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Widespread pruritic papules on the trunk and extremities

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The patient

A 42-year-old Caucasian man presented with a 15-year history of a widespread, persistent, pruritic eruption over his trunk and extremities. He was not on medication and had no relevant family history. On physical examination, multiple discrete, pinkish, hyperkeratotic 1 to 2 mm papules, arranged in a rippled pattern, were detected on the trunk and extremities (Figure 1).

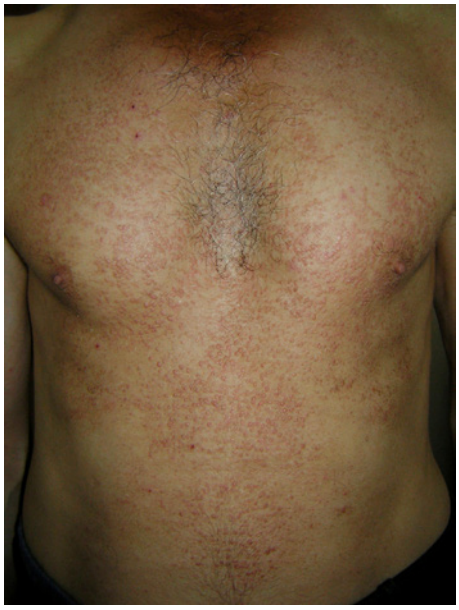

Figure 1. Multiple pinkish, hyperkeratotic papules are observed on the trunk. [Copyright: ©2014 Sezer et al.]

A biopsy was performed; photomicrographs are presented in Figure 2A, B. What is your diagnosis?

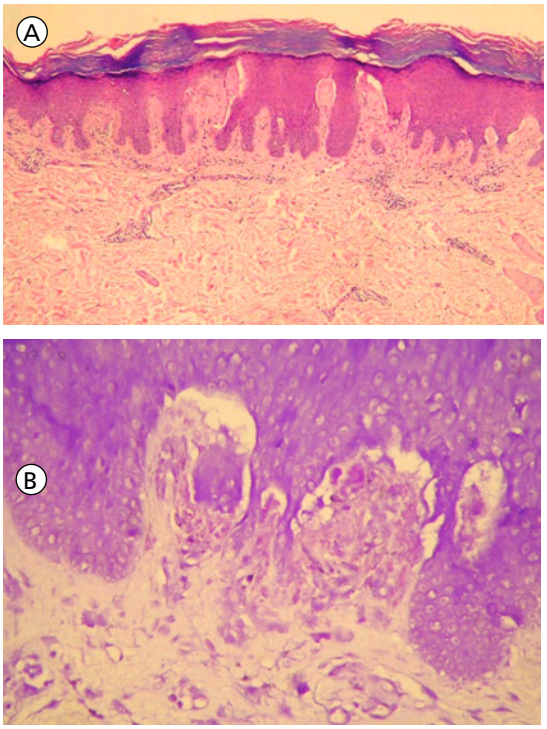

Figure 2. (A) Histopathological examination revealed amorphous eosinophilic deposits in the papillary dermis with hyperkeratosis, hypergranulosis, and marked acanthosis. (B) Crystal violet staining highlighted the eosinophilic deposits in the upper dermis. [Copyright: ©2014 Sezer et al.]

Answer and explanation

Generalized lichen amyloidosis

On histopathological examination, epidermal changes including marked hyperkeratosis, hypergranulosis, and irregular acanthosis were observed. An additional finding was the presence of eosinophilic, hyaline, homogeneous deposits confined to the papillary dermis, which showed metachromatic staining with crystal violet. Sparse melanophages were observed in the dermis. The vessels of the dermis and hypodermis were normal. A diagnosis of generalized lichen amyloidosis, based on clinical and histopathological findings, was made.

Lichen amyloidosis (LA) is a form of primary cutaneous amyloidosis without systemic involvement, characterized by extracellular deposition of amyloid material in the papillary dermis [1]. LA is mainly observed in adult patients, without gender predilection. Although the disorder is rare in Western countries, it is relatively common in Asia and some South American countries, indicating a genetic background. On physical examination, discrete, shiny, hyperkeratotic papules are observed mainly in a localized fashion over the shins and extensor aspects of the arms. Biphasic amyloidosis is characterized by overlap of lichen and macular amylodosis with tiny papules superimposed upon a background of hyperpigmented skin. In our case, lack of the hyperpigmented component helped us rule out biphasic amyloidosis.

Generalized LA is vanishingly rare with only five cases reported in the English-language literature. Compared with the conventional form, generalized LA is characterized by widespread involvement of pruritic papules over the trunk and extremities. Its association with pruritic disorders, including lichen planus and chronic urticaria in two reported cases, suggests that pruritus is a precipitating factor in generalized LA [2,3]. Compared with those cases, there was no history of a previous skin disorder in our patient. The duration of lesions in the reported cases varied between 3 and 19 years, which is in accordance with that of our patient (15 years). Lack of involvement of the axillae and the antecubital and popliteal fossae, which correspond to areas of regionally elevated temperature (monitored by infrared thermography) in a patient with generalized LA, raises the possibility of cutaneous temperature as a factor in the distribution of amyloid deposition of this entity [4]. Transformation from amorphous aggregates toward mature amyloid fibrils has been shown as a temperature-dependent process. Our patient's condition was in keeping with previously reported cases, which have shown sparing of lesions over these anatomical sides. Other atypical clinical presentations of LA include blistering and dyschromic variants, characterized by vesicles and dotted, reticular hyper- and hypopigmented spots [5].

Histopathologically, eosinophilic deposits, together with marked epidermal acanthosis, hypergranulosis, and hyperkeratosis, as observed in the skin biopsied specimen of our patient, were detected. Interface changes, including basal cell hydropic degeneration, colloid bodies, and rare satellite cell necrosis, have also been described. Subepidermal vesicles with papillary dermal eosinophilic amyloid deposits have been reported in vesicular LA.

The etiology of LA is unclear. Chronic friction, rubbing, and scratching that result in discharge of epidermal keratinocytes into the dermis and conversion into amyloid deposition have been considered in the etiopathogenesis of LA. The dermal amyloid material is thought to be derived from keratin peptides and other proteins such as apolipoprotein E2. Immunohistochemically, reaction of amyloid deposits with antihuman keratin antibodies such as CK 5/6/18 and MNF 116 also supports this hypothesis [6].

The treatment results of LA are disappointing. Topical and intralesional corticosteroid injections, topical tacrolimus, topical and oral dimethyl sulphoxide, photo(chemo)therapy, retinoids, cryotherapy, and carbon dioxide laser ablation have all been tried with mixed results [7]. However, in our patient, pruritus was markedly improved with narrow-band ultraviolet B phototherapy even though the lesions persisted post-treatment. A trial treatment of photodynamic 5-aminolevulinic acid on a small abdominal area proved ineffective. Finally, we suggest that the diagnosis of generalized LA be considered in cases of widespread, pruritic, and hyperkeratotic papules over the trunk and extremities.

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