Pretibial Epidermolysis Bullosa with Vulvar Involvement

Sir,

As venereologists are often confronted with patients with longstanding genital complaints, it is of importance to be aware of the possibility of a chronic bluossus 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 vulgar 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 immunofluorescence 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 epithelium 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 mupirocin 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.
dermolyssis bullosa. In this disease (5), as in cases of pretibial epidermolyis bullosa (2), rudimentary anchoring fibrils and a decreased number of fibrils have been detected in apparently normal skin. Why our patient had varal lesions remains unclear, except for the patient's moderate adipsias.

REFERENCES

Accepted August 15, 1995.
Carsten Sand Petersen, Kim Brooks, Kaare Weismann, Takayasi Kobayasi and Henrik Klein Thomsen
Departments of Dermatology and Venerology, Bispebjerg Hospital and Rigshospitalet and Department of Pathology, Bispebjerg Hospital, University of Copenhagen, DK-2400 Copenhagen NV, Denmark.

Intralesional Bleomycin for the Treatment of Non-genital Warts in HIV-infected Patients

Sir,
Warts in individuals infected with the human immunodeficiency virus (HIV) are common, frequently disfiguring and a cause of considerable distress to those afflicted. There is a poor response of warts to conventional therapies in HIV-infection, which may reflect the inherent immune dysfunction. Bleomycin has been shown to be effective in treating resistant warts (1, 2) and warts in immunocompromised transplant patients (3). We have investigated the efficacy of direct intralesional delivery of an antimetotic agent (bleomycin) to treat warts in patients with HIV-infection.
Four HIV-infected patients with long-standing, disfiguring hand and finger warts were treated. The mean patient age was 54.7 years, with known duration of HIV-infection from 6 to 8 years (mean 7.25 years). CD4 cell counts ranged from 0.008 to 0.35 x 10⁹/l (mean 0.23 x 10⁹/l). Ten warts from 2 mm to 12 mm diameter were treated, with 10 untreated warts used as controls.
Using a careful, aseptic technique with the physician wearing suitable clothing and eye protection, selected warts were anaesthetised with plain 1% lidocaine. A multiple puncture technique (4) was then used to inoculate bleomycin sulphate

(1 U/ml dose range 0.1 to 0.4 ml depending on wart size) into the wart tissue.
Warts were treated with bleomycin sulphate at 3 weekly intervals for up to 3 cycles of treatment. Warts of similar size and duration on the same subject were selected as controls and received no treatment. Response, side-effects and patient satisfaction were assessed.
Overall, complete resolution was observed in 5 warts, with partial resolution in 5 warts (Table). No regression of the control warts was observed during the study period.
The procedure was well tolerated in all subjects. Side-effects were limited to local pain in all subjects, and one subject experienced mild hand swelling. Simple oral analgesia was required after the procedure in 2 subjects. Three of the subjects were keen to have the treatment again and completed 3 cycles of therapy. All 4 subjects were pleased with the results of treatment.
Intralesional bleomycin for the treatment of recalcitrant warts, unresponsive to conventional therapy, is a well-tolerated procedure with a good response rate in HIV-infected individuals. The mechanism by which bleomycin acts is as yet unknown. However, its exact mode of action may be related

Acta Derm Venereol (Stockh) 1996