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CASE REPORT

CHRONIC RECALCITRANT DERMATOSIS: EROSIVE PUSTULAR DERMATOSIS OF THE SCALP TREATED WITH ACITRETIN AND TOPICAL TACROLIMUS

ABSTRACT

Erosive pustular dermatosis of the scalp is an uncommon disorder of the scalp, which causes chronic sterile pustules and non-healing erosions, leading to cicatricial alopecia and skin cancer. The disease usually affects older individuals. The exact pathogenesis of erosive pustular dermatosis of the scalp is unknown but microscopic examination reveals inflammation with neutrophils. As a result of limited efficacy of various medical and surgical modalities, erosive pustular dermatosis of the scalp patients present a therapeutic challenge. In this report, we describe the case of a 78-year-old male patient having chronic recalcitrant ulceration on the scalp for 4 years, which was misdiagnosed and inappropriately treated with various topical agents. The use of oral acitretin 50 mg/day and tacrolimus ointment led to a complete and permanent remission.

Key Words: Acitretin; Scalp; Scalp dermatosis; Tacrolimus? Ulcer

OLGU SUNUMU

KRONİK DİRENÇLİ DERMATOZ: ASİTRETİN VE TOPIKAL TAKROLİMUS İLE TEDAVİ EDİLMİŞ SKALPİN EROZİV PÜSTÜLER DERMATOZU

Öz

Skalpin Eroziv Püstüler Dermatozu, skatrisyel alopesi ve deri kanseri ile sonuçlanabilen kronik steril püstüllere ve iyileşmeyen erozyonlara neden olan skalpte yerleşim gösteren nadir bir hastalıktır. Hastalık genellikle yaşlı kişileri etkilemektedir. Skalpin Eroziv Püstüler Dermatozu hastalığının esas patogenezi bilinmemektedir ancak mikroskopik muayenede nötrofilik inflamasyon izlenir. Çeşitli medikal ve cerrahi tedavilerin sınırlı etkinlik göstermeleri sebebi ile, Skalpin Eroziv Püstüler Dermatozu hastalarının tedavilerinde güçlük yaşanabilmektedir. Bu çalışmada dört yıldan beri mevcut olan skalp yerleşimli, yanlış tanı almış ve çeşitli topikal ajanlarla uygun olmayan şekillerde tedavi edilmiş kronik dirençli ülserasyonu olan 78 yaşında bir erkek hastayı sunmaktayız. Oral asitretin (50 mg/gün) ve takrolimus pomad kullanımı tam ve devamlı remisyonu sağlamıştır.

Anahtar Sözcükler: Asitretin; Skalp; Skalp dermatozu; Takrolimus; Ülser

INTRODUCTION

Erosive Pustular Dermatitis of the Scalp (EPDS) is an unusual inflammatory condition characterized by chronic amicrobial pustular erosions and crusts causing scarring alopecia. It typically affects the chronically photo-damaged skin of the elderly. Its exact pathogenesis is unclear; however, local trauma, radiotherapy, laser therapy, photodynamic therapy, herpes zoster disease and application of imiquimod and 5% fluorouracil cream have been indicated as potential causes (1-4). In addition, it is associated with autoimmune conditions (5). As the disease exhibits a progressive and chronic course, it is associated with a potential risk of neoplasia and may mimic recurrent skin cancer when it occurs at surgical sites(6). Secondary colonization is possible, but the cultures of typical lesions test negative. Histopathological examinations reveal nonspecific neutrophilic inflammation. The typical history and clinical presentation are the main indicators for diagnosing EPDS. Topical and systemic steroids are the most commonly used treatment options with limited efficacy.

Here we report an elderly male with a 4-year history of moderate-to-severe disease that was misdiagnosed as a chronic non-healing ulcer, which was unresponsive to several topical treatments. The patient achieved complete remission after subsequent treatments with oral acitretin and topical tacrolimus.

CASE

A 78-year-old man had been suffering from painful erosions and plaques with dirty crusts on his scalp for 4 years. He had no history of radiation, surgical or physical trauma preceding lesion development. His family history was unremarkable. He was previously diagnosed with coronary artery disease, diabetes mellitus, hypercholesterolemia and prostate hyperplasia. He had been admitted to various clinics and had been unsuccessfully treated with topical emollients, antifungals agents and antibiotics, as indicated by a non-healing scalp ulcer. Topical steroids that were prescribed for the provisional diagnosis of pemphigus foliaceus demonstrated limited efficacy. A scalp examination revealed multiple yellowish-green hyperkeratotic crusts and oozing erosions over the alopecic skin with actinic damage (Figure 1). A systemic examination revealed normal results with no lymphadenopathy. A provisional diagnosis of EPDS was made. Two skin biopsy specimens were obtained from the affected area on the scalp for histopathological and immunofluorescence examinations. Bacterial and fungal cultures were performed. The complete blood count, urinalysis results and serologic profile, including anti-nuclear antibody, were normal. Biochemical tests revealed mild hyperglycemia. Radiological investigations revealed normal findings. A histopathological examination (Figure 2) revealed subcorneal pustule formation and significant dermal oedema with stromal mucin. Alcian blue immunohistochemical staining demonstrated slight positivity.



Figure 1. Dirty yellow thick crusts and erosions on the scalp before treatment.



A direct immunofluorescence study revealed negative results for IgA, IgG, IgM and C3 deposits on the EPDS-affected skin. A potassium hydroxide examination revealed normal results. Bacterial and fungal cultures from the scalp discharge were negative. The clinical findings and chronicity of the patient and the therapy-resistant characteristic of the lesions together with supportive histological features and negative immunofluorescence findings suggested the diagnosis of EPDS. Prior treatments with antiseptics, topical steroids (mometasone furoate 0.1% and clobetasol propionate 0.05%) and antimicrobials (mupirocin 2%) were unsatisfactory. We prescribed the application of tacrolimus ointment

0.1% twice a day, which demonstrated minimal clinical improvement after 4 weeks. As the patient had significant co-morbidities, oral acitretin 50 mg/day was initiated instead of systemic steroids. After approximately 2 months of the combination therapy of acitretin and tacrolimus, a complete clearance of active pustules, crusted ulceration with severe skin atrophy was observed. However, the patient's adherence to acitretin therapy was poor because of xerosis and intractable pruritus. Therefore, the dose was tapered to zero over the next month. Tacrolimus was used as a monotherapy twice a week for the following 2 months. No recurrence occurred after 18 months of follow-up (Figure 3).

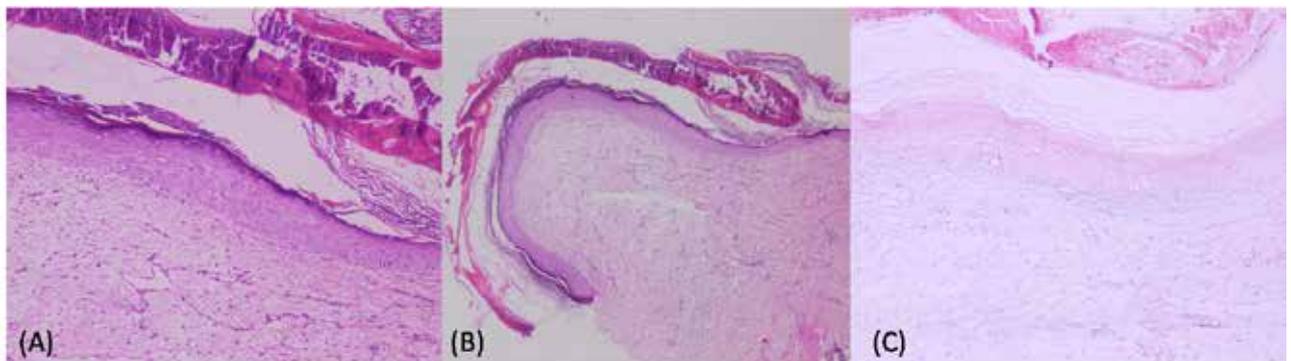


Figure 2. Subcorneal pustule formation is easily detected (Fig 2A, 2B). Dermis shows significant edema (Fig 2B) and Alcian Blue revealed faint positivity of under the epidermis (Fig 2C). H&E, Original magnification, A, C X100, B, X40



Figure 3. Resolution of the lesions after the treatment.

DISCUSSION

The typical clinical appearance on the atrophic and photo-damaged scalp of an elderly man made us suspect EPDS. Multiple differential diagnoses of this condition exist. In our patient, pustular psoriasis and subcorneal pustular dermatosis were excluded by histopathological examination. Immunofluorescence staining revealed no finding associated with immunobullous disorders, including pemphigus foliaceus, IgA pemphigus and cicatricial pemphigoid.

EPDS is generally accepted to be a rare, distinct entity, despite the publication of recent studies on it. The disease is difficult to treat. Typically, potent topical corticosteroids can only partially improve the clinical manifestations; however, recurrences are common after withdrawing the therapy. Numerous treatment options with various therapeutic benefits have been suggested (7-9). Among these, in recent years, tacrolimus has been more frequently recommended in anecdotal reports (10-12). Despite the common views asserting its carcinogenic effects with long-term use on UV-damaged skin, some authors have suggested tacrolimus as a first-line therapy for EPDS (11).

It is well known that retinoids have biological effects, including the alteration of follicular keratinization and suppression of sebaceous gland activity. Isotretinoin has been used for treating EPDS with successful results (13-15). To our knowledge, one report in the literature supports the efficacy of acitretin in treating EPDS (6). Notably, our patient exhibited satisfactory disease remission with acitretin as a systemic therapy in combination with topical tacrolimus 0.1%, without any recurrence observed during follow-up. This treatment protocol was chosen because of the prior long-term use of potent steroids and his poor medical condition.

In our patient, acitretin and topical tacrolimus proved to be highly beneficial. Regarding predisposing factors for skin cancer including sun damage and chronic ulceration, this combination seems to be preferable for compensating for the carcinogenic potential of tacrolimus in particular for fair-skinned individuals. Nonetheless, we would like to highlight that long-term, regular follow-ups are crucial, particularly for tacrolimus-treated elderly patients with photo-damaged atrophic skin.

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