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From the Dermatologikum Hamburg: Quiz

Almut Böer-Auer, M.D.[1]

[1] Dermatologikum Hamburg, Germanya

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Corresponding author: Almut Böer-Auer, M.D., Dermatologikum Hamburg, Drehbahn 1–3, 20354 Hamburg, Germany.
Tel. +49. 40. 3510750. Email: boer@dermatologikum.de.

The patient

A 78-year-old man presented with a keratotic and crusted plaque on the scalp (Figure 1). He had a history of multiple actinic keratoses with various attempts at treatment. The lesion was excised under the clinical diagnosis of squamous cell carcinoma. Photomicrographs of a section are presented in Figures 2A–S. What is your diagnosis?

Answer and explanation

Erosive pustular dermatosis of the scalp

The section pictured in Figure 2A–S shows an eroded and partly ulcerated epidermis (Figure 2G) covered by extensive scale crust housing numerous neutrophils (Figure 2S). Some neutrophils are scattered within the epidermis (Figures 2D and 2H). A diffuse infiltrate of neutrophils, lymphocytes, plasma cells, and histiocytes is present in the dermis (Figures 2I, 2A, 2Q). Fibrosis can be seen around infundibula of terminal hair follicles (Figures 2M, 2O). Out of the clinical context, these findings are not unequivocally diagnostic and bacterial infection, deep mycosis, or dissecting cellulitis of the scalp may be considered in the differential diagnosis.

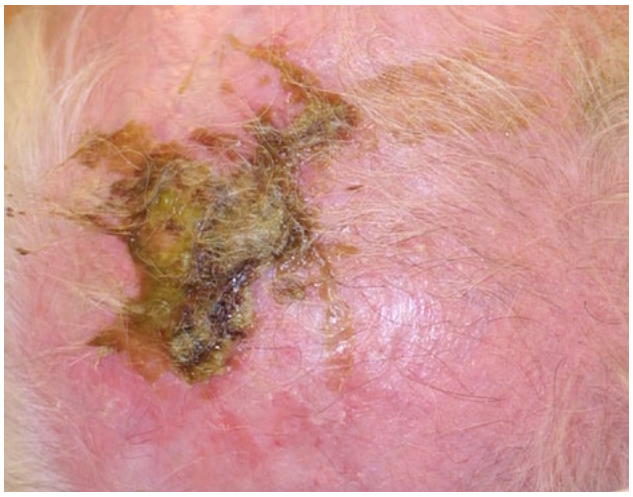

Figure 1. Clinical appearance. [Copyright: ©2012 Böer-Auer.]

In correlation with the clinical picture of a large crusted lesion on the scalp, however, the changes have to be interpreted as erosive pustular dermatosis of the scalp. In this patient, the lesions worsened after the surgical procedure (Figure 3A) and improved only with treatment with oral corticosteroids (Figure 3B).

Erosive pustular dermatosis of the scalp (EPDS) was first described by Pye and colleagues in 1979. They had observed six patients, all elderly women, who presented themselves with pustules and erosions on the scalp [1]. Lesions were not responsive to antibiotics but resolved on topical treatment with potent corticosteroids.

Since 1979, a number of case reports and a few studies on patients with similar findings appeared in the literature of dermatology and have recently been reviewed [2]. Clinical descriptions of patients diagnosed with erosive pustular dermatosis of the scalp are very similar. Most patients were women presenting with crusts, pustules, and erosions on the scalp. In some patients, alopecia was said to have been accompanying. While many reports state that patients were elderly, a few reports told of patients less than 50 years of age. Lesions were said to have been present for months or years in most patients.

Figure 2A–S. Histopathology. What is your diagnosis? [Copyright: ©2012 Böer-Auer.]

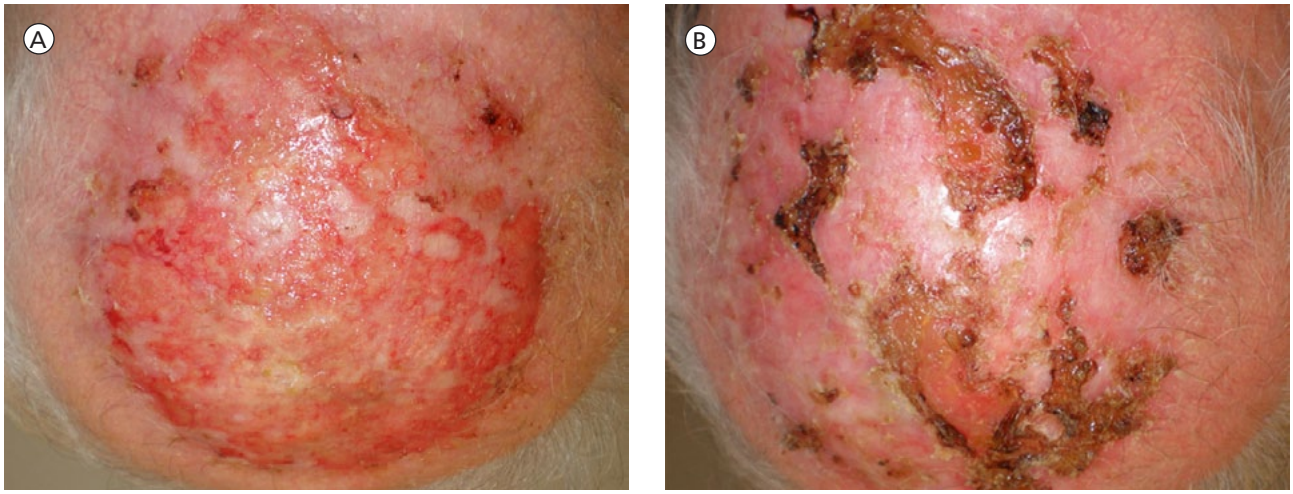

Figure 3. (A) Exacerbation after excision and transplantation; (B) Improvement on corticosteroids. [Copyright: ©2012 Böer-Auer.]

Clinically, there are similarities between lesions of erosive pustular dermatosis of the scalp and folliculitis decalvans, but in the latter condition erosion is not a typical feature. Moreover, male patients have been described with erosive pustular dermatosis of the scalp on sites affected also by androgenetic alopecia. In contrast, folliculitis decalvans is seen only on sites baring terminal hair follicles.

Histopathologic findings of erosive pustular dermatosis of the scalp are not presented in detail in most of the articles [2]. Features mentioned included "atrophy of the epidermis," "chronic inflammation," "plasma cells," and, occasionally, "polymorphous infiltration" and "spongiform pustules" [3]. Even though clinical descriptions of the condition include alopecia as a common clinical feature, involvement of follicles is mentioned rarely in descriptions of histopathologic findings. Pye et al, however, had emphasized destruction of follicles as a histopathologic finding [1]. Just as in the case presented here, patients described in the literature often presented with a quite advanced stage of the disease, and it is not clear to date what early lesions looked like, i.e., whether pustulation started as an infundibulitis or within surface epidermis.

A broad variety of events have been documented to evoke erosive pustular dermatosis of the scalp, among them being local trauma, synthetic fiber implantation, craniotomy for removal of a suprasellar meningioma, surgical excision and grafts for squamous cell carcinoma and basal cell carcinoma, cochlear implant surgery, CO2 laser vaporization, radiation therapy, cryotherapy, photodynamic therapy, topical tretinoin, topical imiquimod, topical 5-fluorouracile to treat multiple actinic keratosis, gefitinib and radiotherapy for brain metastases, and zoster ophthalmicus [2,4–6]. In the patient presented here, one or more of the various attempts to treat actinic keratoses, such as photodynamic therapy and repeated shave excisions, might have been among the initiating events. The denominator all the possible cases mentioned is that they all represent damage to the tissue followed by local inflammation.

The initiation of pustular dermatosis of the scalp by trauma, no matter of what kind, seems to suggest that the condition is mediated by a pathergic phenomenon, similar to the situation in pyoderma gangrenosum. The connection of erosive pustular dermatosis of the scalp to local trauma is a feature that differentiates it from folliculitis decalvans, a disease that develops without any previous tissue damage.

In many patients diagnosed with erosive pustular dermatosis of the scalp, response to potent topical corticosteroids was favorable [2]. Other medications that proved to be effective were other immunosuppressive agents such as acitretin, tacrolimus and calcipotriol [2,7,8]. Failure to respond to antibiotic treatment militates strongly against a bacterial cause of the condition and against similarities in pathogenesis to folliculitis decalvans and its analogues. In patients diagnosed with erosive pustular dermatosis of the scalp who responded to oral administration of zinc, a possible differential diagnosis could be zinc deficiency, which may manifest itself also with pustules and erosions on the scalp.

Even though erosive pustular dermatitis of the scalp seems to be distinctive clinically and pathogenetically, histopathologic findings of the condition are still poorly characterized. The patient presented here illustrates, however, that a sure diagnosis can be made based on clinicopathological correlation.

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