82-Year-Old Female with Crusted, Eroded Plaques the Central Scalp

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Abstract

Erosive pustular dermatosis (EPD), as its name suggests, is characterized by pustules and erosions clustered together. The most common distributions are localized to either the scalp or the legs. Most individuals are older, although rare cases have been reported in children. Diagnosis may be delayed due to overlap with actinic damage and infection. Diagnosis is generally made through a combination of biopsy, treatment failure, and therapy response. Histopathology may show a neutrophilic pustular inflammatory process, leading many to categorize EPD as a neutrophilic dermatosis. The etiology is still unknown, but trauma of any means tends to be a common preceding factor. Treatment is most commonly with topical steroids. Many other proposed treatments are mentioned in the literature. Prognosis is generally good, and many patients achieve long-term resolution of EPD.

Keywords

Erosive pustular dermatosis · EPDS · Cicatricial alopecia · Corticosteroids · Photodynamic therapy · Tacrolimus

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An 82-year-old female presented with scattered eroded plaques and pustules on the scalp for 2 months. She reported hitting her head on a car door a week around the time the eruption started. Her history was significant for actinic keratoses on the scalp and a squamous cell carcinoma on the left frontal scalp that was treated successfully with Mohs surgery.

On physical examination, there were green to yellow crusted plaques that revealed red, friable skin underneath the central scalp (Fig. 16.1). The rest of the scalp exhibited actinically damaged skin. The eyebrows and eyelashes were of normal density. The fingernails were normal. A skin biopsy showed thinning of the epidermis with partial erosion and necrosis. There was a mixed dermal infiltrate of lymphocytes, neutrophils, and a few plasma cells. A reduction of hair follicles was present. Direct immunofluorescence was negative.

Based on the clinical case description, what is the most likely diagnosis?

1. Mucous membrane pemphigoid
2. Dissecting cellulitis
3. Erosive pustular dermatosis
4. Tinea capitis

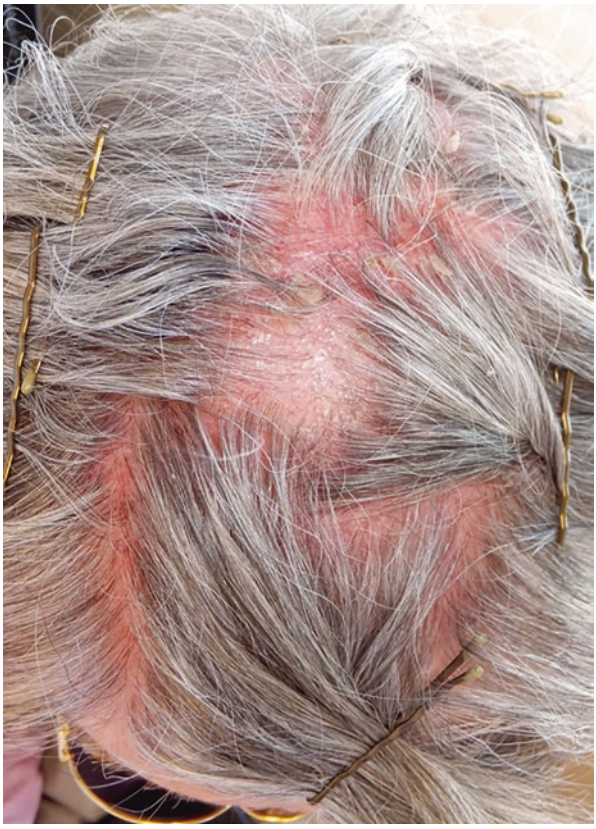

Fig. 16.1 Erythematous, friable skin present on the central scalp. Green-yellow crusted plaques are also present. Image courtesy of Dr. Melissa Piliang and Janine Sot, MBA medical photographer

Diagnosis

Erosive pustular dermatosis.

Discussion

First described in 1979, erosive pustular dermatosis (EPD) is a condition that appears as extensive areas of erosion with numerous scattered pustules overlying the affected region [1, 2]. Other names include erosive pustulosis and erosive pustular dermatosis of the scalp (EPDS), due to it classically involving the scalp [3]. EPD is most commonly located on the scalp or the legs of older individuals. There are contradictory reports in the literature regarding the prevalence in men versus women, although the consensus is that elderly women have the highest incidence and prevalence [4, 5]. The overall incidence and prevalence of EPD is hard to estimate because the disease is most likely under/misdiagnosed and under-reported [6].

EPDS is classically associated with trauma or pathergy to the afflicted region prior to development of the lesion. The most common trauma is actinic damage on the scalp of older individuals [6]. However, there are many reports of other traumatic associations, such as inflammatory or iatrogenic sources. These include surgery, topical therapies, systemic medications, and inflammatory diseases. Interestingly, surgical studies have determined that skin grafting may have the highest association with EPDS compared to other methods of closure [7]. Photodynamic therapy has also been considered a provoking factor, while also being described as a treatment modality [8, 9]. Thus, the etiology of EPDS is still very unclear.

EPDS generally presents as a large, poorly defined erythematous to eroded plaque on the vertex of the scalp. Pustules throughout the plaque often leave behind crusting [4]. The appearance can be similar to diffuse actinic damage on the scalp. Differential diagnosis includes actinic keratoses, squamous cell carcinoma, pemphigus foliaceus, and kerion formation [10]. While there may or may not be symptoms associated with the plaque, the most common symptoms include itching and burning [1]. EPDS is generally a scarring alopecia thought to be caused by regression of the plaque over time, leading to a loss of hair follicles [11]. Dermoscopy, or more appropriately trichoscopy, shows loss of the follicular ostia, dilated vessels, and hyperkeratosis [12]. Histopathological evaluation of the plaque can vary. EPDS can present as a neutrophilic, pustular folliculitis with features of spongiosis, but the presence of plasma cells and lymphocytes may be important in making the diagnosis [4, 13]. Histopathology is key in distinguishing EPDS from other scarring alopecias and conditions. Although there may be a background of actinic damage, there will not be evidence of actinic keratoses or squamous cell carcinoma. It is important to diagnose EPDS as early as possible to avoid irreversible damage. An early biopsy is important for many types of scarring alopecias as it provides cues to diagnosis and treatment.

Treatment

Treatment of EPDS is typically potent topical steroids with or without occlusion [5]. Oftentimes, this treatment helps support the diagnosis when histopathology and clinical features alone are not enough to make the diagnosis [14]. Regrowth of hair has been reported to occur but is not common [15]. Even though steroids are considered very effective for EPDS, there are many other treatment options available. If the patient has a history of herpes zoster on the scalp or head and neck region, topical steroids may not be the best option for treatment. There have been reports of reactivation of herpes zoster involving treatment of EPDS with chronic topical steroid use [16].

Other treatment modalities, reported in various case reports, have been noted to be effective for EPDS. These include non-steroidal immunosuppressive therapies such as topical tacrolimus [5]. The use of sulfasalazine in combination with a 308-nanometer excimer laser was successful in one case [17]. Topical zinc oxide has been reported for treatment of the leg, with a recent report indicating effectiveness for the scalp [18, 19]. As mentioned earlier, photodynamic therapy (PDT) is reported both as a cause and a treatment of EPDS. Case reports have combined PDT with surgical silicone gel or the fractional 1927 nanometer Thulium laser with effectiveness [20, 21]. Tetracycline antibiotics, oral retinoids, and oral Janus kinase inhibitors are examples of systemic agents that have also been used to treat EPDS [22–24].

Due to the unclear etiology of EPDS, no treatment modality can be universally recommended for the condition. Currently, first-line therapy for patients without contraindications is potent topical steroids. No randomized controlled trials regarding EDPS have been conducted. Future therapies may involve topical tacrolimus, sulfasalazine, topical zinc oxide, and PDT. As there is currently little evidence supporting these therapies, further testing must be performed.

Key Points

- Erosive pustular dermatosis (EPD) is rare and presents as chronic, eroded plaques with an overlying crust that cause a scarring alopecia when located on the scalp.
- The cause of EPD is most often caused by preceding trauma, such as actinic damage.
- EPD of the scalp is generally treated with topical steroids but some case reports suggest other therapies such as PDT, topical tacrolimus, and sulfasalazine may be effective.

References

1. Karanfilian KM, Wassef C. Erosive pustular dermatosis of the scalp: causes and treatments. Int J Dermatol. 2021;60(1):25–32.
2. Pye RJ, Peachey RD, Burton JL. Erosive pustular dermatosis of the scalp. Br J Dermatol. 1979;100(5):559–66.
3. Marsh RL, Spohn GP, Kaffenberger JA. Erosive pustular dermatosis of the scalp. Dermatol Online J. 2020;26(8):13030/qt8ks9j8sv.
4. Saridakis S, Giesey RL, Ezaldein HH, Scott JF. Erosive pustular dermatosis of the scalp following surgical procedures: a systematic review. Dermatol Online J. 2020;26(4):13030/qt9d80k39g.
5. Nasturica IE, Busila I, Lallas A. Erosive pustular dermatosis of the scalp. JAMA Dermatol. 2019;155(6):734.
6. Tomasini C, Michelerio A. Erosive pustular dermatosis of the scalp: a neutrophilic folliculitis within the spectrum of neutrophilic dermatoses: a clinicopathologic study of 30 cases. J Am Acad Dermatol. 2019;81(2):527–33.
7. Piccolo V, Russo T, Bianco S, Ronchi A, Alfano R, Argenziano G. Erosive pustular dermatosis of the scalp: why do we miss it. Dermatology. 2019;235(5):390–5.
8. Reschke R, Grunewald S, Paasch U, Averbeck M, Simon JC, Wetzig T. Erosive pustular dermatosis of the scalp: clinicopathological correlation leading to a definition of diagnostic criteria. Wounds. 2021;33(6):143–6.
9. Madray VM, Kent SM, Davis LS. Aminolevulinic acid photodynamic therapy-induced erosive pustular dermatosis of the scalp. Dermatol Surg. 2021;47(8):1140–2.
10. Light JG, Bomar L, McMichael A. Erosive pustular dermatosis in a patient with lichen planopilaris treated with aminolevulinic acid photodynamic therapy. Photodiagn Photodyn Ther. 2021;33:102207.
11. Sechi A, Piraccini BM, Alessandrini A, Patrizi A, Tabanelli M, Sacchelli L, et al. Post-traumatic erosive dermatosis of the scalp: a hypergranulated variant. Australas J Dermatol. 2019;60(4):e322–6.
12. Magri F, Pranteda G, Federico A, Muscianese M, Pigliacelli F, D'arino A, et al. Kerion mimicking an erosive pustular dermatosis of the scalp. G Ital Dermatol Venereol. 2019;154(1):96–8.
13. Martorell M, García-García JA, Ortiz C, Pérez-Vallés A, Calabuig C, Gómez-Cabrero D, et al. Prevalence and distribution of human papillomavirus findings in swab specimens from gynaecology clinics of the east coast of Spain. Scand J Infect Dis. 2010;42(6–7):549–53.
14. di Meo N, Corneli P, Retrosi C, Conforti C, Fagotti S, Longone M, et al. Erosive pustular dermatosis of the scalp: therapy is the diagnosis. Dermatol Ther. 2019;32(6):e13128.
15. Teng C, Yu J, Taylor J, Rubin AI, Treat JR. Erosive pustular dermatosis of the scalp in an adolescent with near-total hair regrowth: case report and review of the literature. Pediatr Dermatol. 2019;36(5):697–701.
16. Moustafa F, Starace M, Ravaioli G, Piraccin BM. Recurrence of herpes zoster infection in the setting of topical immunosuppression for erosive pustular dermatosis of the scalp. Int J Trichol. 2020;12(1):29–31.
17. Seckin D, Tekin B, Güneş P, Demirçay Z. Erosive pustular dermatosis of the leg in a young girl successfully treated with sulfasalazine and 308 nm monochromatic excimer light. Photodermatol Photoimmunol Photomed. 2020;36(6):496–8.

18. Di Altobrando A, Tabanelli M. Topical zinc oxide: breaking the vicious cycle of erosive pustular dermatosis of the scalp. Int J Dermatol. [published online: August 17, 2021]. https://doi.org/10.1111/ijd.15867
19. Di Altobrando A, Patrizi A, Vara G, Merli Y, Bianchi T. Topical zinc oxide: an effective treatment option for erosive pustular dermatosis of the leg. Br J Dermatol. 2020;182(2):495–7.
20. Cunha PR, Tsoukas MM, Kroumpouzos G. Erosive pustular dermatosis of the scalp treated with aminolevulinic acid photodynamic therapy and postprocedure silicone gel. Dermatol Surg. 2019;45(5):740–3.
21. Brianti P, Paolino G, Mercuri S. Fractional 1,927 nm thulium laser plus photodynamic therapy for the treatment of erosive pustular dermatosis of the scalp. Dermatol Ther. 2020;33(2):e13246.
22. Maglie R, Quintarelli L, Caproni M, Antiga E. Impressive response of erosive pustular dermatosis of the scalp to lymecycline monotherapy. J Dtsch Dermatol Ges. 2019;17(11):1177–8.
23. Jaller JA, Borda LJ, Kirsner RS. Erosive pustular dermatosis of the leg successfully treated with an oral retinoid and a bi-layered skin substitute. J Drugs Dermatol. 2019;18(3):301–2.
24. Leung N, Eldik H, Ramirez MR, Sodha P. Oral tofacitinib treatment of erosive pustular dermatosis of the scalp. JAMA Dermatol. 2019;155(6):752–4.