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A Case of Paediatric Linear Morphea with Infraorbital Atrophy: A Clinico-dermoscopic-histopathologic Association

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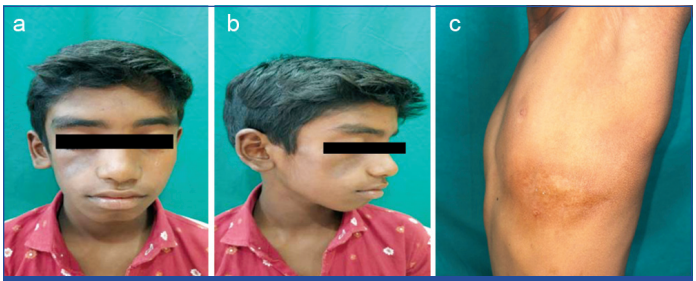
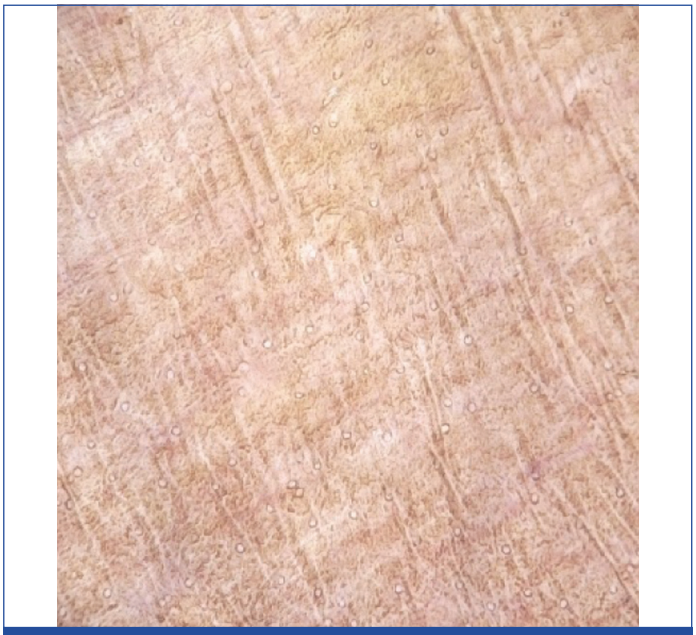
An 11-year-old male presented to the Outpatient Department (OPD) of Dermatology with depression and darkness under his right eye for the past two years. He developed a light-coloured lesion as the condition gradually progressed over a span of 1.5 years to involve the left side of the chest. Thereafter, the lesions remained stable, with no new lesions or increase in the size of existing lesions observed. Similar lesions were not noted in family members. His general condition appeared fair, and his build and weight were normal for his age.

On clinical examination, he had a 3×4 cm, well-defined, localised, oval depression with hyperpigmentation of the skin surface extending from the infraorbital region of the right eye to the mid-cheek [Table/Fig-1a,b]. He also had a 6×5 cm, ill-defined, oval, non tender, hypopigmented atrophic plaque with surrounding hyperpigmentation present over the left inframammary region extending towards the back [Table/Fig-1c]. No limb length discrepancy, bony abnormalities, tongue atrophy, gingivitis, contractures, nail fold changes, or stiffness of the fingers were noted. X-ray skull, ultrasound, and computed tomography showed no underlying bony abnormality. Routine blood tests and other investigations, including antinuclear antibody and acute inflammatory markers, were within normal limits. Hence, a provisional diagnosis of morphea was established. Differential diagnoses included systemic sclerosis, Parry Romberg syndrome (progressive hemifacial atrophy), focal dermal hypoplasia (Goltz syndrome), linear lichen sclerosus et atrophicus, linear atrophoderma of Moulin, Lupus Erythematosus Profundus (LEP), also known as lupus panniculitis, and hemifacial microsomia.



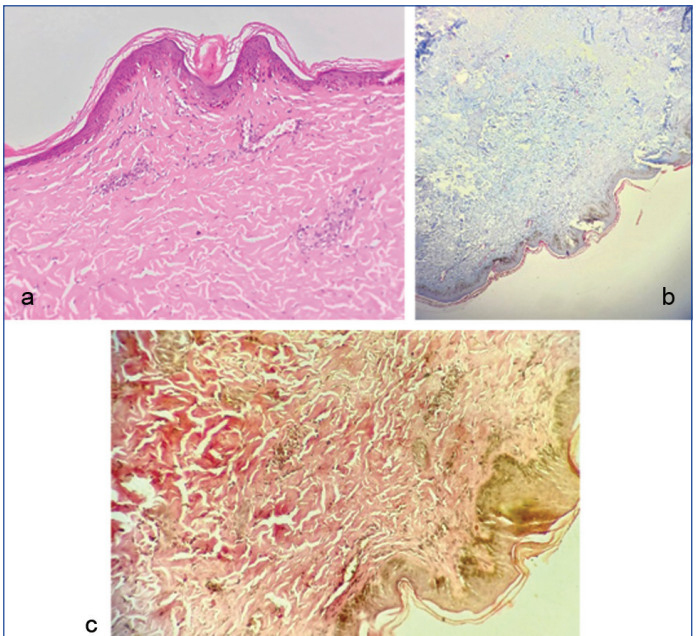
[Table/Fig-1]: a,b) 3×4 cm, well-defined, oval, noticeable localised depression of skin surface and hyperpigmentation extending from the infraorbital region of right eye to the mid-cheek; c) A 6×5 cm, ill-defined, oval, non tender, hypopigmented atrophic plaque with surrounding hyperpigmentation present over the left inframammary region extending towards the back.

Dermoscopy (80X Polarised) revealed a reticular pigment pattern, homogeneous ivory white structureless areas, linear telangiectasias, and reduced follicular openings [Table/Fig-2]. A 4 mm punch biopsy from the lesion on the left side of the chest was performed. Haematoxylin and Eosin (H&E) staining (200X) revealed atrophic epidermis, thickened collagen bundles in the papillary and reticular dermis, and mild perivascular and periadnexal lymphocytic infiltrate. Entrapped atrophic eccrine glands showed loss of surrounding adipose tissue. There was no evidence of panniculitis. Therefore, the features were suggestive of morphea [Table/Fig-3a]. Masson's



[Table/Fig-2]: Dermoscopy revealed reticular pigment pattern, homogenous ivory white structureless areas, linear telangiectasias and reduced follicular openings (80X, polarised).

Trichrome stain (200X) highlighted thickened collagen bundles in the dermis in blue [Table/Fig-3b]. Verhoeff's Van Gieson stain (200X) highlighted thickened collagen bundles in red [Table/Fig-3c].



[Table/Fig-3]: Histopathology report revealed atrophic epidermis. a) Entrapped atrophic eccrine gland shows loss of surrounding adipose tissue [H&E stain (200X)]; b) Thickened collagen bundles in dermis in blue [Masson's Trichome stain (200X)]; c) Thickened collagen bundles in red [Verhoeff's Van Gieson stain (200X)].

On the basis of the history, clinical examination, dermoscopy findings, and histopathology report using H&E stain, as well as special stains like Masson's Trichrome and Verhoeff's Van Gieson, the differentials were ruled out, and a final diagnosis of linear morphea was established.

The patient was started on excimer therapy for his chest lesion and was treated with topical tacrolimus 0.1% ointment and oral betamethasone 1 mg OD, taken at 8 am after food on weekends for a period of six months. Regular follow-up showed no further progression. The patient was advised to undergo autologous fat grafting for the under-eye lesion.

Morphea is a fibrosing disorder of the skin and subcutaneous tissue, characterised by an increase in collagen production, which progresses through two stages: an active (inflammatory) stage and a "burnt out" stage [1]. Treatment is focused on the active phase to stabilise the current lesions and prevent the occurrence of new lesions. Linear morphea is the most common form seen in childhood [1]. Its incidence is estimated to be 0.4 to 2.7 per 100,000 population [1]. Juvenile Localised Scleroderma (morphea) has five subtypes: circumscribed, linear, generalised, pansclerotic, and mixed. Rarely, systemic sclerosis and localised scleroderma may co-exist in a patient [2]. Signs and symptoms of systemic sclerosis include microvascular injury, fingertip ulcers, Raynaud's phenomenon, telangiectasia, and involvement of the lungs, heart, kidneys, and gastrointestinal tract in addition to skin fibrosis [2].

Antinuclear Antibody (ANA) positivity in a patient should therefore alert the physician [2]. Clinical scoring tools for morphea disease activity and severity include the visual analogue score, the Dyspigmentation, Induration, Erythema and Telangiectasia (DIET) score, the Modified Rodnan Skin Score (MRSS), and the Localised Scleroderma Assessment Tool (LoSCAT) [3]. The most common dermoscopic findings are white clouds, which are small, opaque, poorly demarcated areas corresponding to dermal sclerosis with increased deposition of thickened collagen fibres, and the most common vascular pattern observed is linear branching vessels [4]. Homogenisation in dermal collagen, a decrease in skin appendages, and the line sign (for lower extremities) can be used as histopathological clues in the diagnosis [5].

In the present case, systemic sclerosis was ruled out as a differential diagnosis due to the absence of Raynaud's phenomenon, eye changes, dysphagia, dyspnoea, neurological abnormalities, sclerodactyly, and calcinosis cutis, which are diagnosed through serologies, capillaroscopy, and pulmonary testing [6]. Parry Romberg syndrome (progressive hemifacial atrophy) was excluded as a differential because there were no neurological signs such as seizures or migraine, no bone and joint involvement, no associated hair loss, or the presence of moderate sclerosis [7].

Focal dermal hypoplasia (Goltz syndrome) typically presents at birth and features atrophic lesions that are distributed in a blaschkoid pattern and are not indurated. It is generally associated with other skeletal, ocular, and dental anomalies [8]. Linear lichen sclerosus et atrophicus can mimic morphea but is seen more often in the genital area and is characterised by follicular plugging, allowing for differentiation based on histopathology [9]. Linear atrophoderma of Moulin can be distinguished from morphea by the absence of dermal fibrosis on histopathology [10]. LEP, also known as lupus panniculitis, presents with tender plaques, usually shows cosmetic disfigurement, and is characterised by lobular lymphocytic panniculitis and hyaline fat necrosis on histopathology [11]. Hemifacial microsomia is characterised by facial asymmetry, which is not observed in this patient [12].

Milder cases are generally addressed with topical therapies, including corticosteroids, tacrolimus, imiquimod, vitamin D derivatives, or phototherapy [13]. Moderate-to-severe forms can be treated with immunosuppressive medications, with a methotrexate-based regimen as the first-line therapy (with or without intravenous or oral corticosteroids). Mycophenolate mofetil may be used in relapsing cases or in those that are refractory to methotrexate [13]. Cyclosporine, hydroxychloroquine, azathioprine, retinoids, intravenous immunoglobulin, rituximab, and infliximab can be effective agents in the most severe cases [13]. However, their routine use requires caution and long-term monitoring. Refractory cases can be managed with tocilizumab, sarilumab, abatacept, imatinib, pamrevlumab, Janus Kinase (JAK) inhibitors, and Autologous Stem Cell Transplantation (ASCT) [13].

Albadr F et al., reported a case of linear hypopigmented atrophic plaques in a nine-year-old girl, present over the mid-forehead, left eye, and neck, which was diagnosed at the age of seven and treated with topical tacrolimus and steroids [14]. Mendiratta V et al., reported a case of linear morphea with lipoatrophy and segmental vitiligo, which presented as a hyperpigmented plaque over the left lower limb involving the buttock in a 13-year-old girl. For this patient, oral betamethasone at 3-4 mg every week, along with 10 mg of methotrexate per week and hydroxychloroquine at 200 mg twice a day was prescribed for 4-5 years, which led to softening of the plaque with no further lesions [15]. Magri F et al., reported a case of segmental morphea in an eight-year-old boy in the left lumbar region, which was histopathologically confirmed, and treatment was initiated with topical vitamin E [16].

In conclusion, the most prevalent type of localised scleroderma in children is linear morphea, which necessitates early diagnosis to avoid cosmetic and functional consequences. Diagnosis is aided by accurately distinguishing it from imitating illnesses and using clinical, dermoscopic, and histopathological tools. The severity of the condition determines the course of treatment, which can range from topical agents for mild cases to systemic immunosuppressants for moderate to severe forms. Early, individualised, and multidisciplinary management is key to improving outcomes.

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