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CASE REPORT

Erosive pustular dermatosis of the scalp – a case report

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Summary

Introduction: Erosive pustular dermatosis of the scalp (EPDS) is rare inflammatory dermatosis of unknown etiology, which mainly affects the scalp. The condition is characterized by sterile pustules, often with secondary bacterial colonization, erosions and crusts which lead to scarring alopecia. EPDS occurs in the elderly, more frequently in women.

Patient report: A 55-year-old Caucasian female presented with a large atrophic erythematous plaque on the scalp, coated with pustules on the sides and covered with thick crusts and erosions. The patient reported a history of painless, moderately pruritic, enlarging lesions during three months prior to the admission to our department. She had no history of trauma, excessive sun exposure or some medical or cosmetic treatment (topical and systemic). Serological and immunologic tests (Hepatitis B and C, HIV; antinuclear antibodies, ANA) were negative. Fungal cultures were negative. Bacterial cultures grew *Staphylococcus aureus*. A scalp biopsy revealed polymorphous inflammatory infiltrate in the dermis of neutrophils, lymphocytes and plasma cells. Periodic acid Schiff (PAS) stain was negative. Direct immunofluorescence test was negative. Treatment with an oral antibiotic (rimfapicine) and potent topical corticosteroids led to marked and quick improvement but with remaining scarring alopecia.

Conclusion: EPDS is a diagnosis of exclusion, based on clinical presentation, disease course and histopathological findings**.** It is necessary to exclude other differential diagnoses - autoimmune bullous disorders, malignancies, neutrophilic dermatoses, bacterial and fungal infections**.** A prompt diagnosis and treatment will reduce scarring.

Key words: erosive pustular dermatosis, trauma, diagnosis, treatment

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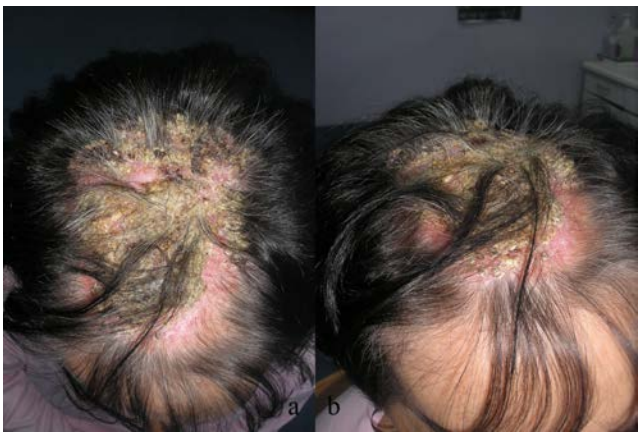
INTRODUCTION

Erosive pustular dermatosis of the scalp (EPDS) is a rare inflammatory disease, with a slow onset (1). EPDS has been reported after local injuries as well as some medication treatments (1). The disease tends to develop on sun-damaged skin of the scalp, in the elderly, more frequently in women (1, 2). Pathogenesis is not well known, predisposing factors (skin atrophy, androgenetic alopecia) and triggering factors such as trauma or damage are possibly involved (3, 4). EPDS is a diagnosis of exclusion (2, 4). The condition is characterized by sterile pustules that often become secondarily colonized, erosions or superficial ulcerations and crusts (1, 4, 5). The choice of treatment depends on age, severity and the extent of the disease.

CASE REPORT

A 55-year-old healthy Caucasian female presented with a large atrophic erythematous plaque on the scalp, coated with pustules on the sides and covered with thick crusts and erosions **(Figures 1a and b)**.



Figures 1a and b. Clinical features of a patient showing erythematous plaque covered with hyperkeratotic crusts, erosions and pustules

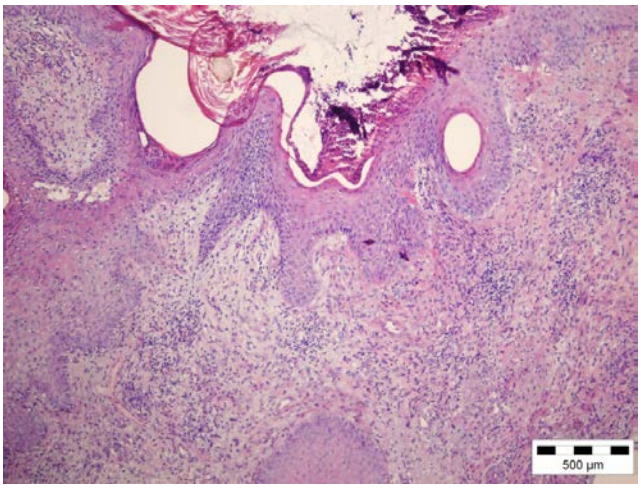


Figure 2. Histopathological finding (hematoxylin-eosin stain; original magnification x 200)

The patient reported a three-month history of painless, moderately itching, enlarging lesions. She had no history of trauma, sun exposure or a cosmetic treatment. She was initially treated with oral antihistamines and antibiotics with no or minimal improvement. Except for an elevated erythrocyte sedimentation rate (40 mm/h) and C- reactive protein (5.9 mg/L), all other tests were within the normal range. Serological and immunologic tests were negative. Fungal stains were sterile. Bacterial cultures grew *Staphylococcus aureus*. A scalp biopsy revealed polymorphous inflammatory infiltrate in the dermis consisting of neutrophils, lymphocytes and plasma cells **(Figure 2)**.

Direct immunofluorescence test was negative. Treatment with oral antibiotics (Rimfapicine, 600 mg daily, during 2 weeks) and potent topical corticosteroids led to a marked and quick improvement **(Figures 3a and b)**

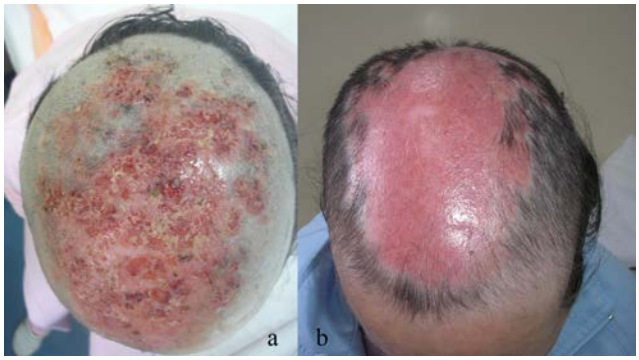


Figure 3a and b. Clinical presentation after 2 weeks of therapy with remaining scarring alopecia (Figures 4a and b).

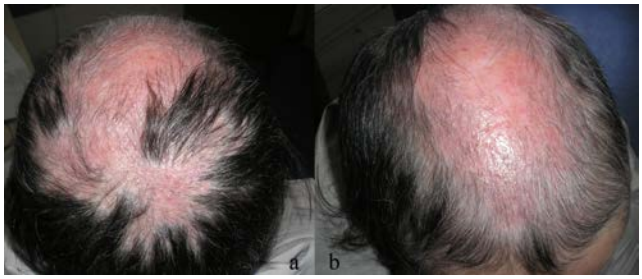


Figure 4a and b. Complete clinical regression of skin lesions with remaining scarring alopecia

In the follow-up period of 18 months she experienced only one mild recurrence and was treated shortly with topical steroids. The patient was later lost to follow-up.

DISCUSSION

EPDS predominantly occurs on the scalp, mostly on the vertex and the crown, but other sites may also be affected, including the extremities (5). EPDS is usually characterized by the association of pustules, erosions, and serous, yellow- brownish or hemorrhagic crusts on atrophic skin (6). The clinical course is protracted, with intermittent improvement and recurrences, resulting in scar formation.

The etiology and pathogenesis of EPDS is still not clear. Some factors are assumed to lead to the development of the disease. Predisposing factors include areas of actinic damage, skin atrophy and androgenetic alopecia (4). It is postulated that the immune mediated reaction after skin trauma resulting in secondary inflammation and impaired microcirculation may play a role in etiopathogenesis (6). EPDS is mostly a disease of the elderly, so a term called 'immunosenescence' (a decrease in the specificity and the efficacy of the immune response that develops with age) has been implicated (4). Aberrant immune response-loosing tolerance to self-antigens, leading to increased "self-reactivity", along with other predisposing factors may account for the development of EPDS (4, 7). Although similar to bacterial or fungal infection, infectious etiology is not thought to play a role in inflammation in EPDS, as cultures often demonstrate only occasional colonization (6, 8). However, *Herpes zoster* is reported as a possible trigger for EPDS (9). According to literature, local trauma, burns, contact dermatitis, laser treatment, cryotherapy, topical photodynamic therapy (PDT), radiotherapy, as well as some medications, cosmetic or surgical treatments are mentioned as provoking factors (2, 3, 10). The recurrence of EPDS following trauma such as skin graft, has been described in the literature indicating that removing affected skin did not control the disease (11). The presence of neutrophils in the inflammatory infiltrate is probably the consequence of skin trauma triggering autoimmune reaction resulting in a secondary inflammation (6). Recently, systemic medications such as epidermal growth factor receptor (EGFR) inhibitors have been reported as triggers (12). Most patients, however, present with a history of spontaneous onset of one or multiple confluent skin lesions (1) which was also the case in our patient.

Establishing an EPDS diagnosis can be challenging because numerous other diseases, such as primary scarring alopecia, skin malignancies, bacterial and fungal infections, inflammatory conditions (autoimmune blistering disorders in particular) may clinically mimic EPDS (13,14). Biopsy and clinicopathologic correlation are required to distinguish between EPDS and aforementioned entities. Although histopathology of EPDS is nonspecific, it is mandatory for the diagnosis to differentiate ESPD from other clinically similar diseases. The fact that chronic nonspecific inflammation is present in skin biopsies indicates that the inflammation plays a significant role in the pathophysiology of this disease (15). Considering that neutrophils are almost always present in the histopathological findings, it suggests that EPDS could or should be classified as part of neutrophilic dermatosis, but they are not predominant in the same way as they are in other conditions (5, 15).

Trichoscopy and dermoscopic examination can be useful in the diagnosis of EDSP and excluding other disorders, especially non-melanoma skin cancer (basocellular and squamocellular carcinoma) (3, 6, 13, 14). We did not use these diagnostic tools in our case.

The treatment depends on age, severity and the extent of the disease. Local treatment is given in early stages of the disease. According to literature, the best results have been obtained with high potent topical steroids, as the first line of treatment for EPDS (1, 3, 14). Besides topical steroids, topical tacrolimus and calcipotriol cream has been suggested as alternate treatments for EPDS (2, 7, 16). Although PDT has been reported as a successful treatment option it should be considered with particular attention as data also suggest PDT is a triggering factor for EPDS (2). Other treatment alternatives include oral retinoids, oral and topical dapsone and, in more severe cases, oral steroids (18, 19). Systemic antibiotics are also used to treat bacterial superinfection that occurs frequently in patients with EPDS, which was the case in our patient.

CONCLUSION

EPDS is a diagnosis of exclusion based on clinical presentation and course. The disease is likely underreported. The nonspecific histopathological pattern, the evolution leading to scarring alopecia, and the frequent response to topical steroids, favor the diagnosis. Once the other causes of the inflammatory process have been ruled out, the diagnosis of EPDS must be seriously considered. Given the lack of a large comprehensive case series and the unknown etiology, there is no general treatment recommendation for EPDS. Avoidance of precipitating factors and understanding the risk of development of actinic damage is very important.

Conflicts of interest: None to declare.

Author contribution: All listed authors contributed equally to the conception of the work, the interpretation of data, preparation of the draft of the manuscript and the interpretation of the revised version.

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EROZIVNA PUSTULARNA DERMATOZA SKALPA – PRIKAZ SLUČAJA

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Sažetak

Uvod: Erozivna pustularna dermatoza skalpa (EPDS) je retka, inflamatorna dermatoza, nepoznate etiologije, koja se uglavnom javlja u kapilicijumu. Karakteriše je pojava sterilnih pustula, često sa sekundarnom kolonizacijom, erozijama i krustama, što dovodi do ožiljne alopecije. EPDS se javlja kod starijih osoba, češće kod žena.

Prikaz slučaja: Predstavljamo slučaj pacijentkinje uzrasta 55 godina, sa velikim atrofičnim eritematoznim plakom na kapilicijumu, prekrivenim brojnim pustulama, erozijama i debelim krustama. Anamnestički, tokom tri meseca pre prijema dolazi do uvećanja promena, koje su bile bezbolne, praćene umerenim svrabom. Negirala je postojanje prethodne traume, intenzivnije izlaganje suncu, upotrebu medicinskih ili kozmetičkih sredstava (topikalnih i sistemskih). Serološki (Hepatitis B i C, HIV) i imunološki testovi (antinuklearna antitela, ANA) su bili negativni. Mikološke kulture su bile sterilne. U bakteriološkim kulturama izolovan je *Staphylococcus aureus*. U histopatološkom preparatu promene dobijene biopsijom, nađen je polimorfni zapaljenski infiltrat u dermu, sačinjen od neutrofila, limfocita i plazma ćelija. Bojenje po PAS-u (eng. *periodic acid Schiff,* PAS) je bilo negativno. Direktni imunofluorescentni test je bio negativan. Terapija oralnim antibiotikom (rimfapicin) i topikalnim potentnim kortikosteroidima dovela je do značajnog i brzog poboljšanja, ali sa posledičnom ožiljnom alopecijom.

Zaključak: EPDS je dijagnoza *per exclusionem*, na osnovu kliničke prezentacije, toka bolesti i histopatološkog nalaza. Potrebno je isključiti druge bolesti - autoimunske bulozne dermatoze, malignitete, neutrofilne dermatoze, bakterijske i gljivične infekcije. Brzo postavljanje dijagnoze i lečenje smanjuju stepen ožiljavanja.

Ključne reči: erozivna pustularna dermatoza, trauma, dijagnoza, lečenje

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