Pretibial Epidermolysis Bullosa with Vulvar Involvement

Sir,

As venereologists are often confronted with patients with long-standing genital complaints, it is of importance to be aware of the possibility of a chronic bullous disease with manifestations in the ano-genital area. Hereditary epidermolysis bullosa is the name of a group of disorders characterized by the formation of blisters following minor trauma (1). A rare variant is pretibial epidermolysis bullosa, a probably dominantly inherited disorder with onset at a young age, usually between 11 and 24 years. Slowly healing pruritic papulo-nodular crural lesions are usually found (2, 3). A woman with pretibial epidermolysis bullosa developed severe macerative bullous lesions on the vulva and perigenital area as part of the clinical manifestations.

CASE REPORT

A 45-year-old otherwise healthy woman was referred to the clinic due to a painful blistering eruption of 3 months' duration, located on the vulva. The history was unremarkable except for a tendency since childhood to develop slowly healing bullous lesions on the pretibial areas after even minor trauma. None in the family had similar skin problems. Clinical examination revealed remnants of bullae, erosions and erythema in the vulval area (Fig.1). Milia were noticed in the genital lesion. Milia, cicatrices and slight erythematous nodular lesions could be detected on the crura (Fig. 2). A single bulla was seen on the medial aspect of the left lower leg. The nails were dystrophic, but hair and teeth were uninvolved. Pathogenic bacteria, *Candida albicans* or herpes simplex virus could not be demonstrated in the genital lesions. A histopathological examination of biopsy specimens from the genital and crural lesions showed subepidermal bulla formation with no or only slight lymphohistiocytic inflammation. Direct immunofluorescense findings of perilesional skin were negative for immunoglobulins and complement. Electron microscopic examination showed that epidermis was separated from dermis. A large blister had formed under the epidermis (Fig. 3). The blister roof was epidermis with basal lamina. Remnants of the dermal tissue were attached to the basal lamina. Anchoring fibrils were not found. The blister floor was dermal connective tissue. Symptomatic treatment with mupirocine ointment to the ulcerated lesions was initiated, with good symptomatic effect. Varying vulvar lesions are still seen after 4 years.

DISCUSSION

The history and clinical findings in our patient are in accordance with the diagnosis of pretibial epidermolysis bullosa (1). None of the previously described cases have shown ano-genital bullous lesions. Pretibial epidermolysis bullosa is considered a minor or localized variant of dominant dystrophic epi-

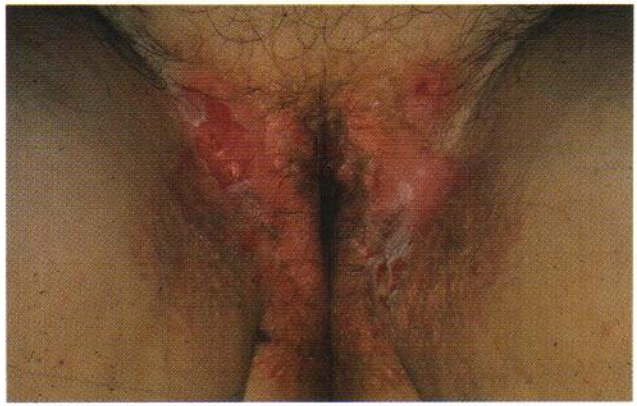

Fig. 1. Vulval erosions and erythema in a patient with pretibial epidermolysis bullosa.

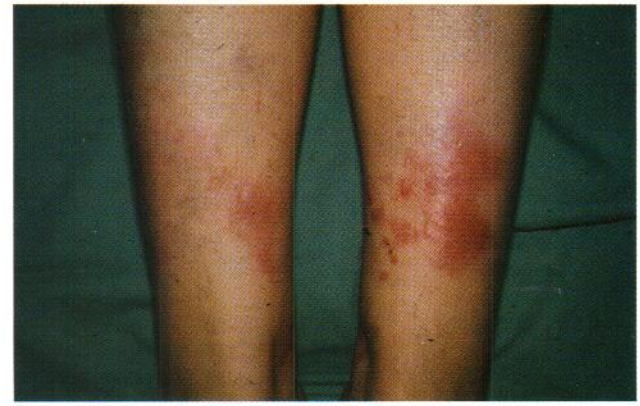

Fig. 2. Pretibial erythematous lesions in a woman with pretibial epidermolysis bullosa.

dermolysis bullosa. In this disease (5), as in cases of pretibial epidermolysis bullosa (2), rudimentary anchoring fibrils and a decreased number of fibrils have been detected in apparently normal skin. Why our patient had vulvar lesions remains unclear, except for the patient's moderate adipositas.

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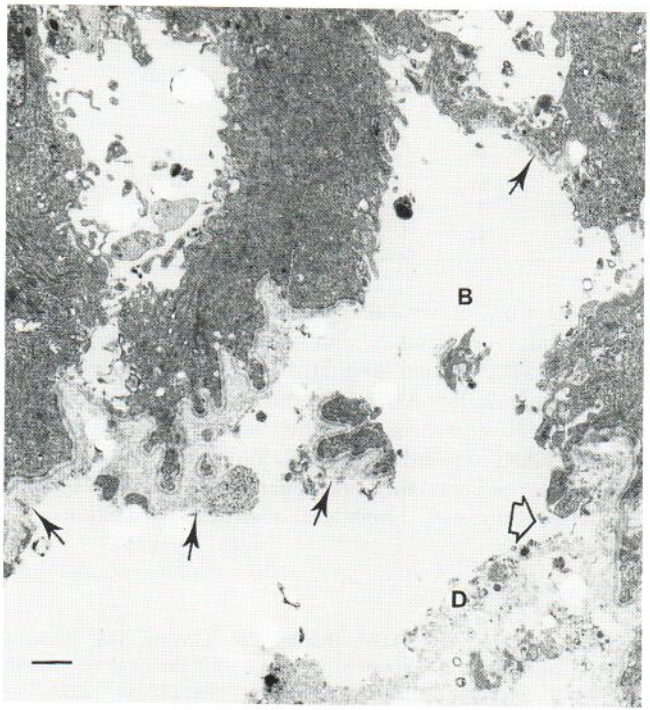

Fig. 3. A blister under the epidermis. Blister cavity (*B*). Basal lamina remnants of dermal tissue in the blister roof (*arrows*); the blister floor was dermal connective tissue (*D*). The edge of the blister (*framed arrow*). Scale indicates 1 mcm. × 6000.