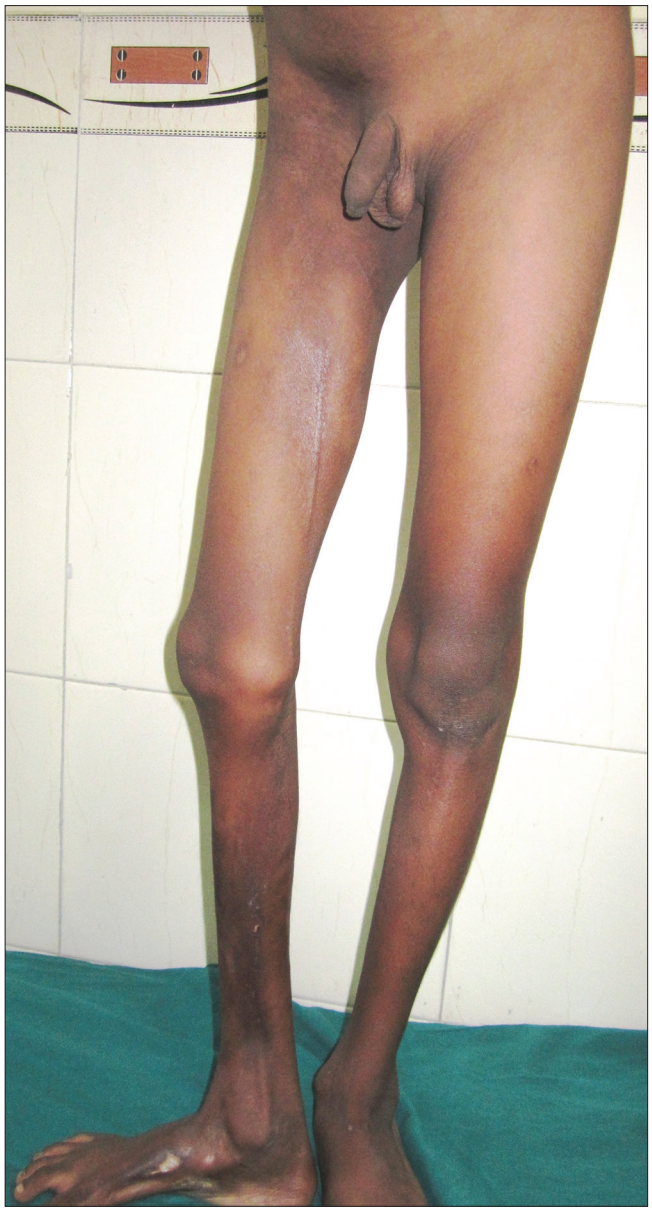

Figure 1: An 8-year-old male boy with hemiatrophy of right lower limb, fixed flexion deformity of right ankle joint and characteristic skin lesions

DISCUSSION

Morphea, also known as localized scleroderma, is a disorder characterized by excessive collagen deposition leading to thickening of the dermis, subcutaneous tissue, or both. The exact cause is unknown; however, trauma, excessive physical exertion, immobilization, radiation, infection by *Borrelia burgdorferi*, febrile illness, and vaccinations have been proposed as trigger factors. The incidence of morphea has been estimated as approximately 0.4-2.7/100,000 people.[3] Morphea is classified into guttate, plaque, linear, bullous and deep morphea according to the clinical presentation and depth of tissue involvement.[2] Pansclerotic morphea is a variety of deep subcutaneous subtype. Plaque and linear types are the commoner varieties seen in children, whereas, the pansclerotic type is still very rare.

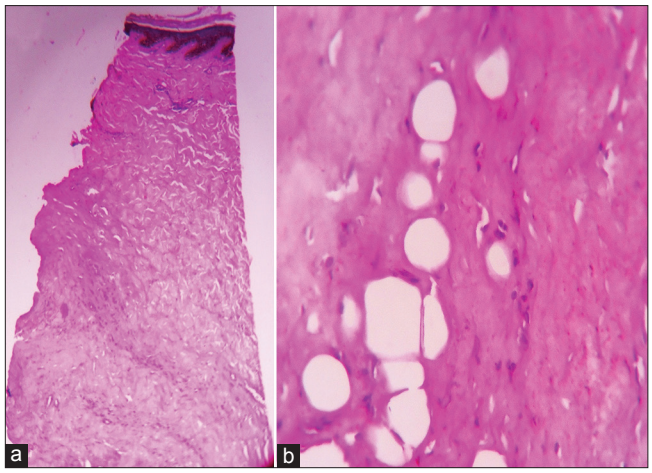

Figure 2: Skin biopsy showing (a) atrophy of epidermis with loss of appendageal structures, collagenization of dermis, (b) diminished subcutaneous tissue

Pansclerotic morphea is a rare atrophying and sclerosing variant that affects the subcutaneous tissue, muscles, tendons, and even bones. It may develop from linear morphea and can have a relentless, mutilating course leading to complications that include contractures, deformities, ulcers, and even squamous cell carcinoma.[4,5] It usually starts before the age of 14 years and a marked female preponderance is observed at a 3:1 ratio,[6] though our patient was a male with onset of the lesion as early as at 5 years of age.

Ipsilateral atrophy of the limb with contracture, along with lesion extending to the lower trunk was observed in our patient. However, he did not have arthritis, cramps or convulsions as described by the earlier authors.[7,8] When compared to adults, children are less likely to show serological abnormalities with ANA positivity encountered in about one third of those with localized disease.[9] It is very interesting in our case that ANA and Anti scl-70 both were found to be positive, making the case unique and interesting.

The contracture at the ankle and wasting of muscles reinforce the diagnosis of pansclerotic morphea. The extensiveness of his disease is complimented by positive ANA and anti scl-70 which are indicative of the ongoing disease activity.[10,11]

Diagnosis is essentially made by clinical examination aided by skin biopsy. The disease is usually slowly progressive and spontaneous remission can occur. There is no effective treatment available. Different therapeutic modalities tried with varying results are oral steroid, Ultraviolet light, calcitriol, D-penicillamine, low dose methotrexate, tropical calcipotriene, diphenylhydantoin and antimalarials. However, none of these has shown any significant promise till date.

Thus, this case of pansclerotic morphea with hemiatrophy and deformity of a limb in a male child presenting at a reasonably earlier age with ANA and Anti scl-70 positivity is an extremely rare entity, considering all these features occurring in a single case.

REFERENCES

1. Rowell NR, Goodfield MJ. The connective tissue diseases. In: Champion RH, Burton JL, Burns DA, Breathnach SM, editors. Rook Wilkinson Ebling Textbook of Dermatology. 6th ed., Vol. 3. UK: Blackwell Sciences; 1998. p. 2437-575.
2. Diaz-Perez JL, Connolly SM, Winkelmann RK. Disabling pansclerotic morphea of children. Arch Dermatol 1980;116:169-73.
3. Fett N, Werth VP. Update on morphea: Part I. Epidemiology, clinical presentation, and pathogenesis. J Am Acad Dermatol 2011;64:217-28.
4. Peterson LS, Nelson AM, Su WP. Classification of morphea (localized scleroderma) Mayo Clin Proc 1995;70:1068-76.
5. Padmavathy L, Rao LL. Unilateral linear pansclerotic morphea affecting face and limbs. Indian J Dermatol Venereol Leprol 2005;71:192-4.
6. Lewkonia RM, Lowry RB. Progressive hemifacial atrophy (Parry-Romberg syndrome) report with review of genetics and nosology. Am J Med Genet 1983;14:385-90.
7. Lapresle J, Desi M. Scleroderma with progressive facial hemiatrophy and atrophy of the other side of the body. Rev Neurol (Paris) 1982;138:815-25.
8. Thomas J, Janaki VR. Disorders of connective tissue. In: Valia RG, editor. IADVL Text book and Atlas of Dermatology. 2nd ed. Mumbai: Bhalani Publishing House; 2001. p. 911.
9. Rosenberg AM, Uziel Y, Krafchik BR, Hauta SA, Prokopchuk PA, Silverman ED, *et al*. Antinuclear antibodies in children with localized scleroderma. J Rheumatol 1995;22:2337-43.
10. El-Azhary RA, Aponte CC, Nelson AM. Do antihistone autoantibodies reflect disease activity in linear scleroderma? Arch Dermatol 2004;140:759-60.
11. Falanga V, Medsger TA Jr, Reichlin M. Antinuclear and anti-single-stranded DNA antibodies in morphea and generalized morphea. Arch Dermatol 1987;123:350-3.

Cite this article as: Dasgupta MK, Patra C, Sarkar S, Das S. Pansclerotic morphea: A male child with hemiatrophy of lower limb. Indian Dermatol Online J 2014;5:170-2.

Source of Support: Nil, **Conflict of Interest:** None declared.