A case of erosive pustular dermatosis of the scalp in a pediatric patient

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INTRODUCTION

Erosive pustular dermatosis of the scalp (EPDS) is a rare inflammatory disease with predominance in the elderly population. Clinically, the condition is characterized by sterile pustules and chronic crusted erosions. Bacteriologic and mycologic workups tend to have negative findings, and histopathologic evaluation findings are nonspecific. Affected areas undergo a continuous cycle of healing and recurrence, resulting in atrophic skin with new areas of pustules and erosions. If left untreated, these areas may progress and result in scarring alopecia or cutaneous malignancies. Although the exact etiology remains unknown, EPDS tends to occur in areas of cutaneous atrophy because of numerous etiologies such as long-standing solar damage, surgical/medical/physical trauma, or, in the case of our patient, a burn.[1] Although EPDS has been documented after skin grafts in elderly patients, to the best of our knowledge, it has yet to be described in a pediatric patient after skin graft for a burn.

Abbreviations used:

EPD:	erosive pustular dermatosis
EPDS:	erosive pustular dermatosis of the scalp
IF:	immunofluorescence
SCC:	squamous cell carcinoma
sPG:	superficial pyoderma gangrenosum

CASE REPORT

A 12-year-old boy with a medical history of a third-degree burn affecting his scalp 3 years ago presented from a burn hospital for the evaluation of a scalp rash. The rash began about 2 years after an autologous graft due to a severe burn. The rash slowly worsened over the year with associated pruritus, erythema, and a thin film developing over the affected area. Previous treatments with 5% doxepin topical solution helped the itch, but topical antibiotics and antibacterial soaps did not ameliorated the condition, thus provoking the surgery team to consider regrafting out of fear of infection. The patient had a personal history of atopic dermatitis, and his mother had a history of hidradenitis suppurativa. On physical examination, the affected area (Fig 1) encompassed approximately 70% of the patient's scalp, radiating out from the vertex scalp. The rash consisted of diffuse erythema, weeping shallow erosions with some overlying yellow-brown crusting and atrophic skin bordering the grafted area. Previous bacterial cultures were obtained by the burn hospital and were all negative. A diagnosis of EPDS was suspected, and the patient was started on topical fluocinonide 0.05% cream twice a day. The diagnosis was confirmed with clinical improvement and negative microbial laboratory values at follow-up 2 weeks later (Fig 2).

DISCUSSION

EPDS is a rare, inflammatory disease of unknown etiology that is clinically characterized by chronic inflammation and recurring sterile pustules, erosions, and crusts ultimately leading to scarring alopecia.[1,2] EPDS most commonly presents in the elderly population and appears to be associated with chronic actinic damage, trauma, medication, or surgery.[3] Its presentation in the pediatric population

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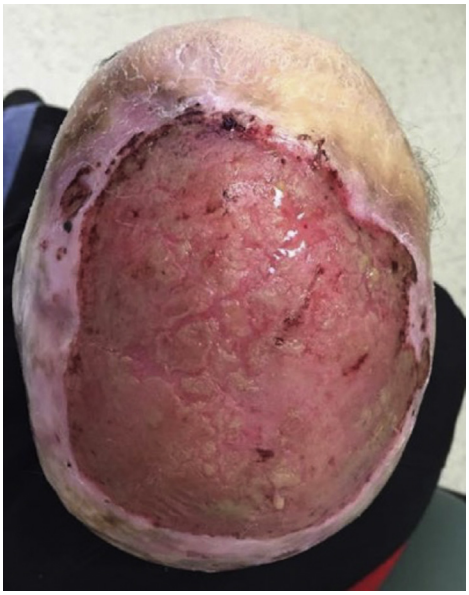

Fig 1. Erosive pustular dermatosis of the scalp. Diffuse erythema, weeping shallow erosions with some overlying yellow-brown crusting and atrophic skin bordering the grafted area.

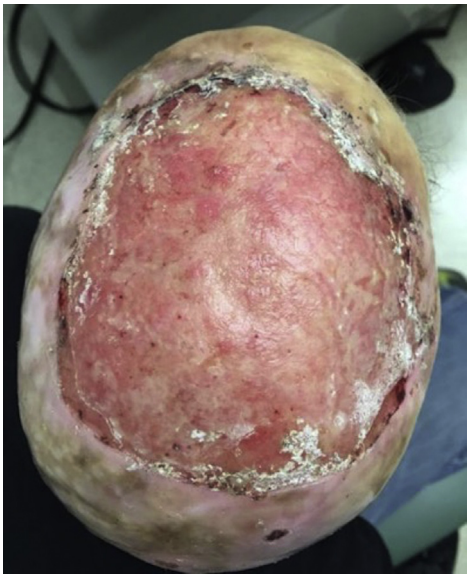

Fig 2. One-week follow-up. Significant decrease in erythema and pruritus with some focal areas of healing at the graft's edges.

has been linked to prolonged labor, caput succedaneum, and surgical trauma, among other conditions.[4,5] Although the pathogenesis is not clear, its classification is split between a chronic, inflammatory dermatosis and an immunologic dysfunction.[2] Causative agents such as local trauma (eg, burns, skin grafts, cryotherapy, radiation therapy, photodynamic therapy, hair transplantation) and topical medications (including retinoids, imiquimod, and 5-fluorouracil) damage the skin and result in acute inflammation, sterile pustules, and erosions/ulcerations.[2] After these areas of inflammation heal, the resulting atrophic skin can be reinjured, and the cycle can repeat itself.

Because of the possible misdiagnosis of EPDS with cutaneous malignancy, squamous cell carcinoma (SCC) and basal cell carcinoma should be ruled out in suspected elderly patients with EPDS and considered for all treatment-refractory patients with EPDS.[2,3] Because of the rarity of EPDS, the incidence and prevalence is unknown, thus the risk of concurrent nonmelanoma skin cancer has never been estimated.[3] Nevertheless, patients should be monitored for possible development of these or other cutaneous malignancies. For this patient, a broad differential diagnosis includes EPDS, SCC, bacterial folliculitis, kerion, other cutaneous fungal infections, atypical varicella zoster virus infection, histiocytosis X, pemphigus foliaceus, superficial pyoderma gangrenosum (sPG), and factitial dermatitis. Bacterial, viral, and fungal cultures may be helpful to identify an infectious cause. A psychology consult is helpful in evaluating for any new stressors that may suggest factitial dermatitis.

A deep biopsy can be performed with subsequent histologic and immunofluorescent (IF) examination to rule out other causes on the differential diagnosis.[6] If a biopsy was performed, nonspecific histologic findings of EPDS could have included early stages of the condition showing sterile pustules, epidermal changes including hypertrophy or atrophy, edema, and erosions. Density of hair follicles is generally normal with increased ratio of catagen to anagen follicles.[7] In addition, early findings can include a polymorphous dermal inflammatory infiltrate composed of neutrophils, lymphocytes, plasma cells, and even foreign body giant cells.[2,5] Although neutrophils, plasma cells, and giant cells are commonly seen in EPDS, these findings should not be considered pathognomonic because of their prevalence in many other conditions. Hematoxylin-eosin stain can also be used to rule out cutaneous malignancies, such as SCC and basal cell carcinoma, histiocytosis X, and neutrophilic dermatoses, such as sPG. Direct IF and special stains should be used to rule out autoimmune blistering conditions, such as PF.

There are multiple similarities between EPDS and sPG such as local trauma, or a pathergy phenomenon, as the causative agent and resolution with topical steroids. sPG may also be associated with immunosuppression and have a similar clinical presentation and histopathology as EPDS. The ways in

which these disorders may be related, especially when presenting on lower extremities, has yet to be elucidated.[3] In our patient, the lesion had well-defined, unchanged, nonundermined borders and was not painful, which helped establish the clinical diagnosis of EPDS as opposed to sPG.

Because of the rarity and unknown etiology of EPDS, there is no established treatment recommendation, but treatment is usually conservative.[2,8] Possible treatments include systemic or topical corticosteroids, topical tacrolimus, oral or topical retinoids, topical calcipotriol, photodynamic therapy, dapsone gel, silicone gel, and oral zinc sulfate.[2,3,9] Corticosteroids, tacrolimus, and wound dressings appear to be more effective and are considered first-line treatments.[3,6] High-potency topical corticosteroids are the most frequently used treatment and are established as effective and safe.[6] However, some disadvantages of topical steroids include the risk of skin atrophy and EPD relapse.

This case of EPDS demonstrates many aspects: how clinical acumen may appropriately diagnose the disease, how topical corticosteroids are an effective treatment for this condition, and how this condition may be found in skin-graft-recipient pediatric patients. For this patient, the diagnosis centered on a recent history of a traumatizing burn and autologous skin graft, lack of response to antibiotics, characteristic physical examination findings, negative bacterial and fungal cultures, and an excellent response to topical corticosteroids. A biopsy was deferred because of the patient's young age, risk of physical and mental trauma, and the fact that it would not have altered the treatment plan. If the patient had not responded to empiric treatment, then a biopsy would have been considered. The patient did not have any signs of recurrence and was scheduled for a 6-month follow-up or to present sooner if any signs of inflammation recurred. Clinicians are hesitant to make a diagnosis of a disease that lacks pathognomonic histologic and laboratory findings, which may leave EPDS frequently unrecognized and underreported.[4] This case represents a unique presentation of EPDS because it occurred in a pediatric patient after a skin graft because of a burn. Dermatologists should be aware of the possibility of EPDS developing in children who have received a skin graft.

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