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Original Research Paper

Dermatology

A RARE OCCURRENCE OF BOTH PLAQUE AND EN COUP DE SABRE MORPHEA

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ABSTRACT En coup de saber and plaque morphea is a rare atrophying and sclerosing disorder of the subcutaneous fat, muscle and bone. In some patients the atrophic lesion extend to involve the ipsilateral face with radiological evidence of hemiatrophy. We report a case of a 4 year old child with deformity on right side of face that started as a indurated plaque on forehead and cheek. The case is being reported in view of its rare occurance.

KEYWORDS :

INTRODUCTION

Morphea (also known as localized scleroderma) is a disorder characterised by varying degrees of sclerosis, fibrosis, atrophy in the skin, subcutaneous tissue and sometimes extending deep into muscles, bone and brain[1]. It occurs at least 10 times more commonly in children than pediatric systemic sclerosis.[1] The condition has an estimated incidence of 0.4 to 1 per 100,000 in children.[1] We here report a case of morphea on the face of a 4-year-old child with both linear and plaque morphology.

CASE REPORT

A 4-year-old male child, born of non-consanguineous marriage came with complaints of binding down of skin over right side of face for 3 months and hemiatrophy of right side of face for 3 months. It was gradual in onset and progressive in nature. No history of preceding trauma/ vaccination/ infection/ drug intake/ radiation exposure/seizure/headache/any neurological complaint/ocular complaint/hearing difficulty/difficulty in opening mouth or dental complaint/ any other systemic complaints. On examination, the child was underweight and severely waisted for his age. On cutaneous examination, there were two well defined hyperpigmented atrophic plaques, one linear shaped on the right forehead and another irregular shaped on the right side of face with intervening skin in between the plaques non-indurated. (Fig 1) The linear plaque on forehead (3×8cm) was indurated and extended till frontal scalp and had hypopigmentation at places along with atrophy in the centre. (Fig 2) A well-defined shiny atrophic plaque of size 10x15cm on the right side of face extended from right temple above to upper1/3rd of neck below, medially till lateral to ala nasi and laterally till preauricular area. It was indurated, non tender with hypopigmentation present on the margin of the plaque. Appendages were lost over the plaques. (Fig 3) Partial madarosis was present on middle of right eyebrow and complete madarosis of lower right eye lid and medial aspect of right upper eyelid. (Fig 4) Hemiatrophy of right side of face along with deviation of angle of mouth. (Fig 5) Oral mucosal examination showed atrophy of right side of tongue and decrease mouth opening as well as deviation of tongue to right side. (Fig 6) Rest cutaneous and systemic examinations were normal. On investigating the child, all haematological, biochemical and radiological investigations were normal except orthopentogram showed retracted mandible on right side of face. ANA was negative. Histopathological examination of skin biopsy taken from the forehead plaque showed homogenization of collagen in the dermis and apparent pooling of appendages. (Fig 7) Mild lymphocytic infiltrate present in the dermis. On Von Geison stain, there was staining of collagen. (Figure 8). With these finding we kept a diagnosis of extensive linear morphea of head and neck en coup de sabre and plaque type). The child was started on IV methyl prednisolone pulse monthly and tablet methotrexate 5 mg daily. Currently there is halting of the progression of the lesions and child has received 3 pulses.

DISCUSSION

Scleroderma is a word used to describe a spectrum of conditions characterized by hardening and/or thickening of the skin and fibrosis of the tissues involved; it is, didactically, divided into systemic and localized forms. [2,3] The localized form, also known as morphea, is characterized by predominant skin involvement, with occasional involvement of subjacent muscles, usually sparing internal organs [2,3] Based on clinical criteria, morphea can be classified into several subtypes. These include plaque type morphea (including guttate, bullous and keloidal); generalized morphea; morphea profunda; linear morphea (also called linear scleroderma and includes morphea en coup de sabre [ECDS] and progressive hemifacial atrophy [PHA]); pansclerotic disabling morphea; and mixed forms.[1]

The en coup de saber type may evolve with bone atrophy, mandibular deformity, abnormal positioning of the teeth and aesthetic damage due to hemiatrophy or cranial and CNS deformity. [4] Plaque type morphea most commonly involves the trunk, whereas linear morphea primarily affects the face and extremities[1] The most common form of morphea is linear (51.4%) morphea in children with almost twice as many patients showing linear lesions on the extremities than the ECDS or PHA forms of linear morphea followed by plaque morphea (26%), generalised morphea (7%) and deep morphea (2%).[5]

The course of morphea is usually 3 to 5 years, but many patients have demonstrated periods of quiescence, only to be followed by reactivation of the inflammation, induration, and/or tissue loss that characterize the disorder. Diagnosis is based on the clinical features; biopsy specimen is confirmatory, but often not needed, especially in pediatric cases. Morphea therapy is mostly guided by clinical findings. Laboratory tests do not reliably predict the disease course. Patients need to balance the risks of systemic therapy with the risk of untreated disease. They must be counseled that treatment is aimed at active disease in the hopes of preventing enlargement of already-present lesions and the development of new lesions. Patients with linear morphea of the head and neck or limbs are at significant risk of facial deformity, limb length discrepancy and contractures and should, therefore, be treated with systemic therapy. Based on the available evidence, methotrexate in combination with a short course of systemic steroids is first-line therapy.[6] In our case we have

started the patient on systemic pulse steroid as well as daily methotrexate. The progression of the lesion has stopped according to child's parents.

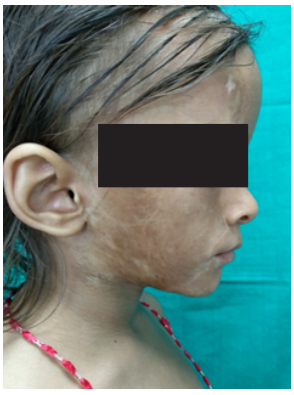

Figure 1: Two atrophic plaques on right side of face with normal intervening skin

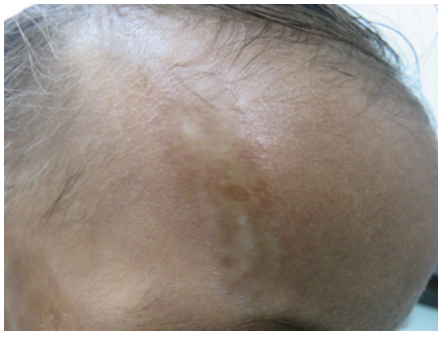

Figure 2: A linear plaque of 3×8 cm on right forehead

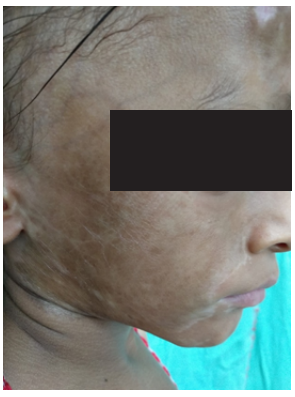

Figure3: A irregular shaped atrophic plaque of 10×15 cm on right side of face

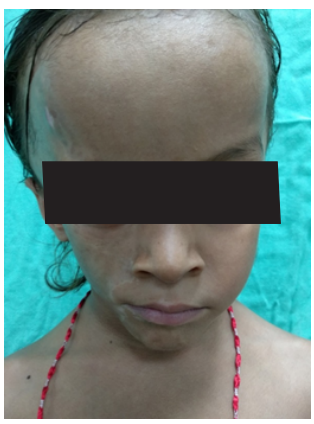

Figure 4: Partial madarosis of right upper eye brow and complete madarosis of right lower eyelid

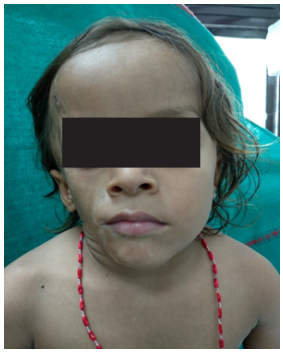

Figure 5: Hemiatrophy of right side of face along with deviation of angle of mouth

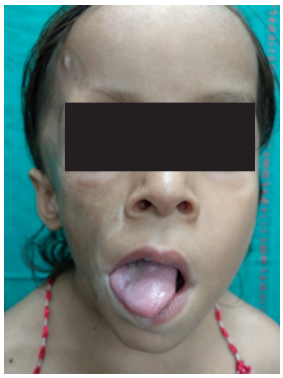

Figure 6: Deviation of tongue to right side

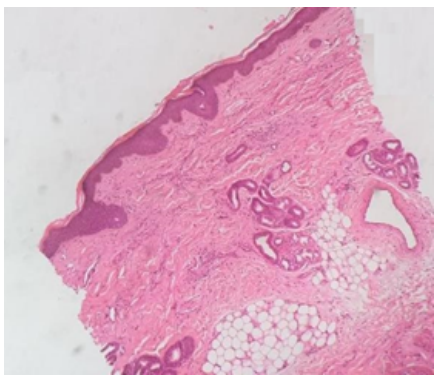

Figure 7: Histopathological examination revealing homogenization of collagen in the dermis and apparent pooling of the appendages

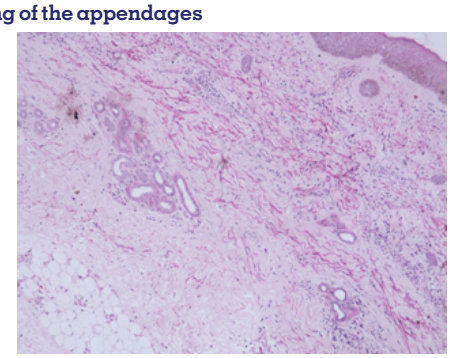

Figure 8: Von Gieson staining of collagen fibres

CONCLUSION

In our case the child had extensive linear morphea of head and neck (en coup de sabre and plaque type morphology) with deeper involvement with atrophy of rt side of tongue, retraction of right-side mandible and apparent deviation of angle of mouth to right side. Such a rapid progression in 3 months with both en coup de sabre type and plaque type on

the same side of face are rarely reported. So, all these children should be started on therapy early in order to halt the progression of the disease and prevent further sequele.

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