Linear Atrophic Lesion on Forehead

A 17 year old girl presented with asymptomatic linear depressed groove involving the midline of the forehead region which was progressively increasing over the past 1 year. The lesion was slowly extending to the scalp which became a major concern to the patient. There was no history of trauma preceding the development of the lesion. On examination, an ivory sclerotic depressed linear lesion mainly involving the mid forehead with slight extension to the scalp was noted (**Fig**. **1**). No other remarkable cutaneous or systemic finding was noted. We considered a differential diagnosis of "en coup de sabre" morphea, and linear atrophoderma. On histopathology, squared-off edge of the biopsy specimen with mild superficial and deep inflammatory infiltrate with collagen fibres, so a diagnosis of "en coup de sabre" morphea was made.

The name en coup de sabre is a french phrase and came from its resemblance to sabre cut. It is a localised type of linear scleroderma. The lesion usually starts with contraction and firmness of the skin over the affected area. Subsequently, an ivory irregular sclerotic plaque develops, sometimes with telangiectatic vessels coursing over it, together with hyperpigmentation at the edge. It may involve the scalp, producing a linear zone of alopecia, which may be preceded by bleaching of the hair. The groove may extend downwards into the cheek, nose and upper lip, and involve the mouth and gum. Subcutaneous tissue, muscle, and, occasionally, bone are involved; this ipsilateral form is known as progressive facial hemiatrophy or Parry-Romberg syndrome. Severely affected patients may have neurological manifestations due to involvement of the meninges resulting in seizures, so it is very important to diagnose and treat this condition at an early stage to avoid complications. Most cases of linear scleroderma are self-limited, with clinical activity apparent for an average of 3 to 5 years but en coup de sabre may have an insidious course lasting for decades. Treatment mainly is directed towards the inflammatory component and consists of corticosteroids, vitamin D analogues, methotrexate, cyclophosphamide, azathioprine, hydroxychloroquine, intralesional interferon-α, D-penicillamine and psoralen with ultraviolet A therapy (PUVA).

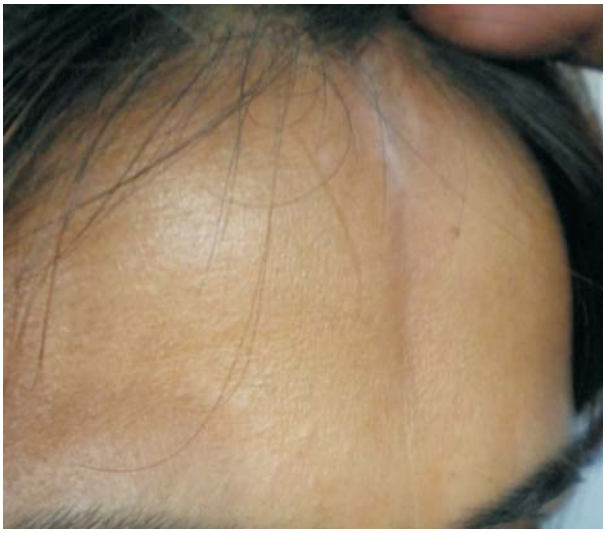

FIG. 1 *Linear atrophic lesion on forehead.*

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Epidermal Nevus Syndrome

A 6-year-old girl presented with multiple, brownish-black, well-demarcated papillomatous plaques over face, neck, trunk, and arm, arranged in a linear configuration along Blaschko's lines alongwith asymptomatic nodule of size 2cm x 1cm in her left eye, over limbus, since 6 months of age (***Fig.* 1**). Radiological investigations showed no abnormality. Histopathology of skin lesion was consistent with the diagnosis of verrucous epidermal nevus and excised tissue from limbus showed features consistent with dermoid.