

Wide-spread Pruriginous Lesions with Flagellate Scarring

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Case Presentation

A 47-year-old man had pruriginous nodules evolving into flagellate scarring for 5 years. Family history was negative. On examination, pruriginous papules and nodules were present on the trunk and extremities associated with hypertrophic scars in a flagellate or linear configuration (Figure 1). A biopsy of a pruritic papule showed a subepidermal blister with a dermal proliferation of fibroblasts. The mutation was identified in the COL7A1 gene: c.6846G>C, compatible with epidermolysis bullosa pruriginosa (EBP). Thalidomide 100 mg/day was used for three months with some relief from itching.

Teaching Point

EBP is a hereditary disease that commonly arises in infancy or early childhood. Occasionally, it occurs in the third decade or the fifties. EBP starts as pruriginous papules or plaques

associated with intense itch. Due to scratching, makes intact blister are rarely observed. Recurrent excoriations may stimulate skin to develop hypertrophic scars in a linear or figurate configuration [1]. Histology shows cell-poor subepidermal splits or blisters. Direct and indirect immunofluorescence studies are negative. EBP can be confirmed by a mutation analysis of COL7A1, which disrupts the normal functions of anchoring fibrils and leads to sublamina densa blistering.

The late-onset and widespread lesion in EBP differs from the pretibial form of dystrophic epidermolysis bullosa. The intense pruritus in EBP may similarly appear in lichen simplex, nodular prurigo, pemphigoid nodularis, and lichen planus. The subepidermal blistering made all these skin disorders unlikely in our patients. Treatment strategies for this condition include topical and intralesional corticosteroids, thalidomide, cyclosporine, phototherapy, and dupilumab. However, most patients are refractory to them [2].



Figure 1. (A) Numerous violaceous papules and nodules are seen on the trunk and extremities with excoriations. (B) Hypertrophic scars are scattered among them or arranged in flagellate or linear configurations.

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