A Boy with Claw Fingers

An 8 years-old boy, presenting with clawing of left little and ring fingers for 2 years was referred to Department of Dermatology for evaluation of Hansen's disease. There was no history of trauma or loss of sensation in affected limb or elsewhere, but had history of skin tightening of left limb and few patchy areas on trunk 2 years back, which resolved within a year. The clawing of fingers followed this skin tightening. Examination revealed no hypopigmented hypoaesthetic lesions anywhere on the body and no peripheral sensory loss was appreciable. The left hand was atrophic on medial side and was notable for linear indurated lesion extending from medial arm to palm and medial two fingers. Overlying skin was hyperpigmented and could not be pinched off. Similar indurated plaques were found on trunk and medial thighs too (***Fig.*1**). All peripheral nerve trunks appeared to be normal in thickness. A clinical, diagnosis of Morphea (Linear morphea- hand lesion and classical plaque type-trunk lesions) was made. Histopathology was consistent with the diagnosis.

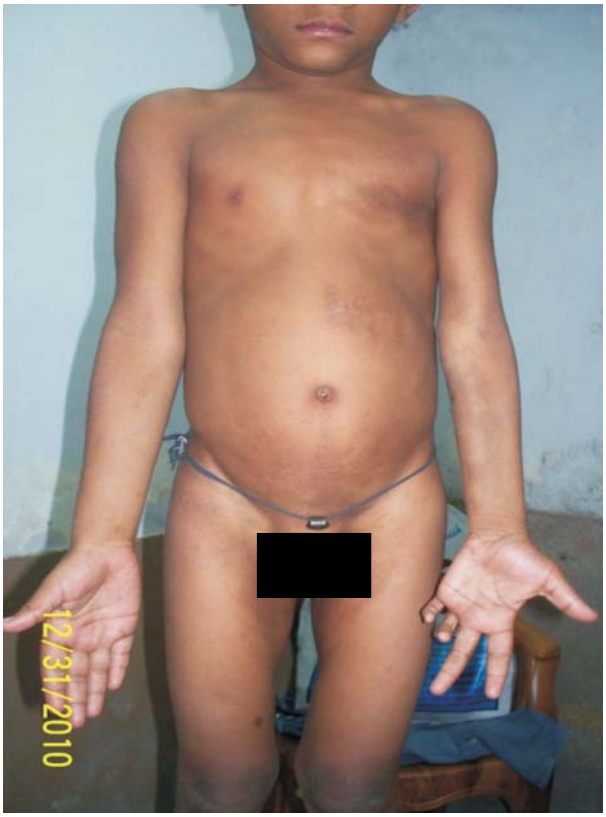

Fig. 1 *Linear indurated plaque on left forearm (linear morphea) causing atrophy of medial forearm and clawing of medial two fingers. Note indurated plaques on trunk (classical plaque type morphea).*

Morphea is a clinically distinct inflammatory condition affecting primarily dermis and subcutaneous fat, which ultimately leads to scar-like sclerosis. Classical plaque type has an asymmetric patchy distribution on trunk; acral parts are uncommonly involved. However, linear form preferentially affects extremities and is much more common in children. Usually, there is no systemic association and it is a relatively harmless disease; however, lesions affecting extremities may result in significant contractures and deformities. Linear lesions are common on face too and are known as "*morphoea en coup de sabre*". Differential diagnosis includes scleroderma (symmetric acral involvement, raynaud's phenomenon, gastrointestinal and pulmonary function compromise), Eosinophlic fasciitis (rapidly onset of edema of extremities following strenuous exercise, dry river bed sign), Lupus panniculitis (tender nodules and plaques on face, trunk and upper arms), and lichen sclerosus (predominantly affects genitalia, hypopigmented lesions, follicular plugging and some evidence of hemorrhage in lesion). Diagnosis is mainly clinical with a supportive histopathology. Early identification and treatment is necessary to avoid lifelong morbidities. Psoralen-UV A therapy (PUVA) and UV A1 therapy are particularly effective. Rapidly progressing disease may require weekly methotrexate and pulsed high dose corticosteroid therapy. The disease has a good prognosis and many lesions heal spontaneously in about 3-5 years. However, older lesions may reactivate and new lesions may appear. The major concern is irreversible fibrosis of skin and subcutaneous tissues and around joints, which necessitates early diagnosis and treatment.

ASHIM KUMAR MONDAL, NIHARIKA RANJAN LAL AND PIYUSH KUMAR
Department of Dermatology, Medical College, Kolkata.
docpiyush@gmail.com