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).

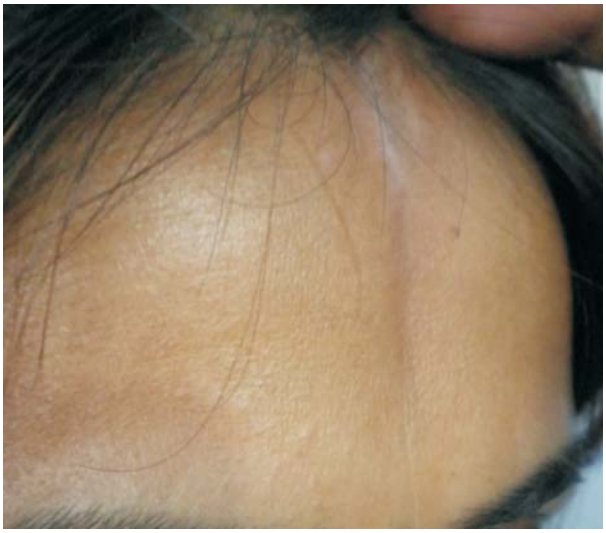

FIG. 1 *Linear atrophic lesion on forehead.*

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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.

Epidermal nevus syndrome is a disease complex consisting of the association of an epidermal nevus with developmental abnormalities of the skin, eyes, nervous, skeletal, cardiovascular and urogenital systems. Around 9 to 30% patients have ocular abnormalities, the commonest of which are colobomas and choriostomas. Choriostomas include dermoid, lipodermoid, single tissue and complex choriostoma.

Linear verrucous epidermal nevus should be differentiated from other dermatoses presenting as linear hyperkeratotic or verrucous lesions: lichen planus, psoriasis, lichen straitus, and porokeratosis. Linear lichen planus is severely itchy and violaceous in color with slight scaling, psoriasis presents with thick silvery white scales. Lichen straitus is an asymptomatic and self-limiting disease, which usually resolves in 1 year. Linear porokeratosis can be differentiated by its pathognomonic cornoid lamellae.

A verrucous epidermal nevus may enlarge slowly during childhood. By adolescence, the lesion usually reaches a stable size and further extension is unlikely. Excision is the most reliable treatment, but not advisable if very extensive or at sites not amenable to simple surgery. Other treatment modalities are electrofulguration, laser, cryotherapy, dermabrasion, and chemical peels.

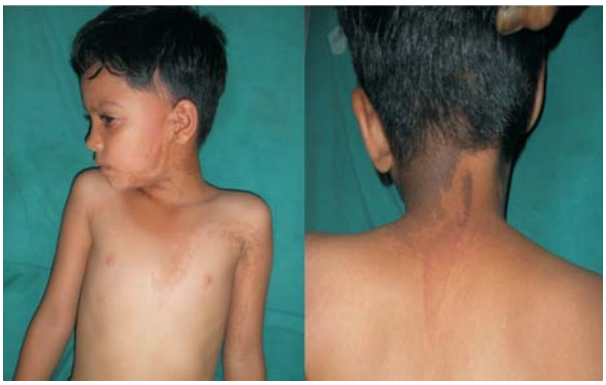

FIG. 1 *Plaques over face, arm, and neck.*

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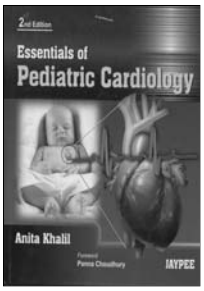
BOOK REVIEW



Essentials of Pediatric Cardiology: 2nd Edition
ANITA KHALIL
Jaypee brothers Medical Publishers (P) Ltd;
New Delhi: 2011.
Pages: 414; Price: 475/-.

Pediatric cardiology super specialty has been growing at a rapid pace specially in the last decade, and increasing number of pediatricians are opting for this specialty as their career options. In this current scenario this book is very good addition to available literature on the subject.

There are 13 chapters covering all the aspect of congenital and acquired heart diseases. Special mention must be made on the chapters on perinatal cardiology and preventive atherosclerotic disease beginning in childhood. The contents are recent, the language is lucid, and the author has reproduced quality photographs, and, judiciously used figures, diagrams and flow charts. Apart from the content, the index is accurate and the book is handy for its size; quality paper has been used, and the overall presentation is good. I would have loved to see a chapter on the non-surgical pediatric interventions which is advancing at a very rapid pace, it has been partly covered under specific lesions. I recommend this book for practicing pediatricians, post graduates, and those aspiring to take pediatric cardiology as their super-specialty.

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