Multiple Brownish Nodules with Blisters on the Trunk and Extremities

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CASE REPORT

A 79-year-old gentleman presented with a 1-year history of many pruritic nodules on the trunk and extremities. Some blisters developed for several weeks before he visited our clinic. On physical examination, there were about 60 discrete, hyperkeratotic, brownish nodules on the trunk and four limbs (Fig. 1). Besides, there were several tense blisters right on some of the prurigo lesions and the normal skin (Fig. 2). No oral erosion or blister was found. Routine investigations including complete blood count, differential counts of white blood cells, renal function and liver function tests were all within normal limits. The biopsy specimen from the blister showed subepidermal vesicle with lymphocyte and eosinophil infiltration in the upper dermis. The other biopsy specimen from the prurigo lesion showed features of prurigo nodularis (Fig. 3). Direct immunofluorescence (DIF) studies of both the peri-blister and prurigo skin revealed linear IgG and C3 depositions at the basement membrane zone (BMZ) (Fig. 4). Indirect immunofluorescence (IIF) studies of his serum detected anti-BMZ autoantibodies. Besides, his serum autoantibodies bound to the epidermal side on salt-split normal human skin. Western blotting using human epidermal extract as substrate revealed the presence of serum autoantibodies against 230 kD antigen (BP230).

Fig. 1
There were many discrete, hyperkeratotic, excoriated, brownish nodules on the back and upper arms.

Fig. 2
Two tense vesicles were noted near the excoriated brownish nodule.

Fig. 3
Histological examination of the nodular lesion showed hyperkeratosis, irregular acanthosis and the elongation of rete ridges. Perivascular lymphohistiocytic infiltration and fibrosis were noted in the dermis. (H&E; x40)

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Fig. 4
DIF of the prurigo lesion showed linear depositions of IgG antibodies. (x200)

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DIAGNOSIS: Pemphigoid Nodularis

DISCUSSION

Several distinct variants of bullous pemphigoid have been described, including localized, vesicular, polymorphic, vegetans, cicatricial, erythrodermic, and nodular forms. The latter, known as pemphigoid nodularis, is characterized by itching prurigo nodularis-like lesions and tense blisters. The tense blisters may arise at the site of the nodules and/or normal skin. The diagnosis depends on the histologic features of prurigo nodularis and DIF findings of linear C3 and/or IgG depositions at the BMZ. Most patients have detectable circulating anti-BMZ antibodies, which may be against the 230 kD antigen (BP230), the 180 kD antigen (BP180), 220 kD antigen, or 150 kD antigen. Pemphigoid nodularis predominantly affects old women and is relatively resistant to therapy.

The coexistence of prurigo nodularis and bullous pemphigoid was initially observed by Provost et al. in 1979. However, the term “pemphigoid nodularis” was first described by Yung et al. in 1981. From 1979 to 2006, more than 30 cases of pemphigoid nodularis have been reported. The mechanism of pemphigoid nodularis remains unknown. Since most patients develop prurigo nodularis before the onset of blisters, it is hypothesized that scratching alters or exposes the encrypted BMZ antigens, resulting in the formation of autoantibodies. It is also possible that these two diseases coexist incidentally.

Another explanation is that subclinical pemphigoid with smoldering inflammation presents with pruritus, leading to the formation of prurigo nodularis. There are some patients of pemphigoid nodularis who never developed blisters. Thus, DIF and IIF studies for persistent prurigo nodularis patients should be performed for early diagnosis of pemphigoid nodularis.

The successful management of pemphigoid nodularis often requires systemic prednisolone, azathioprine, dapsone, or sulfamethoxypyridazine. Our patient didn’t have new blister developed after 2 weeks of oral prednisolone (20 mg QD), and all the prurigo lesions flattened significantly. The dosage of steroid was tapered and discontinued 2 months later. His condition remained stationary without evidence of recurrence during the ensuing 1 year.

REFERENCES