Erosive pustular dermatosis of the scalp: Clinical, trichoscopic, and histopathologic features of 20 cases

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Background: Erosive pustular dermatosis of the scalp is a chronic eruption that leads to scarring alopecia.

Objective: The clinical, dermoscopic, and histopathological features and the course of the disease in 20 patients were reviewed and compared with the reports in the literature.

Methods: Gender, age at diagnosis, age at onset, duration, topography, predisposing factors, concomitant diseases, trichoscopy, histology, treatment, and outcome were taken into consideration.

Results: The mean age was 59.4 years. Androgenetic alopecia was present in 12 patients, 6 of whom showed actinic damage. Trauma was reported in 9 patients. Four patients were affected by autoimmune disorders. The vertex was the most common location. In all 20 patients trichoscopy showed an absence of follicular ostia with skin atrophy. Histopathology revealed 3 different features, depending on the disease duration. A reduction of inflammatory signs was observed in 14 patients treated with topical steroids and in all 3 patients treated with topical tacrolimus 0.1%.

Limitations: The rarity of this disease is a limitation.

Conclusions: The relatively high number of patients allowed us to identify a better diagnostic approach, using trichoscopy, and a more effective therapeutic strategy, with high-potency steroids or tacrolimus, which should be considered as first-line treatment. (J Am Acad Dermatol 2017;76:1109-14.)

Key words: cicatricial alopecia; erosive pustular dermatosis; histology; scalp; trauma; trichoscopy.

Erosive pustular dermatosis of the scalp (EPDS) is a rare form of cicatricial alopecia first described by Burton[1] in 1977 and clinically characterized by a chronic eruption of scalp pustules, erosions, and crusts that leads to scarring alopecia and slowly progresses to involve the adjacent scalp areas. The course is typically chronic and poorly responsive to treatments.

A typical finding of EPDS is that a mechanical or chemical trauma of the scalp frequently precedes its onset[2-4] and some authors have hypothesized that the pathogenesis of this condition is an autoimmune response toward the hair follicles induced by trauma with subsequent chronic inflammation and scarring.[3] This hypothesis is strengthened by the frequent association of EPDS with other autoimmune disorders and by its responsiveness to steroids and topical anti-inflammatory drugs.[4]

Pathologically, EPDS is characterized by atrophic epidermis and chronic inflammation consisting of lymphocytes, neutrophils, and occasionally foreign body giant cells.[5]

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The objective of this article is to review the clinical, dermoscopic, and histopathological features and the course of the disease in 20 patients and compare the findings to the reports in the literature.

METHODS

The study was approved by the university ethics committee.

All medical records of patients with a diagnosis of EPDS confirmed by histopathology at the outpatient consultation of hair diseases of dermatology of the University of Bologna between January 2007 and December 2015 were reviewed. The diagnosis of EPDS was made by the clinical finding of sterile pustules, erosions, and crusted lesions of the scalp leading to skin atrophy and scarring alopecia and histopathology that excluded folliculitis decalvans and other types of cicatricial alopecia. Data about gender, age at diagnosis, age at onset, duration of the disease before diagnosis, topography, predisposing and trigger factors, concomitant diseases, biopsy results, treatment, and outcome were obtained from the patients' charts. Clinical and dermoscopic images were taken and stored using the FotoFinder Dermatoscope (Teachscreen Software, Bad Birnbach, Germany). Trichoscopy was done at high magnifications (×20-70) and in incident light, without the use of immersion oil. Skin swabs for cultures of bacteria and fungi were performed in all patients. Patients were seen every month until remission of symptoms and then every 4 months during follow-up.

CAPSULE SUMMARY

- Erosive pustular dermatosis of the scalp is often misdiagnosed.
- Erosive pustular dermatosis of the scalp presents with characteristic trichoscopic features, namely severe skin atrophy with erosions and crusts and hair bulbs visible through the epidermis, which may allow diagnosis.
- Erosive pustular dermatosis of the scalp responds very well to topical therapy with high-potency steroids or tacrolimus, which should be considered first-line treatment in this condition.

RESULTS

Clinical features

Twenty patients received a diagnosis of EPDS during the 9-year period (2007-2015), including 19 Caucasians (Fitzpatrick II to III phototypes) and 1 Indian (phototype IV), with a mean age of 59.4 years. The demographic characteristics are summarized in Supplemental Table I (available at http://www.jaad.org).

Predisposing/triggering factors included: severe androgenetic alopecia in 12 patients, 6 of whom had severe signs of actinic damage. A previous mechanical or chemical trauma was reported in 9 of the patients. One patient developed EPDS 3 months after starting the application of topical minoxidil lotion. Patch tests excluded allergic contact dermatitis to minoxidil and propylene glycol.

The median time between the triggering event and the onset of the first signs of EPDS was 6 months (range 1-8 months) except for 1 patient (case 3), who reported the onset of EPDS 3 days after child delivery, in the same scalp area where 5 years earlier she had sustained a severe accidental trauma. None of the patients had other skin or mucosal diseases, apart from a woman affected by erosive pustular dermatosis both on the scalp and on the lower limbs.

Family history for EPDS was negative in all patients. Three patients were affected by autoimmune thyroiditis and 1 patient by rheumatoid arthritis. Laboratory investigations revealed elevated values of inflammation markers in 13 patients; 1 patient had asymptomatic antinuclear antibody positivity (1:160).

Previous diagnoses included tinea capitis, treated with topical/systemic antifungals (n = 5), folliculitis decalvans (n = 1), and postherpetic skin scarring in 1 patient.

The vertex was the most common location, followed by the frontal, the parietal, and the temporal region. The hairless area had a linear shape, extending from the frontal to the vertex scalp in 6 patients and in 3 cases it followed the shape of a previous scar from a mechanical trauma; and in 1 case it coincided with an area previously affected by herpes zoster (Fig 1, *A*). A triangular shape was seen in 2 cases, whereas in the other 13 patients the area of alopecia because of EPDS was round, with a mean diameter of 3 to 4 cm. The duration of the disease ranged from 3 to 96 months (mean 53.1 months) and positively correlated with the size of the alopecia. The clinical features also differed according to the duration of the disease. Scarring alopecia with loss of follicular ostia, severe skin atrophy, and diffuse hair thinning was evident in all patients. In EPDS of recent onset (5 patients), the scalp was erythematous and covered by serous or hemorrhagic crusts (Fig 1, *A*); in long-standing cases the scalp was diffusely thinned and atrophic, and erosions and scales were located in the center of the alopecia (5 patients) or in an active area (6 patients) (Fig 2). In 4 patients the scarring area was severely thinned, with evident