Erosive Pustular Dermatosis of the Scalp: A Case Treated Successfully with Isotretinoin

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Sir,

Erosive pustular dermatosis of the scalp (EPDS) is a rare skin disease seen in elderly people. The disease is characterized by chronic pustular and eroded lesions with crusts confined to the scalp that heal with scarring alopecia. It is often preceded by trauma to the skin. We describe here an elderly woman with sun-damaged skin, who developed a wound on the scalp. After visiting several doctors and departments a diagnosis of EPDS was suspected and treatment with oral isotretinoin was initiated with a good final result.

CASE REPORT

An 89-year-old woman developed itchy scaly changes in the skin of the left temple over a period of several months. The lesions progressed to erosions and superficial wounds with severe crusting and purulent discharge. A dermatologist found a superficial scalp wound measuring 10×5 cm in diameter and actinic keratoses in the surrounding skin (Fig. 1a). A wound swab revealed *Staphylococcus aureus* and she was treated with oral dicloxacillin with no effect. After referral to a wound care centre at the University Hospital, a skin biopsy was taken from the lesional area, which raised suspicion of pemphigus vulgaris; however, a later immunofluorescence staining was negative. She was subsequently referred to the department of dermatology, where topical treatment with Fucicort[®] (betamethasone 17-valerate and fusidic acid, LEO Pharma, Ballerup, Denmark) and later Betnovate[®] (betamethasone 17valerate, GlaxoSmithKline Pharma, Ballerup, Denmark) was initiated because of a seemingly inflammatory component. New skin biopsies showed actinic keratosis and non-specific dermal inflammation with atrophy and ulceration. In particular there were no signs of vasculitis, malignancy or dysplasia. The only biochemical abnormalities were a slight anaemia with haemoglobin 6.5 mmol/l (normal range 7–10 mmol/l) and slightly raised creatinine 148 µmol/l (normal range 44-116 µmol/l). Serum zinc was reduced to 7.7 µmol/l (normal range 9.9-19.1 µmol/l). Scrapings were negative for fungi and no herpes simplex was found. S. aureus was repetitively isolated from the lesion and she was given several courses of antibiotics with dicloxacillin (Sandoz A/S, Odense, Denmark) and later roxithromycin (Surlid[®], Aventis Pharma A/S, Hørsholm, Denmark) combined with Aquacel Silver (Convatec, Lyngby, Denmark) as a wound dressing. In spite of these treatments she still had a $10 \times 15 \times 0.5$ cm area with erosions, crusts, pustules and purulent discharge one year after her initial symptoms. Treatment with topical steroids had some effect, but the eroded area enlarged as soon as treatment was withdrawn.



Fig. 1. The patient (a) before and (b) after treatment with isotretinoin.

Based on the clinical picture, with poor healing, superficial ulceration in sun-damaged skin and refractoriness to treatment, we considered a diagnosis of EPDS. The patient started treatment with isotretinoin 20 mg orally daily (0.3 mg/kg) increasing to 40 mg daily; after 2 weeks this was supplemented with Zinklet[®] (Gunnar Kjems ApS, Copenhagen, Denmark) OTC (containing 22 mg zinc) twice daily. She was switched to a milder topical corticosteroid Locoid[®] (hydrocortisone butyrate 0.1%, Yamanouchi, Leiderdorp, Holland). After one month the erosions and superficial ulcer had healed with alopecia (Fig. 1b). Actinic keratoses persisted in the surrounding area.

DISCUSSION

The sun-damaged skin, together with the clinical and histological features, in this patient led us to suspect a diagnosis of EPDS. This entity was first described in 1977 by Burton (1). Since then approximately 40 cases have been reported (2, 3). It tends to affect elderly white women and there is often a precipitating history of trauma to the skin, including sun-damage, skin grafting, laser, cryotherapy and X-ray therapy (2-4). Up to several years after the skin damage a wound develops in the scalp with erosions, pustules, crusts and alopecia. Usually skin swabs and scrapings are negative for bacteria and fungi. If micro-organisms are found it probably represents a secondary colonization rather than primary infection. Biopsies often show histology with atrophy and chronic inflammatory changes with a dermal infiltrate of lymphocytes and plasma cells (3, 5).

In one case a patient with EPDS developed a squamous cell carcinoma in the wound (6). Another case was associated with Hashimoto's thyroiditis, autoimmune hepatitis and Takayasu's aortitis in the same patient, suggesting an autoimmune mechanism (7).

Often there is a good response to potent topical corticosteroids, but typically with a relapse when treatment is stopped. The following treatment regimens have been reported to be effective: oral isotretinoin, zinc sulphate, nimesulide, topical tacrolimus or calcipotriol (3, 8–10).

The effect of isotretinoin has been documented (3). The drug has a well-known anti-inflammatory effect on the pilosebaceous unit of the skin and a normalizing effect on the keratinization, which theoretically could have an effect on the inflammation and dyskeratinization of the affected skin areas.

Our patient was treated successfully with isotretinoin as systemic therapy in combination with a low dose of zinc sulphate. This treatment was chosen because tacrolimus and calcipotriol can be locally irritating and our patient had erosive, ulcerated skin lesions. In addition, the patient had sun-damaged skin and the carcinogenic potential of tacrolimus in such a skin is unknown. We have followed the patient for 6 months after her wound has healed and are now treating her actinic keratoses with cryotherapy, taking care not to precipitate the EPDS. No relapse has occurred.

The authors declare no conflicts of interest.

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