Case 18

Multiple brownish patches involving the limb and trunk

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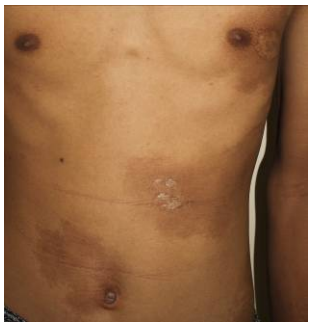
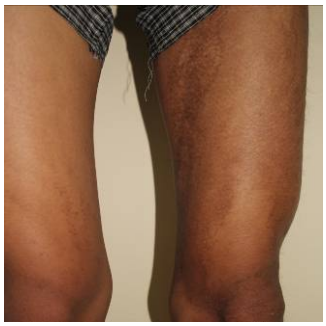
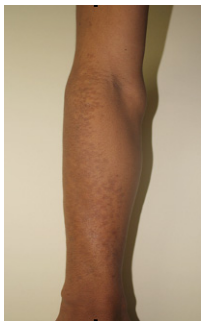
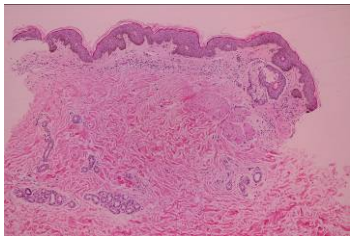
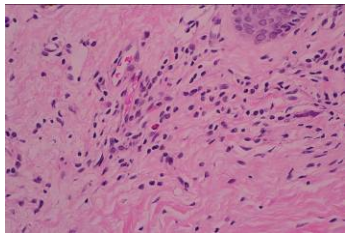
Patient: A 16-year-old Thai Boy from Nakhonratchasrima

Chief complaint: Generalized rashes for 10 years

Present illness:

The patient noticed asymptomatic brownish patches on the left leg for 10 years. The rashes had gradually enlarged and slowly progressed to the right leg, trunk and both arms. His left leg is smaller than the right side, but both legs show equal length. He did not have any lesions on genitalia.

Past history: no underlying disease

Skin examination:

- Multiple, well-defined, brown, slightly depressed macules and patches in linear distribution on all extremities
- Multiple, discrete, brown, slightly depressed, large patches on trunk, some of which lesions had atrophic porcelain white patches with wrinkled surface on the center
- The genital, perianal, and the mucosal areas were normal
- Left leg is smaller than the right side, but no length discrepancy

Histopathology: (S14-9148A, trunk; S14-9148B, forearm)
Trunk: There is superficial lichenoid infiltrate of lymphocyte in the thickened sclerotic papillary dermis.
Forearm: There is perivascular and interstitial infiltrate of lymphocytes in association with focal homogenized collagen bundles in the superficial dermis.

Diagnosis: **Coexistence of lichen sclerosus and localized scleroderma**

Treatment: Due to patient inconvenience, he was referred to other hospital for treatment.

Discussion:

Lichen sclerosus (LiS) and localized scleroderma (LoS) are classified as connective tissue diseases that primarily affected the skin.

LoS is a disorder characterized by excessive collagen deposition leading to thickening of the dermis, subcutaneous tissues, or both. LoS is classified into many subtypes: limited types of LoS, generalized types of LoS, linear types of LoS and deep LoS (deep morphea) according to the clinical presentation and depth of tissue involvement.[1]

LiS is an inflammatory dermatitis affecting primarily vulvar, perianal and perineal skin of prepubertal, perimenopausal and postmenopausal women

The classic clinical presentations of LiS and LoS are markedly different. LiS is characterized by hypopigmented, polygonal and flat-topped papules or plaques with a crinkled or cellophane-like texture. Some patients exhibit thickened, hyperkeratotic skin and accompanying eczematization due to itchiness. In contrast, LoS is characterized by an oval plaque with a shiny smooth surface, often with a purple or brown edge that indurate on palpation. Over months to years, the sclerotic plaque softens and becomes atrophic with hypo- or hyperpigmentation.

Histopathology of LiS shows an atrophic epidermis and a lichenoid lymphocytic infiltrate at the dermal–epidermal junction with homogenized collagen at papillary dermis. In contrast to LoS, the early phase demonstrates an interstitial and perivascular inflammatory cell infiltrate with lymphocytes and plasma cells in the dermis. Then there is homogenization of the papillary dermis and thickened collagen bundles extending to the reticular dermis or deeper structure. And late lesions are characterized by loss of inflammatory cell infiltrate, lessening of sclerosis, and absence of appendegeal structures. Telangiectasia may occur

The pathogenesis of both diseases is unknown. The possible coexistence of both LoS and LiS has been well known for many years.[2-4] The coexistence of LoS and LiS in the same patient suggests that these lesions represent a spectrum which may reflect similar etiologic events or closely related pathologic processes in these two diseases.[4] Although there are some similarities between LiS and LoS, their exact relationship remains debated. A retrospective analysis of 472 patients with LoS showed higher prevalence of LiS in patients with LoS, compared to healthy population, and they showed limited and generalized LoS are at highest risk for the development of LiS.[5] But LiS can rarely occur in linear and guttate subtype of LoS.[6-7] In the majority of cases, patients with LoS and extragenital LiS were reported, which is generally less common than genital LiS.[5] However, a recent prospective multicenter study from France revealed a surprisingly high percentage (38%) of genital LiS in patients with LoS.[6] Most of patients with concomitant LoS and LiS were adults. The majority of extragenital LiS cases were located on the back and shoulders.[5]

The coexistence of LoS and LiS can occur in the same biopsy specimen, the same location, or different location, usually genital LiS.[4-5, 8] It has been

estimated that up to 5% of patients with genital LiS develop vulvar squamous cell carcinoma.[9] Thus, screening these patients for LiS would have an essential impact on cancer prevention.

In our patient, He showed clinical picture of both diseases, LiS: atrophic white patches with wrinkled surface on trunk, and generalized LoS; generalized brown slightly depressed patches on trunk and extremities. And some lesions on trunk also had both clinical signs in the individual lesion. Both lesions were histologically confirmed.

The treatment is depended on the extent and depth of skin and underlying soft tissue involvement. First line treatment included topical treatment; including corticosteroid, calciprotriol and calcineurin inhibitor, phototherapy includes ultraviolet A and B. The systemic treatment is suitable for active disease and large area involvement. Methotrexate, systemic steroid or combination treatment had been effective and widely used.

References:

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