Disseminated Superficial Porokeratosis: An Eruptive Pruritic Papular Variant

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Recently 4 cases of an eruptive pruritic papular variant of disseminated superficial porokeratosis (DSP) were reported by Japanese authors [1,2]. We describe a case with similar clinical features in a Caucasian man.

A 69-year-old apparently healthy Caucasian man presented with intensively pruritic skin lesions on the trunk and extremities. The lesions consisted in scattered, sharply demarcated, brownish-red colored, somewhere excoriated, up to 10 mm wide macules and papules (fig. 1). On the legs, some of the lesions had a verrucous surface while elsewhere they had a slightly elevated rim.

Laboratory investigations

![Fig. 1. Disseminated sharply demarcated brownish-red macules and papules on the trunk and limbs. Some of the lesions showed elevated rim (inset).]
Fig. 2. a Typical cornoid lamella of porokeratosis. b Subepidermal clefts and pronounced vacuolar degeneration of the basal keratinocytes with formation of intra-epidermal vesicles containing degranulated eosinophils with fragmented nuclei.

revealed only a slight eosinophilia 9% (0.504 1071) and elevated IgE level (nephelometry) 222.5 IU/ml (normal range up to 100). Histopathology was typical of porokeratosis, including cornoid lamellae (as many as six per section), atrophy of epidermis, thinning or absence of the granular layer, subepidermal clefts and pronounced vacuolar degeneration of the basal keratinocytes, with formation of intraepidermal vesicles containing degranulated eosinophils with fragmented nuclei (fig. 2). A moderate perivascular infiltrate with lymphocytes, eosinophils, plasmocytes and melanophages was an additional finding. Direct immunofluorescence was negative. In situ hybridization for HPV was negative.

After the failure of 0.77 mg/kg/day prednisone treatment, UVA therapy and heliotherapy, 0.64 mg/kg/day etretinate was introduced. Pruritus totally subsided and lesions gradually faded. Any attempt to taper the dosage below 0.32 mg/kg/day resulted in a relapse of pruritus and lesions. Only after 1 year could etretinate be stopped.

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Our patient was affected by the eruptive pruritic papular variant of DSP. Described by Kanzaki et al. [1] in 1992 and Tanaka et al. [2] in 1995, it has never been reported in Caucasians. Our patient differed in that he had no preexistent lesions, no colloid bodies on histopathological preparations and did not show any tendency to spontaneous resolution over a 16-month follow-up. In addition, the peripheral and tissue eosinophilia and elevated IgE level, though consistent with the pruritus, seems to belong exclusively to our patient.

References

A 35-year-old unmarried female presented with complaints of diffuse shedding of hair from the scalp, mentioning specifically that the hair would easily come out by the roots during washing and combing. The patient related the hair loss to an event that had occurred about months previously – she and her younger sister were attacked by a swarm of honeybees when they were walking through a wood. The bees which were in ‘thousands’ (in her words) bit them in all exposed parts including the scalp, face, neck, upper trunk, forearms, hands and legs below the knees. Both of them became unconscious immediately thereafter. They were admitted into a hospital with severe anaphylactic shock. While the patient survived after an in-hospital treatment of days with intravenous fluids, epinephrine and corticosteroids, her sister died immediately after reaching the hospital.

At the time of presentation, there was no obvious baldness in the scalp, there was no scaling in the scalp and the texture of the skin over the scalp was normal. Her eyebrows, eyelashes, axillary and pubic hair were normal. Telogen count, performed on a sample of hair from the vertex and occipital regions, was above 30%. Her nails and teeth were normal. The patient denied any history of hair loss before the episode of the bee stings, and she had never taken anticoagulants or cytostatic agents in the past. There was no evidence of cutaneous vir-ilism and her menstrual history was normal. Thyroid hormone levels in her blood were within normal limits. There was no history of any other stressful events in the 6 months preceding the onset of hair loss. Apart from her hair loss, she was in a good state of health.

The presentation with diffuse shedding of hair in the absence of any obvious baldness, the trichogram showing telogen count of over 30%, and the clear relation of the hair loss to the episode of multiple bee stings, which preceded it by about 4 months, favour the diagnosis of telogen effluvium in this case.

Telogen effluvium has been defined as a non-inflammatory involvement of hair, wherein some form of stress (for example surgery, parturition, haemorrhage, fever, ‘crash’ dieting, drugs, traction, emotional stress) precipitates the anagen phase into catagen and telogen phases in short order [ 1 ]. The venom of the honeybee contains histamine, mast cell degranulating peptide, melittin, phospholipase A,, hyaluronidase and acid phosphatase [2]. While a stressful event in the form of multiple bee stings leading to severe anaphylactic shock is clearly evident in our case, the possibility of a buildup of significant levels of some chemical (which may be a constituent of the honeybee venom) in the patient’s body at the time of multiple bee stings leading to or contributing to the hair loss cannot be entirely excluded.
It must be admitted here that, in our case, circumstantial evidence pointed towards the diagnosis of telogen effluvium, however, it is difficult to prove conclusively that the diffuse hair loss was due to the multiple bee stings, because it is not easy to evaluate other possible factors like endogenous hormonal alterations, accidental exposure to any chemicals, role of nutritional deficiencies and early androgenetic alopecia. It is arguable that the patient suffered telogen effluvium in association with the emotional stress of losing her sister, although the role of the manifest physiological stress is probably of greater significance.

References

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