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QUIZ SECTION

Itchy Papules and Nodules: a Quiz

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A 54-year-old male patient presented with itchy papules and nodules, which were initially located on both pretibial sides (Fig. 1). The papules and nodules soon began to disperse across the entire body surface, notably along the arms and legs. The patient reported an itch intensity score of 8 on a visual analogue scale (VAS; 0–10). Sensations such as burning, pricking and pain were reported, along with sudden attacks of itching. Symptoms appeared mainly in the evening and at night, causing severe insomnia. Earlier therapies, such as antihistamines, phototherapy (ultraviolet A (UVA)/ultraviolet B (UVB), narrow-band UVB), oral and topical steroids, anticonvulsants and immunomodulatory therapies (e.g. cyclosporine, 5 mg/kg) had not been effective.

What is your diagnosis? See next page for answer.

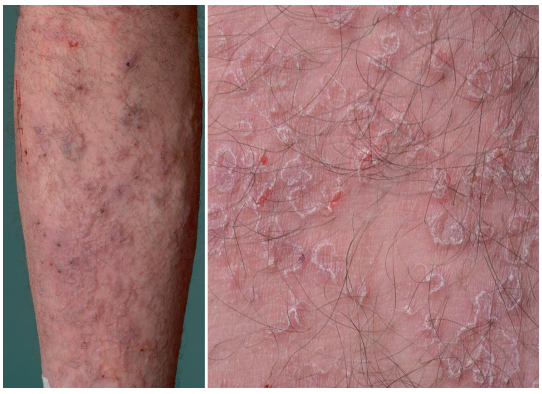

Fig. 1. Multiple, partially crusted or excoriated reddish papules and nodules with peripheral hyperpigmentation and central atrophy, together with annular, keratotic papules, distributed diffusely across the body surface.

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ANSWERS TO QUIZ

Itchy Papules and Nodules: A Commentary

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Diagnosis: Eruptive pruritic papulous porokeratosis

Porokeratoses are diseases of different aetiologies characterized by a disruption of epidermal keratinization (1). The 6 most well-known, clinical forms of porokeratoses can be classified into localized (e.g. Porokeratosis of Mibelli) and disseminated forms (e.g. disseminated superficial porokeratosis (DSP)). Eruptive pruritic papular porokeratosis (EPPP) represents a rare variant of DSP and was first described in 1992 (2). Globally, approximately 10 cases have been described to date, exhibiting a stage-like progression (3). In the initial stage, asymptomatic, slightly raised papules with a brown pigmentation form can be found (3). These can last for up to several years. In the following stage, patients with long-term DSP suddenly develop an abrupt, intensive pruritus (1) and inflammation (3). The pruritus abated over time until the final stage marked by remission with healed lesions and hyperpigmentation (3). Each stage can persist for different lengths of time (3); there is not always a spontaneous improvement in pruritus (4). In many cases, eosinophil granulocytes have been described within the inflammatory infiltrate, in addition to classical histological features of porokeratoses (Fig. 2). Thus, it seems plausible that the origins of pruritus can be attributed to inflammation and, in particular, to the eosinophil granulocytes in the tissue (3). Eosinophil granulocytes release, among other mediators, neuropeptides that are involved in the development of pruritus, such as substance P (5), which causes itch by binding to mast cells and releasing histamine. Antihistamines can therefore provide alleviation of EPPP to a certain extent. Although a perivascular infiltrate was histologically detectable in our patient, there was no clinical response to antihistamines.

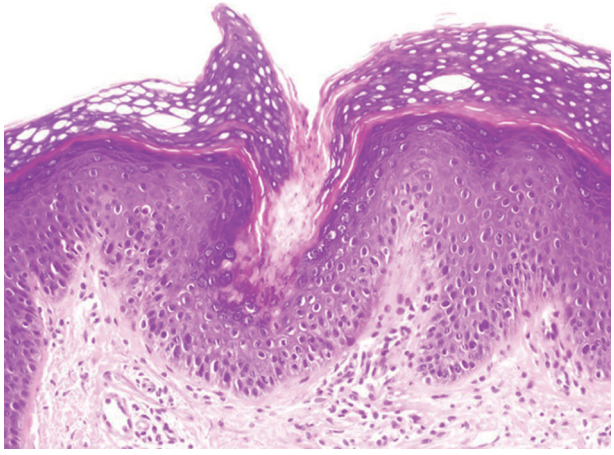

Fig. 2. Histologically, pseudoepitheliomatous hyperplasia with compact orthohyperkeratosis and a central column of parakeratosis with underlying vacuolization and dysmaturation of keratinocytes were observed (original magnification × 400).

Whilst our patient had had EPPP for over 12 years, he developed prurigo nodularis from the resulting endless scratching due to the severity of the pruritus. This masked the porokeratosis, hindering diagnostic identification and possible therapies. Prurigo nodularis is a reaction to severe chronic pruritus triggered by constant scratching (6) and characterized by the presence of itchy papules and nodules (6). Until now, no association with porokeratosis has been reported. Other cases have been described in which prurigo nodularis has concealed dermatoses, e.g. dermatitis herpetiformis (7) and bullous pemphigoid (8). Treating prurigo nodularis remains difficult. Treatment is based on both topical and systemic therapies in order to permit the nodules to heal and to suppress the pruritus (9). Treatment for EPPP itself is identical to treatments for other porokeratoses. With neuropeptides in mind, we treated our patient with aprepitant, a neurokinin-1 receptor antagonist that blocks binding of substance P. After a treatment period of one week, there was a remarkable recorded decline of 4 points on the VAS and no new acute scratch marks.

In conclusion, even rare dermatoses, such as EPPP, as presented here, can be masked due to prurigo nodularis. In such cases, histological examinations can provide valuable insight into the underlying dermatosis that the prurigo nodularis is obscuring.

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