CASE REPORTS

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Erosive pustular dermatosis of the scalp – Is it really a rare condition?

Erozivna pustularna dermatoza skalpa – da li je to zaista retko stanje?

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Abstract

Introduction. Erosive pustular dermatosis of the scalp (EPDS) is a rare disorder of unknown etiology that usually occurs in the elderly and is characterised by multiple pustules, erosions and crusts that appear on the scalp leading to scarring alopecia. The histopathology and laboratory tests are not specific which is the reason that EPDS is a frequently misdiagnosed condition. **Case report.** We presented two patients with EPDS. The first patient had the known history of local trauma, both patients had chronic recidivant process, classic clinical presentations, and nonspecific histological findings. Each patient had prompt therapeutical response to potent topical steroids. **Conclusion.** The diagnosis of EPDS can be made if a condition fulfills the following criteria: atrophic or actinic damaged skin, clinical association of erosions, pustules, scales and crusts, no specific histopathology, no infectious agent found responsible for the condition, and chronic course leading to scarring alopecia, and prompt respnse to the treatment with topical steroids. The history of chemical or physical trauma is often present.

Key words:
skin diseases; scalp dermatoses; diagnosis; therapeutics; adrenal cortex hormones; treatment outcome.

Apstrakt

Uvod. Erozivna pustularna dermatoza skalpa (EPDS) je retka dermatoza nepoznate etiologije. Javlja se kod bolesnika starijeg životnog doba, a karakteriše pojavom brojnih pustula sa erozijama i krustama koje rezultuju ožiljnom alopecijom. U EPDS patohistološki i laboratorijski nalazi nisu specifični, što rezultuje čestim neprepoznavanjem ovog poremećaja. **Prikaz bolesnika.** Prikazali smo dva bolesnika sa EPDS hroničnog recidivantnog toka. Kod jednog bolesnika pojavi EPDS predhodila je trauma, a oba su imala klasičnu kliničku prezentaciju i nespecifičan histološki nalaz. Kod oba bolesnika postignut je brz terapijski odgovor na potentne topikalne kortikosteroide. **Zaključak**. Dijagnoza EPDS postavlja se ako su prisutni atrofična ili aktinički izmenjena koža, pustule, erozije, kruste i ožiljci, bez specifičnog patohistološkog nalaza i uzročnika u vidu infektivnog agensa, kao i hroničan tok koji vodi u cikatricijalnu alopeciju, uz brz terapijski odgovor na topikalne kortikosteroide. Često je prisutna istorija hemijske ili fizičke traume.

Ključne reči:
koža, bolesti; poglavina, dermatoze; dijagnoza; lečenje; kortikosteroidni hormoni; lečenje, ishod.

Introduction

Erosive pustular dermatosis (EPD) of the scalp (EPDS) is a rare disorder that mainly occurs in elderly patients often with previous trauma to the scalp [1]. It was first described in 1979 by Pye et al. [2]. Less than 50 cases have been reported since then. The first described patients were all women with involvement of the scalp. Biopsy specimens revealed marked chronic inflammatory changes with prominent plasma cells, epidermal atrophy, and, in most patients, epidermal erosion. The number of hair follicles appeared to be diminished. Patients were successfully treated with potent topical steroids. In instances when EPD has not been correctly diagnosed, the course of the process is continuous, and treatment lasts for years [3].

Case 1

A 59-year-old man with the 5-year history of erosive, pustular and crusted lesions on his scalp (Figure 1) was presented to our Clinic. The patient had previously been treated with antibiotics and sun protective cream without showing signs of improvement. A histopathological examination revealed the presence of unilocular subcorneal pustule in epidermis with flattened rete ridges. Solar degeneration of collagen fibers was present in the dermis (Figure 2). Direct immunofluorescence did not reveal deposits containing IgG, IgA, IgM, or C3 antisera. Wound swabs did not result in the isolation of pathogenic agents. The changes were treated with a potent topical corticosteroid – fluocinoloneacetonide ointment, with removal of crusts prior softened by a physio

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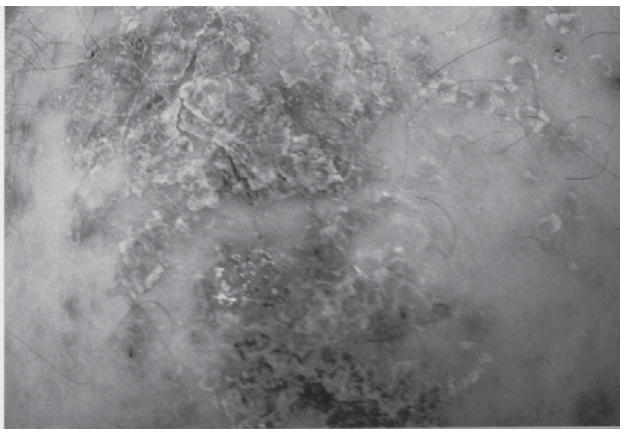

Fig. 1 – Erosive, pustular and crusted lesions on the scalp of the first patient before the treatment in our clinic.

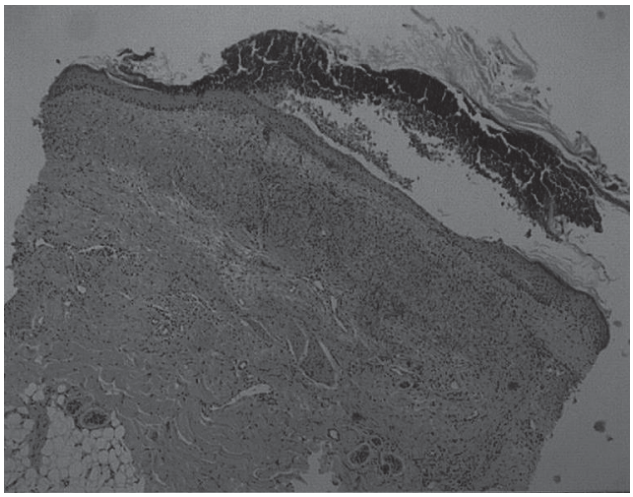

Fig. 2 – Unilocular subcorneal pustule in epidermis with flattened rete ridges; solar degeneration of collagen fibers (histopathological examination of the scalp lesions of the first patient) (HE, ×50).

logical solution and a rinse with 3% boric acid. Therapeutic response at the end of the second week of the treatment showed a 60% improvement, with the presence of rare changes: 2 pustules, 3 erosions, without crusts. At the end of the fifth week of the treatment the skin was completely epithelialized (Figure 3).

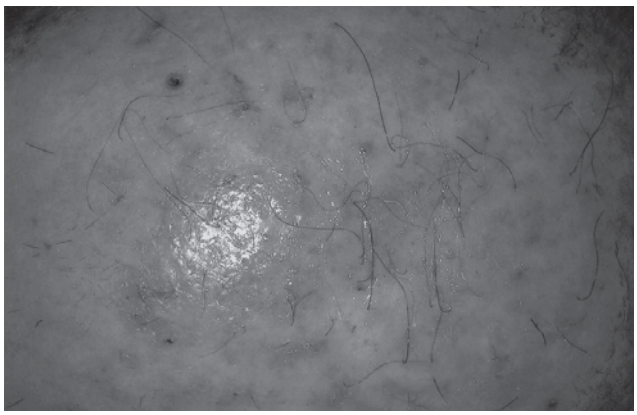

Fig. 3 – The completely epithelialized skin of the scalp of the first patient at the end of the fifth week of the treatment.

Case 2

A 87-year-old woman was presented to our Clinic because of the 5-year history of crusted lesions and inflamed erosions on her scalp (Figure 4).

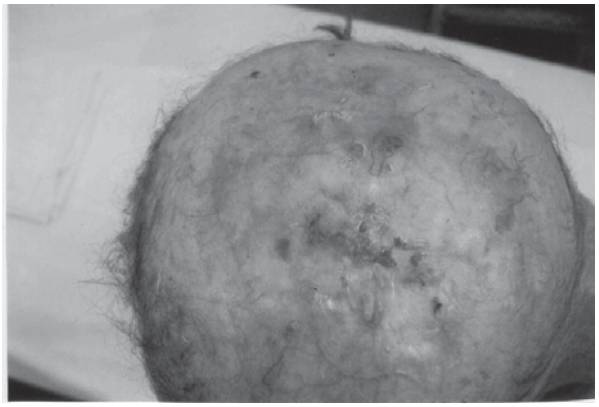

Fig. 4 – Crusted lesions and inflamed skin erosions on the scalp of the second patient (the first examination in our clinic).

Nine months after the onset of the condition, the patient underwent a radical excision procedure for a suspect tumour of the vertex of the scalp, and a split-thickness skin graft 70 mm in diametar was placed. Histopathological finding was as follows: pemphigoid dermatitis, with no tumour cells present. The acceptance of graft was pour with pustules, erosions and crusts occurring at the site of graft and the rest of the scalp. There was no necrosis of the graft. Excisional biopsy was taken by plastic surgeon with histology reporting chronic cutaneous ulcus. The patient was treated with an hydrocolloid wound compress with ionic silver after rinsing with a H_20_2 3% solution, after which occurred a temporary improvement, then a relapse without any provoking factors.

A subsequent skin biopsy from a temporal scalp region revealed hyperkeratosis, a flattened rete ridges, atrophy of the epidermis, and ulceration in the reticular dermis, and reduced appendages. Dense inflammatory infiltrate, numerous plasma cells, and histiocytes were found on the edges of ulceration (Figure 5). Direct immunofluorescence did not re-

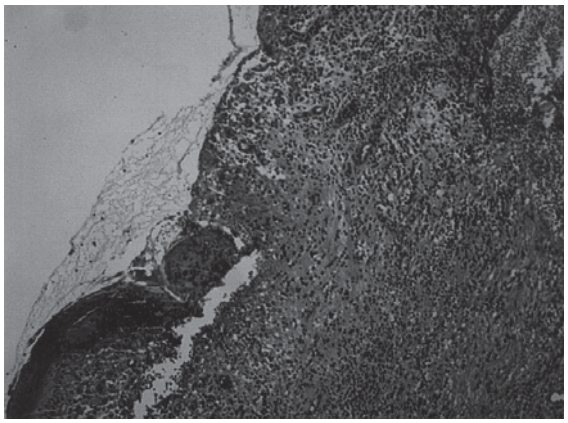

Fig. 5 – The dense inflammatory infiltrate, numerous plasma cells, and histocytes are found on the edges of ulceration (HE, ×100).

veal deposits containing IgG, IgA, IgM, or C3 antisera. Treatment with a potent topical corticosteroid ointment (betamethasone began), with removal of crusts prior softened by a physiological solution and a rinse with 3% boric acid. Three weeks of the therapy led to a significant improvement, approximately 70%. After five weeks of the therapy, there were three erosions of the skin, which was clearly atrophic, with cicatricial alopecia (Figure 6). Therapy with a corticosteroid lotion (mometasone furoate) continued, only to the areas where new pustules were forming.

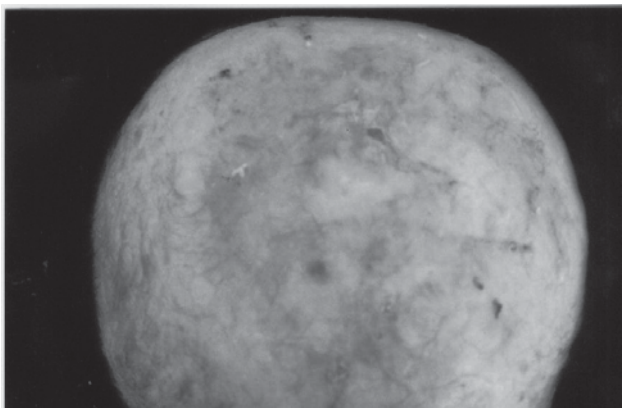

Fig. 6 – Three erosions of the skin, clearly atrophic, with cicatrial alopecia are shown after five weeks of the therapy.

Discussion

This is the first case report of EPDS in Serbia. Both patients presented to our clinic within one calendar year, with the history of unrecognised EPDS for longer than five years. Both were previously seen and treated by dermatologists for longer than 3 years under different diagnoses and with mild or no therapeutical response.

The exact cause of EPDS is unknown. Previous reports suggest that the development of EPDS is often associated with antecedent trauma to the skin and the areas of long-standing solar damage and that usually occurs in the elderly patients [4].

Actinic keratoses can sometimes be present clinically and/or histologically. Because this is the sun-damaged skin, it is not surprising that malignancy can either precede or follow EPD. For this reason, it may appear that epidermal dysplasia is causing the erosive disease. Arguments against this supposition are as follows: the treatment with cryotherapy, electrosurgery, laser or topical chemotherapy do not improve the condition and occasional development EPD in non sun-damaged skin of the legs. Conclusion is that atrophy caused by actinic damage, rather than actinic damage itself, is responsible for the development of EPD [3].

It is belived that trauma is one of the precipitating factors. A history of preceding tissue damage such as that caused by herpes zooster or iatrogenic insult: cryoteraphy, topical chemotherapy, laser therapy, electrosurgery, excisional surgery, grafting, is often found [5].

Hormones might have some influence in the pathophysiology of EPDS. It has been documented that it develops with a female predominance of about 2:1. There is evidence that in postmenopausal women estrogen loss is accompanied by many skin changes including atrophy and delayed wound healing, which might lead to the development of EPDS [3].

Chronic nonspecific inflammation (presence of plasma cells and lymphocites) is present in biopsy specimens. This observation suggests that inflammation plays an important role in the pathophysiology of the disease. Neutrophils are almost always present. It suggests that EPD should be classified as part of neutrophilic dermatoses. However, neutrophils do not predominate in the same way they do for other conditions classified as the neutrophilic dermatoses [3].

Cultures most often reveal only normal flora. Microorganisms found in EPDS represent a secondary colonization rather than primary infection. Even when *Staphylococcus aureus* is indentified, treatment with appropriate antibiotics leads to little or no improvement [6]. Crusted plaques on the scalp with accompanying hair loss suggest fungal infection of the kerion type. However, cultures and special stains on biopsy specimen rarely reveal the expected dermatophytes and empirical antifungal treatment fails to resolve the lesion [7].

The histopathology is not specific [8], with a spectrum of inflammatory changes involving epidermis and dermis, ulceration, atrophy or hyperkeratosis and reduced number or absence of hair follicles [9]. Occasionally, when neutrophils are numerous, small subcorneal, or mid-epidermal, spongiform pustules may be present [3]. In the biopsy specimens taken from the edge of erosions, epidermis is generally present, usually atrophic, and epidermal dysplasia or even frank actinic keratoses may be present. The dermis is often thinned but it is usually intact as would be expected for a condition that is more erosive than it is ulcerative [3]. Squamous cell carcinoma arising on the scars has also been reported [8].

Clinical hallmark of EPD is the presence of inflamed erosion. Most patients present with thick, yellow, or yellow-brown crusts covering one or more shallow inflamed erosions. Pustules have been reported to be present in many patients, they are usually flattened and contain little fluid. The skin around erosions is almost always atrophic. There is no swelling, warmth and regional lymphadenopathy. Pain or pruritus may be present [3].

EPDS treatment approaches include potent topical corticosteroids and oral isotretinoin; oral zinc sulfate or aspartate have been used, also. The best results have been achieved with topical corticosteroids [8]. The calcineurin inhibitors such as tacrolimus might have theoretic advantage over topical steroids, however, this advantage should be balanced by the possibility that these have oncogenic potential when used on chronically sun-exposed skin [3]. Topical tacrolimus is a potent anti-inflammatory and immunosuppressive molecule, which has been shown to be effective in the management of chronic inflammatory skin disease [9].

Two types of EPD have been discussed in the literature: EPD of the scalp and EPD of the lower legs. The clinical appearance, the histology, and the response to the therapy of lesions in these two sites are essentially identical but the settings in which they occur differ. Scalp lesions develop in the presence of atrophy secondary to actinic damage whereas the leg lesions occur only on non sun-damaged skin of patients with venous stasis disease. Controversy is whether the EPD at these two sites represent a single process or whether they are similar but separate, distinct conditions. It is belived that the unifying factor is predilection for atrophic skin of any cause to develop nonhealing, or slowly healing, shallow inflamed erosions [3].

The differential diagnoses of EPDS include pyoderma gangrenosum, pemphigus, cicatricial pemphigoid, bacterial and fungal infections, discoid lupus erythematosus, pustular psoriasis, subcorneal pustular dermatosis, infected eczema, epidermoid carcinoma, folliculitis decalvans, and lichen planus [10].

Conclusion

EPDS is not as rare condition in dermatology settings as speculated, but often goes unrecognized or misdiagnosed worldwide. These case reports are the first reports of EPDS in Serbia. EPDS is the diagnosis of exclusion, however it is the most likely diagnosis in cases of chronic shallow erosions that do not heal easily, in the background of actinic damaged skin or atrophic skin of the scalp in the elderly, with a prompt therapeutic response to the application of potent topical corticosteroids. These criteria, despite non-specific laboratory and pathological findings, are sufficient for diagnosis.

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