QUIZ SECTION

Tense Bullous Eruptions Arising in Porokeratosis: A Quiz

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A woman in her 60s, with a history of Sjögren's syndrome and rheumatoid arthritis, developed autoimmune hepatitis and primary biliary cholangitis approximately 4 months prior to her first visit to our department. Steroid pulse therapy was administered for these conditions, followed by oral prednisolone, which had been reduced to 8 mg/day by the first visit. During the treatment she noticed round red patches, mainly on her left lower leg. At our clinic, multiple erythematous annular plaques with scaly borders were observed on the lower legs and shortly thereafter spread to the thighs, through both visual and dermoscopic examinations (Fig. 1A, B). A skin biopsy revealed thinned epidermis with cornoid lamella and mild inflammatory cell infiltration in the papillary dermis (Fig. 1C, D). These findings led to a diagnosis of eruptive disseminated porokeratosis, possibly associated with corticosteroid-induced immunosuppression. Vitamin D3 ointment was prescribed. However, approximately 2 weeks later, the patient returned for an unscheduled visit, complaining of tense blisters on the erythematous lesions (Fig. 1E).

What is your diagnosis?

1: Bullous pemphigoid
2: Pemphigus vulgaris
3: Contact dermatitis with vitamin D3 ointment
4: Bullous eruptive disseminated porokeratosis

See next page for answer.

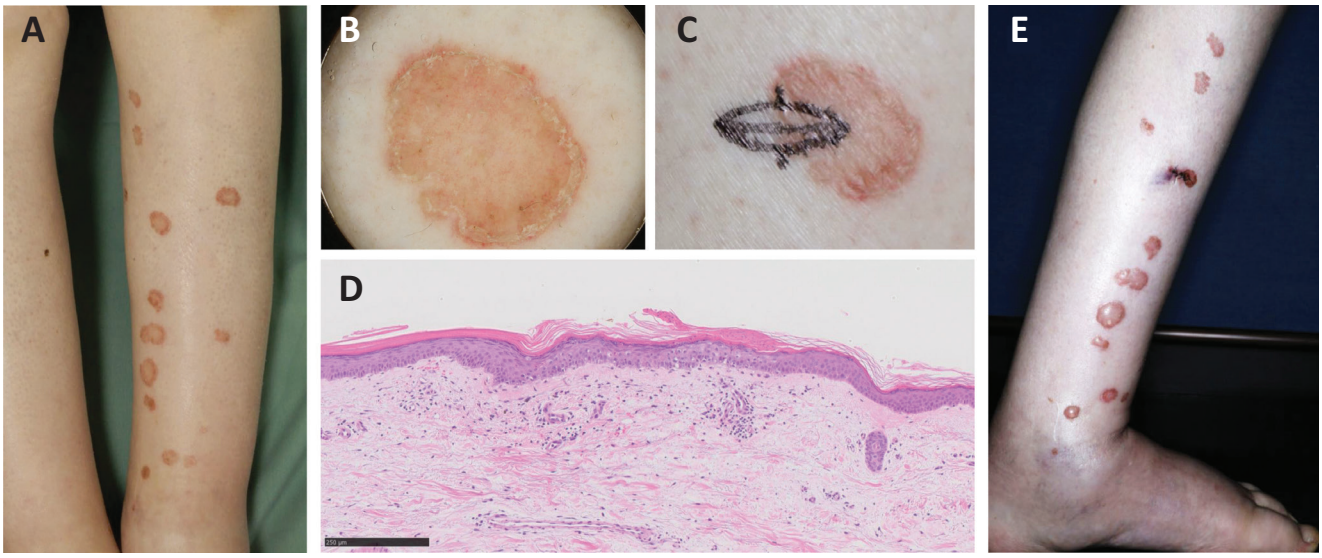

Fig. 1. Clinical, dermoscopic, and pathological findings of the patient. (A) Clinical manifestation on the left leg at the first visit. (B) Representative dermoscopic image of the lesion. (C) Biopsy site. (D) Haematoxylin and eosin-stained tissue histology slide with a characteristic cornoid lamella. Scale bar: 250 μm. (E) Clinical manifestation on the left leg at the unscheduled visit due to blister formation.

ANSWERS TO QUIZ

Tense Bullous Eruptions Arising in Porokeratosis: A Commentary

Acta Derm Venereol 2025; 105: adv43898
DOI: 10.2340/actadv.v105.43898

***Diagnosis*: Bullous eruptive disseminated porokeratosis**

When the tense blisters appeared, pitting oedema in her lower legs clearly worsened. Interestingly, the blisters were observed only on the distal portion of the lower legs, where the oedema was most pronounced. Hypoalbuminaemia (2.9 g/dL) and lifestyle habits such as prolonged sitting and standing were considered causes of the oedema. Bullous pemphigoid (BP) and pemphigus vulgaris were ruled out, as serum anti-BP180, anti-desmoglein 1, and anti-desmoglein 3 antibodies tested negative. Contact dermatitis due to vitamin D3 ointment was also excluded, as the blisters were limited to the distal porokeratosis lesions and there were no eczematous changes on the surrounding skin, despite the ointment being applied over a wide area of the legs. Viral diseases such as herpes simplex or herpes zoster were deemed unlikely because the lesions were not painful. A skin biopsy was not repeated because it had just been performed at the initial visit, and the oedema was thought to have caused the blisters. Compression stockings and elevation of the lower extremities led to an improvement in oedema, which subsequently resulted in the disappearance of the blisters.

Based on the above, this case was diagnosed as bullous eruptive disseminated porokeratosis (BEDP), a condition reported in 2 publications to date (1, 2). In both cases, similar to ours, tense blisters were predominantly observed in porokeratosis lesions on the lower legs, associated with pitting oedema. Although the reason for the tendency of blisters to form in porokeratosis remains unknown, all 3 cases indicate that abnormalities in the structure of the basal membrane may be present in porokeratosis lesions. Indeed, careful observation of pathological samples of porokeratosis sometimes reveals separation of the epidermis at the basal layer (data not shown). These abnormalities could facilitate the entry of interstitial fluid into porokeratosis lesions, leading to the formation of tense blisters. Such symptoms should be differentiated from BP, which has been reported to co-occur with porokeratosis (3). However, tense blisters caused by oedema in BEDP occur only at the sites affected by porokeratosis, whereas in BP, blisters can develop in areas unrelated to porokeratosis. This distinct pattern enables a differential diagnosis through visual examination. As the oedema decreases, the blisters resolve accordingly, allowing management with compression therapy or lifestyle modifications alone, without repeated biopsies, additional oral medications, or discontinuation of the ointment in use. This approach is beneficial for both the patient's well-being and healthcare economics.

The authors have no conflicts of interest to declare.

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