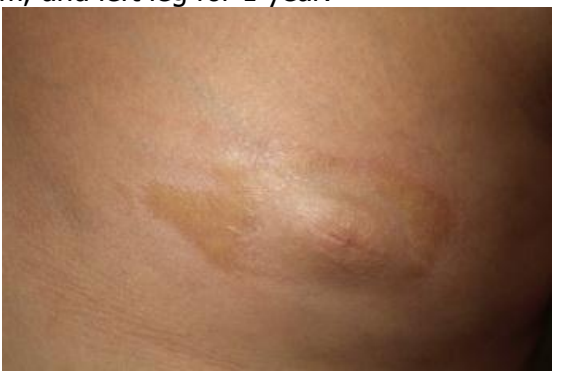
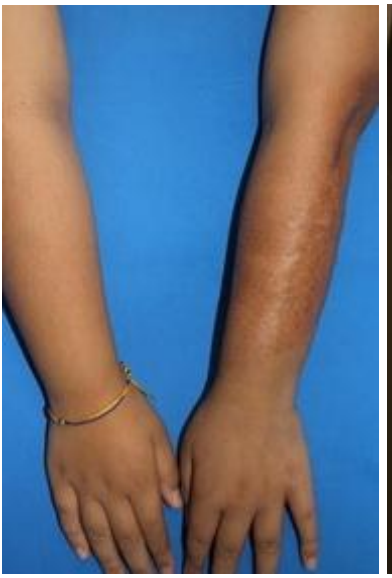
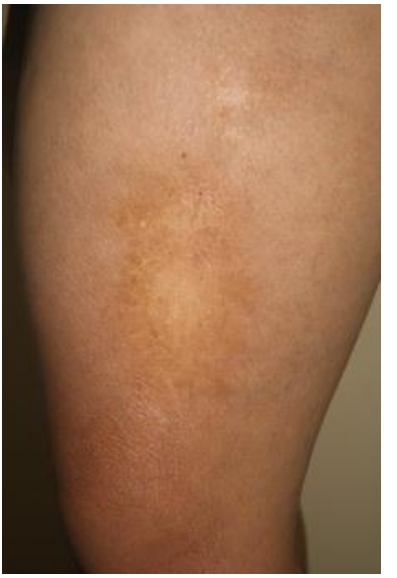
Case 25

A 7-year-old girl from Phuket.

Chief complaint: Asymptomatic sclerotic plaques on her left chest, left arm, and left leg for 1 year.







Present illness: One year previously, the patient gradually developed asymptomatic sclerotic plaques on her left chest, left

arm and left leg. She noted the lesions developed after an insect bite on her left arm. Then the lesions slowly progressed to large plaques. She denied history of dyspnea or dysphagia. There was no limitation of the joint movement.

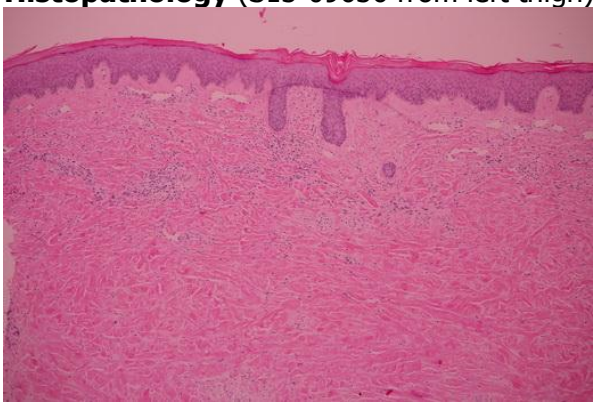
Past history: She denied any underlying disease

Family history: Otherwise normal

Physical examination: multiple sclerotic, slightly atrophic plaques with wrinkle surfaces and brownish borders on the left arm, the left leg and the left chest, no sclerodactyly, no Raynaud's phenomenon, no nailfold capillary changes.

Investigations: ANA 1:40 homogeneous, 1:40 fine speckled

Histopathology (S13-09636 from left thigh)



Thickened, closely packed, hypocellular homogeneous (sclerotic) collagen bundles, involving an entire dermis. Nodular infiltrate of lymphocytes and plasma cells in the deep dermis and subcutaneous tissue.

Diagnosis: Linear morphea

Management: methotrexate (2.5mg) 3 tabs/wk, folic acid 5mg once daily, 0.1% triamcinolone acetonide cream apply twice a day

At one month follow-up, there was slight improvement of the lesions which were softer.

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Discussion

Morphea or localized scleroderma is differentiated from systemic sclerosis by the absence of sclerodactyly, Raynaud phenomenon, nailfold capillary changes and organ involvement.[1] The pathogenesis of morphea is multifactorial, involving genetic factors and environmental exposures, which lead to small vessel damage, the release of profibrotic cytokines and disruption of the balance of collagen production and destruction. [2] Transforming growth factor-β (TGF-β) has been found to be increased in lesion of localized scleroderma. TGF-β stimulates fibroblasts to produce increase amount of glycosaminoglycans, fibronectin, and collagen while decrease extracellular matrix breakdown; and it diminishes fibroblasts susceptibility to apoptosis.[3] There are five morphea variants: circumscribed, linear, generalized, pansclerotic and mixed.[1]

Linear morphea, also known as linear scleroderma, is more common in children and can present in the first or second decade. The three most commonly described variants are en coup de sabre, progressive hemifacial atrophy also known as Parry-Romberg syndrome, and linear limb involvement. All three variants are commonly accompanied by underlying tissue atrophy.[1] Plaque formation is elongated into an appearance of linear bands which are usually transverse in the trunk and longitudinal in the extremities.[4] Linear induration involving the dermis and subcutaneous tissue may include muscle and bone. In more than 90 percent of patients, the involvement is unilateral. It may cross joint lines and sometimes leads to contractures which can be a significant cause of morbidity and deformities.[5]

Choice of therapy of morphea should be based on several factors: relative activity of the disease, depth of involvement, area of involvement and course. To date, methotrexate combined with

systemic corticosteroid and UVA1 have the most convincing data supporting their use. Both of these medications should be reserved for those patients with extensive involvement, facial involvement or involvement across joints.[2] Besides medical therapies, physical therapy is often recommended in patients with morphea, particularly in linear limb, generalized, pansclerotic variants that can cause joint contractures. (Fig. 1)

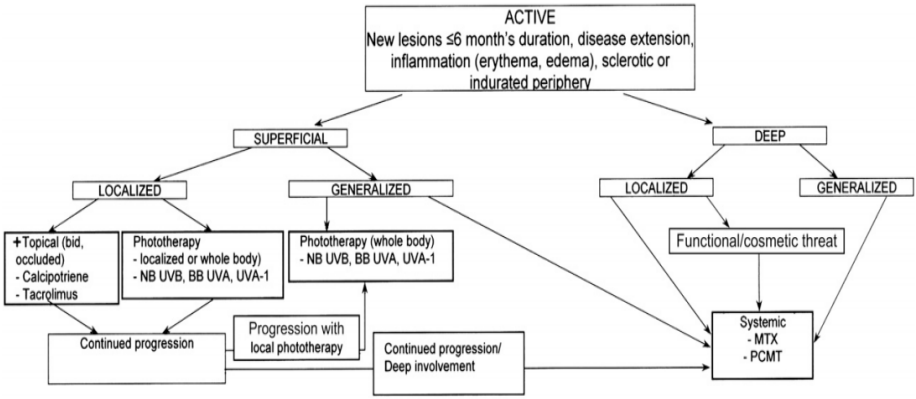

Figure 1: Therapeutic algorithm for morphea based on existing evidence.[6]

References

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