Case 17

A 9-year-old Thai woman from Bangkok

Chief complaint: Hypopigmented patches and plaques on left trunk, upper and lower extremities for 1 year

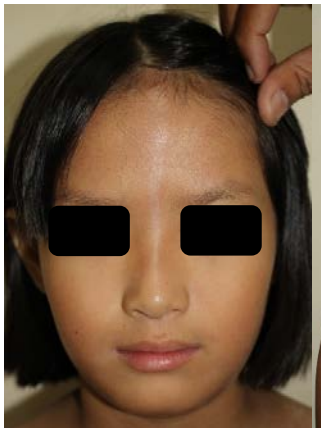

Fig 17.1

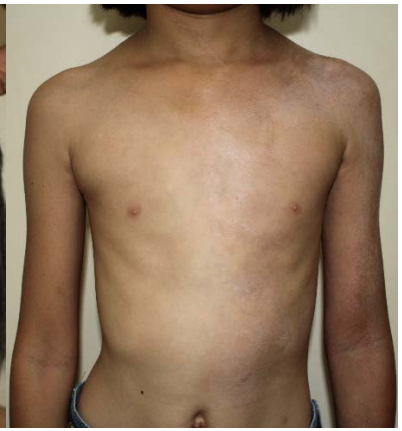

Fig 17.2

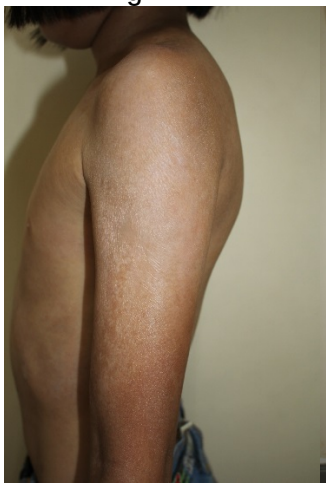

Fig 17.3

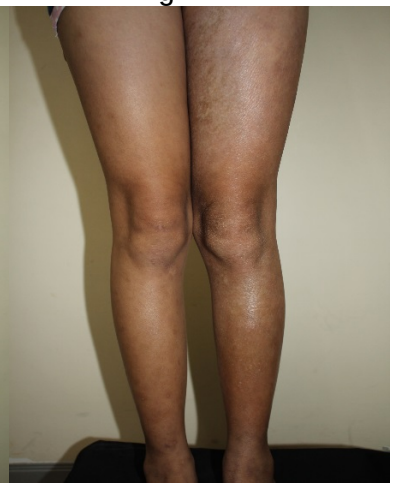

Fig 17.4

Present illness: The patient presented with a 1-year history of asymptomatic erythematous patches that initially developed on left arm and then extended to left trunk as well as left leg. The lesions gradually progressed to hypopigmented patches and atrophic plaques in 6 months prior to visit. She denied history of prolonged fever, Raynaud's phenomenon, digital pitting scar and previous treatment.

Past history

No underlying disease

Family history

No family history of autoimmune disease

Dermatologic examination

(Fig 17.1) Linear atrophic plaque on forehead

(Fig 17.2, 17.3, 17.4) Unilateral atrophic patches and plaques with hypopigmentation on left trunk, left arm and left leg, respectively

Physical examination

General appearance: A Thai female, looked-well

HEENT: No pale conjunctivae, no anicteric sclerae,

Heart: Normal S1S2, no murmur

Lung: Normal breath sound, no adventitious sound

Abdomen: Soft, not tender, no hepatosplenomegaly

Extremities: No limit range of motion on shoulder, elbow, knee or wrist joints

NS: no facial palsy, motor power grade V all extremities

Lab investigations:

- CBC:
  - WBC 10,430 cells/mm^3 (N 61%, L 32%, M 3%, E 3%, Ba 1%)
  - Hct 36.20%, Platelet 353,000 cells/mm^3
- ANA: positive homogeneous titer 1:320
- ANA 12 profile: negative all

Histopathology (S18-012204, Right arm) (Fig 17.5)

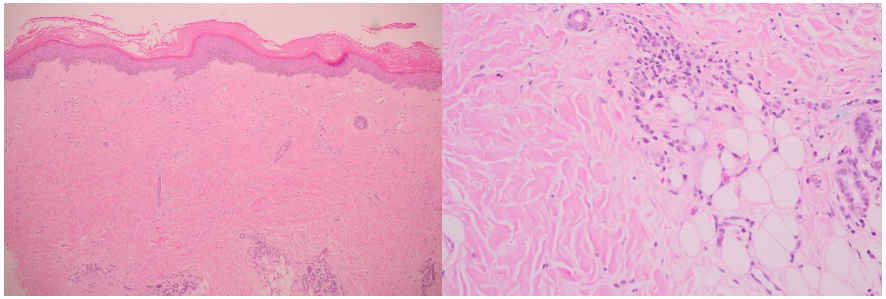

Fig 17.5

Sparse inflammatory cells infiltrate and homogenized collagen bundle in the whole dermis. The inflammatory cells composed of lymphocytes and plasma cells.

Diagnosis: Unilateral generalized morphea

Treatment:

- Methotrexate 5 mg PO once weekly
- Folic acid 5 mg PO daily
- 0.1% Triamcinolone acetonide milk lotion apply on trunk, arm, leg twice daily
- 0.1% Mometasone furoate cream apply on face daily

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Discussion

Localized scleroderma, also known as morphea, is a chronic autoimmune connective tissue disorder characterized by fibrosis of the skin and underlying tissue. Localized scleroderma is different from systemic sclerosis based on the absence of sclerodactyly, Raynaud's phenomenon, and nail fold capillary change.[1] In general, localized scleroderma involve entire dermis and usually extend to the subcutaneous fat tissue, however, it may progress to muscle, fascia and lead to muscle atrophy, growth retardation and significant contracture.[2] Visceral organ involvement is uncommon.

Commonly, localized scleroderma is classified into 5 subtypes; circumscribed (with superficial and deep variants), linear (with trunk/limb variant and head variant), generalized, pansclerotic and mix scleroderma.[1,3]

Unilateral generalized morphea (UGM) is another extremely rare variant of localized morphea. At present, there are few case report in the literature.[2,3] It is commonly found among childhood or adolescent.[4,5] The clinical characteristics consist of early onset, unilateral involvement and positive ANA (no specific antibody pattern), like our patient.[3]

The lesion begins as erythematous patch and plaque that resolve into sclerotic with varying amount post inflammatory hypo/hyperpigmentation. It then spreads to the ipsilateral side (face, neck, trunk, upper and lower extremities).[1,6]

The exact etiology and pathogenesis of localized scleroderma is unknown and may be associated with trauma, intramuscular injection, infection (measles, varicella, Epstein Barr virus, *Borrelia burgdorferi*).[1,7] In UGM, there are some case reports

show the relationship between exposure to silica and vibration in the hand ipsilateral to sclerosis.[6]

The histopathology of UGM shows disorganized collagen fiber extending to subcutaneous fat tissue. In addition, there is some atrophy of follicles and sebaceous glands.[3]

To date, treatments are based on anecdotal case reports. Due to high risk of functional disability in generalized involvement, systemic treatment is essential. Methotrexate alone, pulse high dose corticosteroid alone (PCMT), or combination with methotrexate and pulse high dose corticosteroid (PCMT) showed effective treatment, no further progression was seen in many case reports.[2,8] Low dose UVA1 therapy and physiotherapy were used as adjunctive treatments.[2,9] Plastic surgery indicate in patients with mark atrophy, contracture and facial deformity.[10]

Due to generalized involvement, methotrexate and topical steroid were prescribed in our patient. After 4 months, the lesions markedly improved without new lesion.

In conclusion, localized scleroderma has a wide clinical manifestation, UGM with generalized involvement is extremely rare variant. Early diagnosis and treatment are essential to prevent the development of functional disability and deformities.

References

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