QUIZ SECTION

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A Young Girl with Diffuse White Papules: A Quiz

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A 12-year-old girl presented with a 5-month history of diffuse, asymptomatic skin eruption involving the forehead, upper and lower extremities, abdomen, chest, and back. Serology for hepatitis A, B, C and thyroid antibodies, antinuclear antibodies, and VDRL were normal or negative. She had no relevant medical history and no family history of similar skin problems.

Physical examination revealed the presence of numerous 2–4 mm, hypopigmented, slightly atrophic papules symmetrically scattered over the forehead, trunk, neck, and extremities (Fig. 1A–C). Köbner phenomenon can be seen significantly on the back and limbs (Fig. 1B, C). A biopsy specimen obtained from her back revealed hyperkeratosis with follicular plugging, atrophy of the epidermis, and homogenization of the collagen in the upper dermis. Cleft-like space separating the atrophic epidermis and dermis could be detected at the dermoepidermal junction, and lichenoid lymphocytic infiltration was observed in the mid-dermis (Fig. 1D).

What is your diagnosis?

Idiopathic guttate leukoderma leukoderma

Papular elastorrhexis

White fibrous papulosis

Generalized lichen sclerosus

See next page for answer.

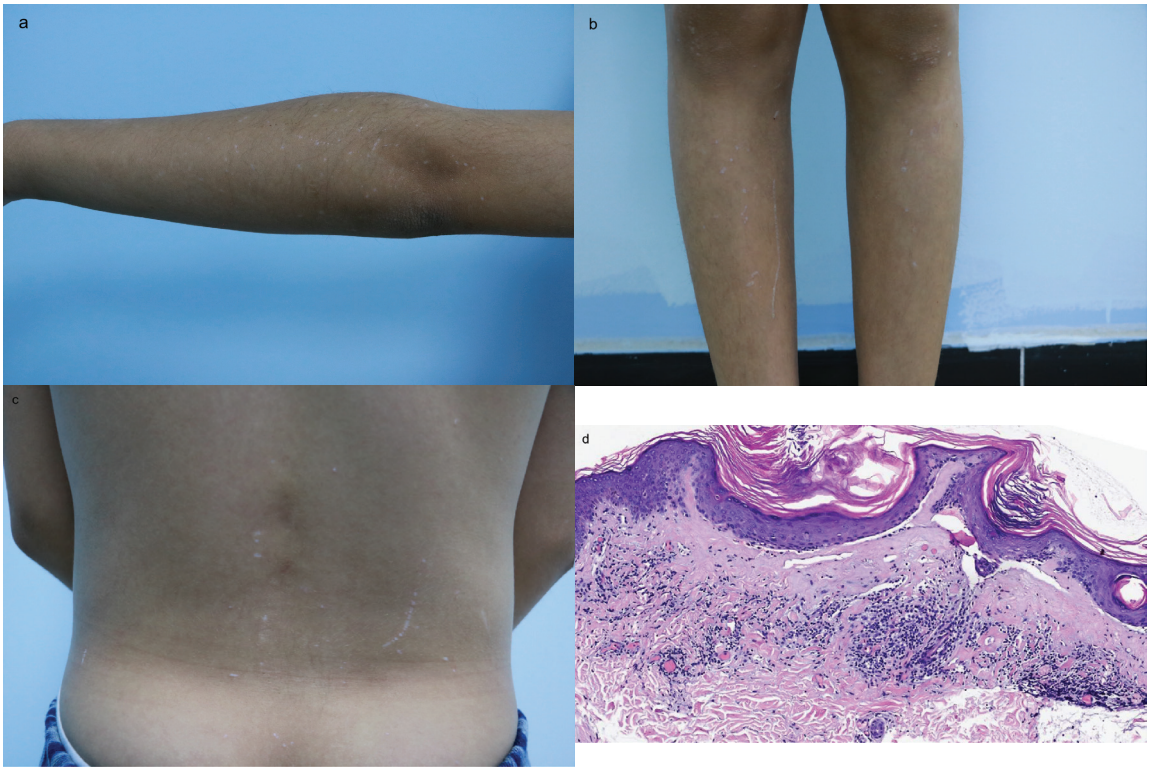

Fig. 1. (A–C) Diffuse white papules and Köbner phenomenon are visible on the patient's extremities and back. (D) Haematoxylin-eosin stain (original magnification ×100).

ANSWERS TO QUIZ

A Young Girl with Diffuse White Papules: A Commentary

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***Diagnosis*: Generalized lichen sclerosus**

Lichen sclerosus (LS) is a chronic mucocutaneous immune-mediated disease that typically involves genital skin (1). Extragenital LS (EGLS) occurs in approximately 15% of patients with LS, and can present in various anatomical areas. Paediatric LS cases represent 5–15% of affected individuals, of which 6% have extragenital manifestations (2). Generalized involvement affecting more than 2 anatomical regions is rare (3). EGLS commonly affects the upper trunk and proximal limbs. The primary lesions begin as asymptomatic to mildly pruritic polygonal white papules, usually on the upper back, chest, abdomen, or neck, which coalesce into well-demarcated erythematous plaques that slowly become atrophic over time, appearing as ivory-white patches with a tendency to scar (4). Köbnerization is very common at extragenital sites, especially at pressure points, old surgical and radiotherapy scars, and trauma sites (5).

Histopathologic features of EGLS include interface dermatitis with epidermal atrophy, orthohyperkeratosis, and follicular plugging (2). The major interventions for EGLS are topical corticosteroids, phototherapy, methotrexate, systemic glucocorticoids, and supportive interventions (1, 6). Disease activity is a critical factor for determining the approach to treatment. Signs of active disease include lesion erythema, new lesions, and expanding lesions. For patients with extensive or spreading active disease, phototherapy or systemic therapy are suggested. For patients with inactive disease, supportive interventions (emollients, wound care for fissures and erosions) rather than anti-inflammatory drugs or phototherapy are suggested. (6). In our case, considering that the lesions are extensive and Köbner phenomenon may indicate active lesions, the patient received a low dose of prednisone (0.5 mg/kg/d) orally. However, there was no improvement after a month's treatment. We observed that the lesion appeared inactive, as there was no erythema, no formation of new lesions, and no expansion of existing lesions. Consequently, we decided to discontinue systemic therapy and recommended the use of emollients. The patient continues to be monitored through regular follow-up appointments.

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