Erosive pustular dermatitis of the scalp: case series

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Summary

Background. Erosive pustular dermatitis of the scalp, also known as amicrobial pustulosis of the scalp, is a rare chronic disorder of unknown etiology mainly reported in elderly individuals. It is characterized by sterile pustules, erosions and crusted lesions, which ultimately result in scarring alopecia. Chronic sun exposure and androgenic alopecia are considered risk factors, and mechanical and chemical traumas are reported to trigger the disease.

Objective. To evaluate prevalence of the disease and risk factors, pathological features and outcome in series of patients.

Methods. All medical records of patients with a diagnosis of EPDS made between January 2007 to June 2013 were reviewed. Evaluated parameters include: age of onset, gender, duration of the disease before diagnosis, topography, predisposing and trigger factors, biopsy results and treatment. All patients were monitored by global photography and videodermoscopy. Cultures for bacteria and fungi were performed in all cases.

Results. A total of 10 patients (5 males and 5 females), ageing from 8 to 95 years (mean age 50.3 years) were diagnosed with EPDS over a period of 6 years. The vertex was the most common scalp localization. Four adult men had severe AGA and 3 of them signs of chronic sun exposure, as predisposing factors. A mechanical trauma was reported as precipitating event in 5/10 (50%) of the patients. The clinical examination revealed in all cases areas of scarring alopecia with skin atrophy, associated with sero-purulent crusts and erosions. The hair were sparse or absent, and sometimes broken a few millimeters above the emergency. The scalp was severely thin, allowing visualization of the superficial vessels. Scalp videodermoscopy showed a very atrophic and thin scalp, with lack of follicular ostia, sparse hair and evident superficial blood vessels. Scalp pathology revealed similar feature in all patients: massive fibrosis of the dermis with considerable reduction of the hair follicle density and absence of sebaceous glands. A neutrophilic infiltrate was present around the hair follicles at the isthmus level. Therapy with clobetasone propionate cream once a day was effective in reducing the inflammatory signs of EPDS in 8 cases, in 4 of which we obtained complete clearing of symptoms.

Conclusion. EPDS is a rare disease that does not seem restricted to an age group, but rather to a precipitating event that involves the scalp, most commonly a mechanical trauma. The clinical history is therefore mandatory to suspect the diagnosis, which is suggested by chronic scalp erosions leading to skin atrophy and scarring.

KEY WORDS: scalp; pustulosis; pustular dermatitis.

Introduction

Erosive pustular dermatitis of the scalp (EPDS) or amicrobial pustulosis of the scalp (APS) is a rare chronic disorder which was first described in 1979 by Pye et al. (1). EPDS or APS is typical of elderly individuals, has unknown etiology, nowadays some authors consider it as a neutrophilic dermatosis and has been included in a recently proposed classification of these diseases (2). EPDS/APS together with Sweet syndrome and pyoderma gangrenosum are inflammatory skin diseases caused by the accumulation of neutrophils in the skin and, rarely, in internal organs.

EPDS is characterized by sterile pustules, erosions and crusted lesions, which ultimately result in scarring alopecia. Patients may refer itch and a sense of nuisance. It may not involve exclusively the scalp, but also the extremities (3, 4).
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Chronic sun exposure and androgenic alopecia (AGA) are considered risk factors, and mechanical and chemical traumas are reported to trigger the disease.

We report 10 patients seen over a period of 7 years at the Outpatient Consultation for Hair Diseases of the University of Bologna.

Materials and methods

All medical records of patients with a diagnosis of EPDS made between January 2007 to June 2013 were reviewed. Demographic data and the following specifics were collected: age of onset, gender, duration of the disease before diagnosis, topography, predisposing and trigger factors, biopsy results and treatment. All patients were monitored by global photography, and videodermoscopy (FotoFinderdermatoscope®, Teachscreen Software, Bad Birnbach, Germany). Cultures for bacteria and fungi were performed in order to exclude other pathologies.

Results

Data about our case series are detailed in Table 1. A total of 10 patients (5 males and 5 females), ageing from 8 to 95 years (mean age 50.3 years) were diagnosed with EPDS over a period of 6 years. Three patients were older than 60 years, 4 were middle aged, 2 were young, ageing 23 and 18 and 1 was a child aged 8 years. The duration of the disease ranged from 2 to 20 months. The vertex was the most common scalp localization of EPDS (8 patients), followed by the frontal (3 patients) and the parietal (1 patient) areas.

Four adult men had severe AGA and 3 of them signs of chronic sun exposure, as predisposing factors. A previous mechanical trauma was reported in 5/10 (50%) of the patients. In most of the cases it was an accidental trauma, which preceded the onset of EPDS from 2 months to 10 years. One patient reported Herpes zoster of the ophthalmic branch of the trigeminal nerve, and laser therapy on the scalp for postherpetic neuralgia 2 years before the onset of EPDS.

One female patient reported the onset of the disease the second day after delivery. The same patient, 5 years previously, had an important trauma on the scalp caused by the corner of an open window.

The clinical examination revealed in all cases areas of scarring alopecia with skin atrophy. The hair were sparse or absent, and sometimes broken a few millimeters above the emergency. The skin of the scalp was severely thin, allowing visualization of the superficial vessels. In 8 patients the alopeic scalp showed blood and

Table 1 - Demographic and clinical data of our patients with EPDS.

<table>
<thead>
<tr>
<th>Sex</th>
<th>Age (in years) at diagnosis</th>
<th>Age (in years) at onset</th>
<th>Predisposing/triggering factors (latency before EPDS appearance)</th>
<th>Treatment that induced remission</th>
<th>Recurrences after treatment withdrawal</th>
</tr>
</thead>
<tbody>
<tr>
<td>M</td>
<td>89</td>
<td>88</td>
<td>Androgenetic alopecia (Hamilton V), Chronic sun exposure</td>
<td>Clobetasone propionate cream</td>
<td>yes</td>
</tr>
<tr>
<td>M</td>
<td>25</td>
<td>23</td>
<td>Mechanical trauma (2 years before)</td>
<td>Clobetasone propionate cream</td>
<td>yes</td>
</tr>
<tr>
<td>F</td>
<td>48</td>
<td>44</td>
<td>Mechanical trauma (5 years before). Delivery (EPDS appeared 2 days after)</td>
<td>Tacrolimus 0,1% ointment</td>
<td>no</td>
</tr>
<tr>
<td>F</td>
<td>81</td>
<td>79</td>
<td>Herpes-Zoster of the ophthalmic branch of the trigeminal nerve and laser therapy on the scalp for postherpetic neuralgia (2 years before appearance)</td>
<td>Clobetasone propionate cream</td>
<td>no</td>
</tr>
<tr>
<td>F</td>
<td>51</td>
<td>49</td>
<td>Unidentified cause</td>
<td>Clobetasone propionate cream</td>
<td>yes</td>
</tr>
<tr>
<td>M</td>
<td>95</td>
<td>94</td>
<td>Androgenetic alopecia (Hamilton VII). Chronic sun exposure</td>
<td>Clobetasone propionate cream</td>
<td>no</td>
</tr>
<tr>
<td>F</td>
<td>8</td>
<td>3</td>
<td>Mechanical trauma (1 month before)</td>
<td>Clobetasone propionate cream</td>
<td>no</td>
</tr>
<tr>
<td>M</td>
<td>50</td>
<td>49</td>
<td>Androgenetic alopecia (Hamilton V). Chronic sun exposure</td>
<td>Clobetasone propionate cream</td>
<td>yes</td>
</tr>
<tr>
<td>F</td>
<td>66</td>
<td>56</td>
<td>Androgenetic alopecia (Hamilton IV). Mechanical trauma (10 years before)</td>
<td>Clobetasone propionate cream</td>
<td>yes</td>
</tr>
<tr>
<td>M</td>
<td>18</td>
<td>18</td>
<td>Mechanical trauma (2 months before)</td>
<td>Tacrolimus 0,1% ointment</td>
<td>no</td>
</tr>
</tbody>
</table>
Erosive pustular dermatitis of the scalp

sero-purulent crusts with erosions (Figure 1), while in 2 patients the alopecic skin was diffusely covered by oily scales and erosions. Scalp videodermoscopy showed a very atrophic and thin scalp, with lack of follicular ostia, sparse hair and evident superficial blood vessels. The hairs had a tortuous and coiled shape; broken hair shafts were also noticed, often embedded in a yellow exudate. The thin skin often allowed visualization of the hair follicle bulbs (Figure 2).

Bacterial and fungal cultures were always negative. Scalp pathology revealed similar feature in all patients: massive fibrosis of the dermis with considerable reduction of the hair follicle density and absence of sebaceous glands. A neutrophilic infiltrate was present around the hair follicles at the isthmus level (Figure 3).

Treatment included application of clobetasone propionate cream once a day, with gradual reduction of the days of application in 6 months, and the use of an antimicrobial shampoo. Avoiding sun exposure was also suggested, such as wearing a hat. Therapy was effective in reducing the inflammatory signs of EPDS in 8 cases, in 4 of which we obtained complete clearing of symptoms. Mean duration of treatment before evident efficacy was 6 months. We visited patients and performed videodermoscopy every 3 months until complete remission. In two cases not responsive to clobetasone propionate, we switched to tacrolimus 0.1% ointment, with mild regression of symptoms. In four cases the condition remained stable after treatment interruption. Five cases relapsed, one of them after a prolonged sun exposure.
Conclusions

The clinical features may not always be suggestive of EPDS and other dermatoses should be excluded such as pemphigous vulgaris, seborrheic pemphigous, superficial pyoderma gangrenosum, herpes zoster in immunosuppressed patients, pyodermitis, tricophytic sycosis, cicatricial bullous pemphigoid, acquired epidermolysis bullosa, folliculitis decalvans, dermatitis artefacta that may also present with scarring alopecia, skin atrophy, erosions and sero-hematic and purulent crusts and erosions.

In literature no incidence of EPDS has ever been calculated. In our center we see about 240 patients a year for hair and scalp pathologies; the calculated incidence of EPDS for 7 years of observation in our center is 0.59%. EPDS seems therefore a rare disease, but it may possibly be underrecognized.

Although the condition is reported to occur in the elderly, the cases of our series were scattered in all ages, and also included an 8-year-old child. This young girl had a mechanical trauma (a broken window during a car accident) to the scalp 3 months before the onset of the disease, and had been treated as a bacterial infection for several years before our diagnosis. The occurrence of EPDS in children has been reported mainly in newborns after perinatal scalp injury (5), but a better awareness of possible occurrence of the disease at any age can possibly lead to an increase of diagnosis of EPDS in young in the future. As a matter of fact, all cases of EPDS occurring in young and adult age in our series had elsewhere been previously diagnosed as bacterial folliculitis and folliculitis decalvans, while the elderly patients with EPDS received the right clinical diagnosis since the beginning and were sent to us for pathology or for treatment suggestion. It seems therefore possible that clinicians suspect the diagnosis of EPDS only in front of elderly persons, despite the clinical features of the conditions, which are rather monotonous. Areas of scarring alopecia with severe skin atrophy, associated with erosions and hematic or purulent crusts and erosions.

Chronic sun exposure is reported as a predisposing factor by several authors (6, 7): in our series it was reported in 3 cases. All of them were males with severe AGA: as the incidence of severe AGA is very high at that age and sun exposure it is frequent, it is difficult to assess the real role of these 2 predisposing factors. As a matter of fact, EPSD appeared in 6 of our patients with a thick head of hair that protected the scalp from UV very well. A mechanical trauma, most commonly accidental, was present in 5/10 patients, confirming its hypothetical role in the pathogenesis of the disease. It's known that the triggering role of mechanical trauma may represent a pathergy phenomenon in a neutrophilic disease which explains how lesions arise at the site of injury.

In literature we found only 3 cases of EPDS with onset after herpes-zoster (6-8). Our patient was also submitted to laser therapy for postherpetic neuralgia. As laser therapy is one reported cause of EPDS (9), we cannot distinguish in our case, which of the 2 factors triggered EPDS. We must also not forget that herpes zoster of the scalp often produces long-lasting erosions and crusts, especially in its necrotic type, and differential diagnosis cannot be easy in these cases (10). An intriguing case was reported by Marzano AV et al. of EPDS after cochlear implant surgery for profound sensorineural hearing loss (11).

EPDS is usually diagnosed on clinical bases, and a biopsy is always necessary to confirm diagnosis. Biopsy must be done in active areas, were erosions and hair are present. The histology although not always typical may show important fibrosis of the dermis with...
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the adnexal destruction. The remaining follicles are surrounded by neutrophilic inflammatory infiltrate. We believe that a quantitative and a qualitative analysis of the biopsy specimen might help the pathologist to do a correct diagnosis. The quantitative evaluation is the numerical counting of the follicles, including the number of the follicles in anagen, catagen and telogen, the number of follicles substituted by fibrotic tracts and those miniaturized. EPDS is characterized by reduced follicle counting and diffuse fibrosis. Miniaturized follicles can be present if the patient is also affected by androgenetic alopecia. As for the qualitative study, there should be looked for a lymphocyte and supplicative infiltrate, fibrosis and the absence of sebaceous glands. Dermoscopy might be useful in order to orientate the clinician to make a correct diagnosis, showing loss of follicular ostia and sever skin thinning. Extreme skin thinning with evident hair follicles bulbs is perhaps a dermoscopic finding exclusive of EPDS, and can be appreciated even with a manual dermatoscope.

EPDS has a chronic course, with slow progression. Treatments reported in the literature gave different results and include topical and systemic drugs, photodynamic therapy and surgery. Broussard et al. report 4 patients with EPDS treated successfully with topical dapsone 5% gel (12). This therapy seems to effective but is the result of a limited experience and in our opinion might be considered as a secondary choice, in case of failure to steroids, and tacrolimus.

In our experience, high potency topical steroids are useful in blocking the inflammation of EPDS and decreasing the subjective symptoms. Recurrences may occur after treatment interruption and can be triggered by traumas, as it was in 1 of our patients who had a prolonged sun exposure. Gradual decrease of frequency of steroid application and maintenance of UV protection are in our experience the best ways to obtain good results. Although squamous cell carcinoma is reported to develop in areas affected by EPDS (13), our follow-up of 7 years did not show any malignant outcome.

To conclude, EPDS is a rare disease that does not seem restricted to an age group, but rather to a precipitating event that involves the scalp, most commonly a mechanical trauma. The clinical history is therefore mandatory to suspect the diagnosis, with is suggested by chronic scalp erosions leading to skin atrophy and scarring.

Disclosure
Founding sources: none.
The Authors do not have conflicts of interest to disclose.

References