Erosive Pustular Dermatosis of the Scalp

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Introduction

Erosive pustular dermatosis of the scalp (EPDS) is a rare chronic disease characterized by sterile painful pustules, erosions and crusts on the scalp. It was first described in the late 1970s by Burton (1) and Pye (2). EPDS affects mainly, but not exclusively, elderly women (mean age 70 years). Young adults can also be affected (3, 4). Lesions extend slowly over months or years, leading to marked cicatricial alopecia (3, 5). EPDS has a protracted evolution, responds poorly to treatment, and has a negative aesthetic impact. Evolution to local squamous cell carcinoma occurs exceptionally (6), probably related to pre-existing lesions, such as actinic keratoses. Regular follow-up is therefore advised. The condition is not well-known, and is therefore often overlooked or misdiagnosed; it may be years before patients are offered adequate treatment (5).

Triggering factors

Underlying androgenetic alopecia and actinic damage are not rare (3). Injury to the scalp (5) is often reported to have occurred months before onset of symptoms. Causes of injury include trauma, wounds (e.g. due to herpes infection), contusions, local burns (e.g. due to irritative local/chemical treatment, such as 5-fluorouracil cream, or topical tretinoin, liquid nitrogen/cryosurgery, photodynamic therapy, CO_2 laser, or radiotherapy), surgery, skin grafts, prolonged exposure of a bald scalp to ultraviolet (UV) light, and zinc deficiency (5). In addition, various autoimmune conditions have been reported to be associated with EPDS (e.g. rheumatoid arthritis, Hashimoto thyroiditis, and autoimmune hepatitis).

Clinical signs

Clinically EPDS affects mainly the vertex. The scalp becomes inflamed, with yellow pustules, yellow or haemorrhagic crusts, atrophy of the scalp skin, hair thinning, which slowly evolves into progressive cicatricial alopecia (3) (Figs 1–2). Association with erosive pustular dermatosis (EPD) of the legs has been reported (3). Some authors consider EPDS and EPD of the legs to represent a single disease (7).

Laboratory and microscopic findings

Laboratory findings in EPDS are usually unremarkable, apart from increased acute-phase reactant level (CRP). Zinc deficiency has sometimes been reported and should be investigated at time of diagnosis, at least. Local bacterial and mycological swabs are usually negative, except in the case of secondary colonization with *Staphylococcus aureus* or *Candida albicans*. Pustule swabs should therefore be repeated if necessary (e.g. in the case of large pustules, recurrence, or treatment resistance).

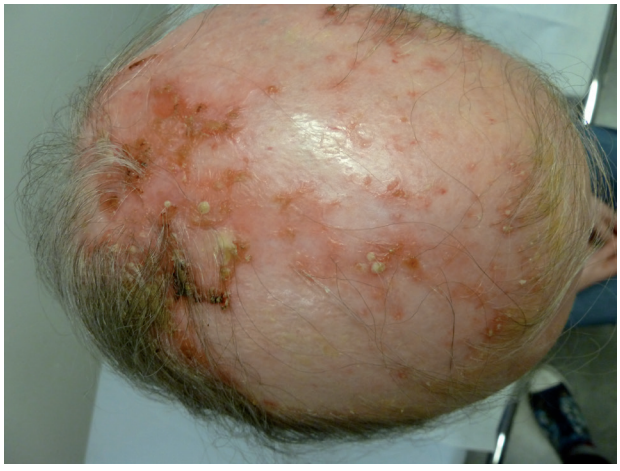

Fig. 1. Cicatricial alopecia of the vertex with erosions, crusts, and yellow pustules in an elderly patient.

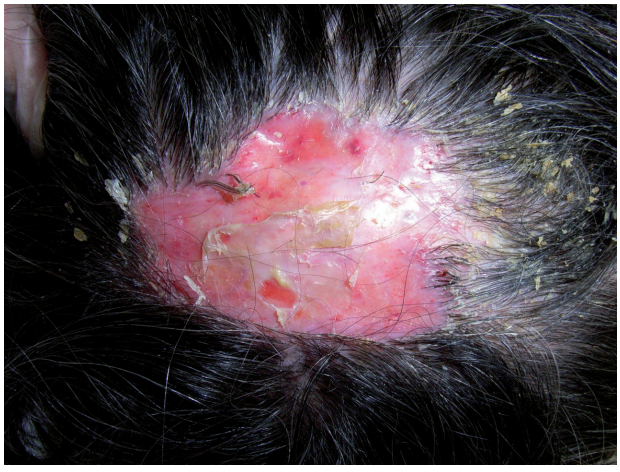

Fig. 2. Localized form of erosive pustular dermatosis of the scalp.

Trichoscopy of bald areas of the scalp will show marked skin atrophy with enlarged blood vessels of the dermis, lack of follicular ostia and reduced number of hairs. Serous or black follicular crusts are seen (3).

Microscopic findings are non-specific and vary according to the type of lesions sampled (e.g. pustule, erosion, scar) and the disease duration. At an early stage histology shows chronic inflammation of the upper dermis, with a focal or diffuse mixed infiltrate composed of lymphocytes, neutrophils and plasma cells. Hair density is normal, with an increased number of catagen follicles. At a more evolved stage, fibrosis of the dermis, absence of sebaceous glands, diminished number or complete absence of hair follicles, and atrophy of hair follicles are observed. Negative direct immunofluorescence will rule out possible bullous pemphigoid. Gram and periodic acid-Schiff (PAS) stainings will be negative as well (3).

Treatment

Treatment of EPDS has not been codified. Several treatments have been tried with varying degrees of success. Local and oral antibiotics and antiseptics are not efficient in the absence of an infectious cause. Ultrapotent local corticosteroid ointments (e.g. clobetasol propionate) are efficient. Efficacy is assessed on the lack of inflammation, the disappearance of erosions and crusts. However, dermocorticosteroids have to be applied for a long time (6 months) before efficacy is evident. Relapses occur within a year of withdrawal, and usually after a mean of 3 months. In addition, skin atrophy may be worsened by corticosteroids. The efficacy of tacrolimus 0.1% once or twice a day, calcipotriol 0.005% daily, and dapsone 5% gel twice a day (5) have been reported in anecdotal cases. Tacrolimus can be used as first- or second-line treatment in case of failure of corticosteroid treatment. The efficacy of tacrolimus is evaluated after 3 months (3). It can be used also as maintenance therapy twice a week to prevent recurrences (3). Because of the risk of squamous cell carcinoma, follow-up of the patient is recommended, especially when tacrolimus is used.

The efficacy of oral zinc has been reported in some cases, even in the absence of zinc deficiency. Treatment with an initial combination of oral zinc (zinc gluconate) and local dermocorticosteroids, followed by zinc alone, may be efficient. Oral isotretinoin may result in improvement after some months (8); however, cutaneous atrophy may be a limiting factor to the use of oral isotretinoin as a first-line treatment. Treatment with oral dapsone does not seem to be efficient according to the few reports in the literature (5). Additional advice for patients includes using sun protection, avoiding additional aggravation to the scalp (e.g. using hot hairdryers, combing, dying, perming or bleaching the hair) (3).

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