CASE REPORT
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GENERALIZED MORPHEA IN A CHILD – A RARE ENTITY

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Abstract

Generalized morphea is a rare presentation, if present in children. We present a 5-year old female child who developed tautness of skin since 3 months.

Key words: generalized morphea, children, rare

Introduction

The term "Morphea" encompasses a group of related conditions characterized by varying degrees of sclerosis, fibrosis and atrophy in skin and subcutaneous tissue, sometimes extending deeply into muscle, bone and brain.[1] Anti-nuclear antibody(ANA) positivity is common but the specific autoantibodies seen in systemic sclerosis are rarely present.[1] Various type of morphea are – Limited morphea, Generalized morphea , Linear morphea & Mixed type. Generalized morphea is a rare condition, it occurs in the cases of dissesminated sclerosis with no systemic involvement. It occurs mainly in adults.It begins in the trunk and causes contractures and deformities in limbs. It differentiates from systemic sclerosis because it does not accompany Raynaud's phenomenon and doesn't show capillaroscopic changes.[2] The disease is extremely uncommon in children.[3]

Case Report

A 5-year female child presented with gradual onset and progressive tautness and tightness of skin since 3 months, which was accompanied by limitation of movement of hands,elbow,ankle and knee joints. Patient also complained of pain over fingers and toes without any change in colour. There was no history of regurgitation of food and difficulty in breathing. There was no history of trauma,bites or history of inflammation over those sites On examination, multiple large shiny atrophic plaques of size ranging from 3 cm to 20 cm were present over bilateral upper limb and lower limb involving the joints [Figure 1 and 2].

Contracture of small joints of hand was present, limiting the movement of hand [Figure 3].

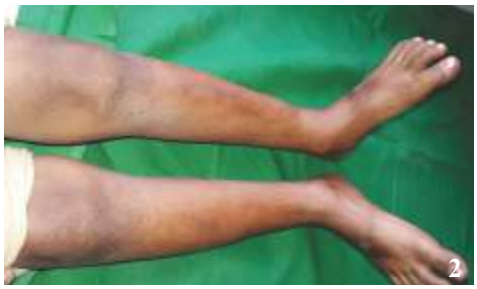
Matt telangiectesia was seen over lips with telangiectesia over bilateral cheeks(without any history of any topical application)[Figure 4 and 5].

Other dermatological examination was unremarkable. Based on history and clinical examination, a differential diagnosis of generalized morphea and CREST syndrome was considered. Complete blood count, ESR thyroid profile, Anti-nuclear antibody(ANA), Extractable nuclear antibody(ENA) profile and upper GI endoscopy were all within normal limit.Histopathology revealed dermal stroma with packed collagen bundles, eccrine glands with replacement of periadnexal fat by collagen and moderate lymphocytes, plasma cells and histiocytes around vessels [Figure 6 and 7].

Based on clinicohistological correlation,a diagnosis of generalized morphea was made. Patient was started with oral prednisolone (10 mg),topical calcipotriol and physiotherapy with an improvement of around 40%.

Discussion

When scleroderma of any type affects children, it is called Juvenile Scleroderma.[4] They constitute about 10% of patients suffering from scleroderma.[5] Generalized morphea is rare in infants. The diagnostic criteria of generalized morphea consists of – 1. 4 or more plaques become larger than 3 cm and merge, 2.Involvement of 2 or more anatomical areas [Head and neck, left or right upper limb or lower limb, anterior and posterior



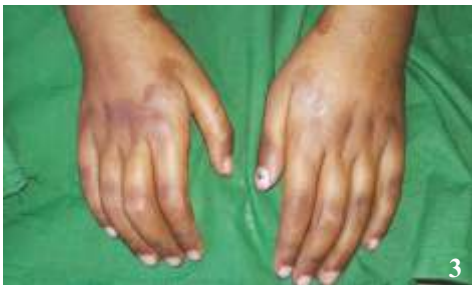


Figure 1,2, 3 : **1**. Large plaques of morphea over right upper limb. **2**. Large plaques of morphea over both legs. **3**. Contracture of small joints of hands

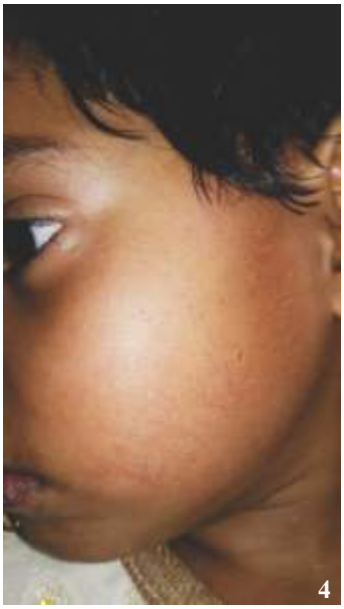
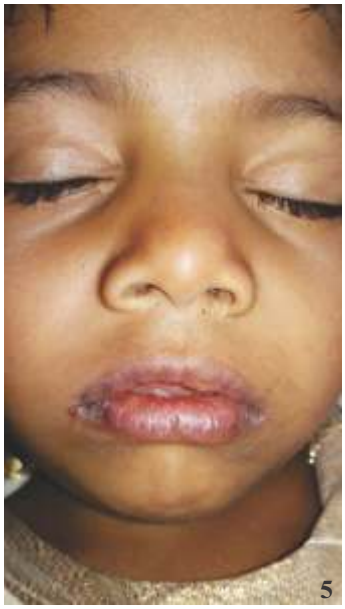



Figure 4, 5 : **4**. Telanigectesia over face. **5**. Matt telangiectesia over lips

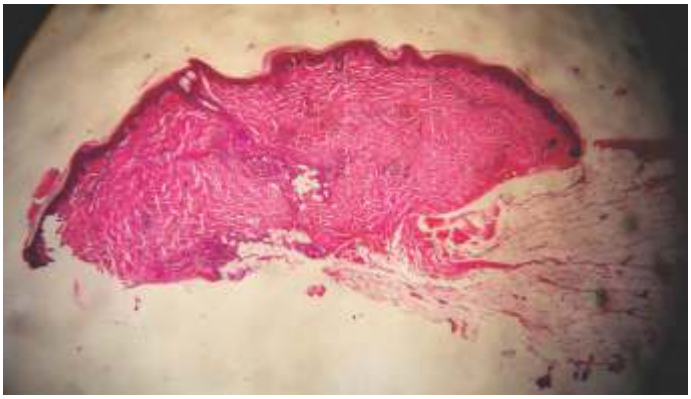


Figure 6 : Epidermal atrophy and dermal stroma with thick collagen bundles. (H & E Scanner View)

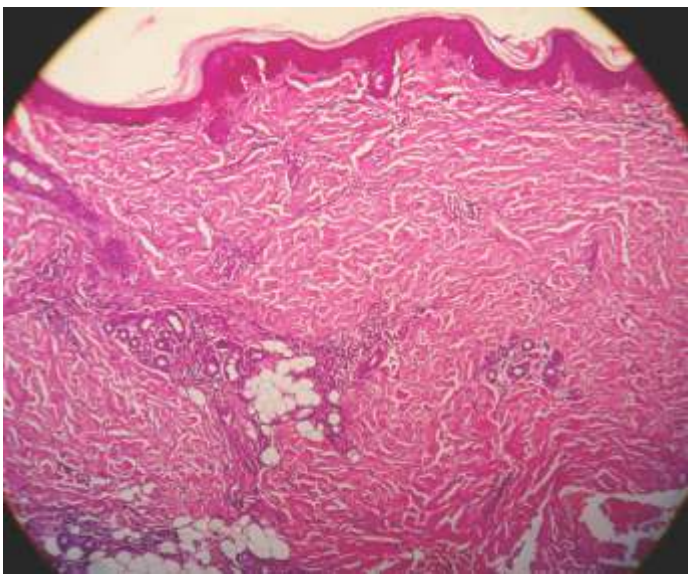


Figure 7 : Dermal stroma with thick collagen bundles, atrophy of pilosebaceous units with hypertrophied piloarrector muscle, presence of eccrine sweat glands in the dermis with replacement of periadnexal fat by collagen and moderate lymphocytes, plasma cells and histiocytes around blood vessels. (H & E 10×)

trunk].[6] The diagnosis of morphea is primarily clinical.

We excluded the differential of CREST syndrome by means of history and negative Anti-nuclear antibody and Extractable nuclear antibody profile

In a large multicentre, international study of Juvenile Localized Scleroderma of 750 children, generalized morphea was present in 7% of patients. It is rare during childhood.[7]

In 1976,Ansell et al[8] described 2 cases (10-year girl and 8-year boy) of generalized morphea with fixed flexion deformities of joints at the site of morphea lesions, similar to our case.

In 2003,Brar BK et al[3] reported a 9-year girl with generalized morphea, following measles vaccination.

In 2015,PK Dey et al[9] described a 10-month male infant with generalized morphea and reported its response to topical steroids.

The above cases lacked presence of telangiectesia over lips and face which was present in our case and drifted us to rule out CREST syndrome.

Only a handful cases have been reported from the Indian subcontinent, hence this case is being reported.

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