Widespread Eosinophilic Pustular Folliculitis in a Nonimmunocompromised Patient

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Abstract
Objective: We present a case of eosinophilic pustular folliculitis, a rare dermatosis which is often associated with HIV infection or internal malignancies. Clinical Presentation and Intervention: We report the case of a 66-year-old man with a medical history of hypertension. Histopathological examination showed a dense follicular inflammatory infiltrate with abundant eosinophils. The clinical response to indomethacin was excellent with no recurrence during the follow-up. Conclusion: The patient responded well to indomethacin treatment.

Introduction

Eosinophilic pustular folliculitis (EPF) is a noninfectious condition characterized by an eosinophilic infiltration of the hair follicles. It was first described by Ofuji et al. [1] in 1970. The etiology of EPF is unknown. Different hypotheses have been postulated including drug hypersensitivity (i.e. to carbamazepine, minocycline or allopurinol), infections (e.g. Demodex folliculorum, dermatophytes, Pseudomonas aeruginosa and larva migrans) and immunological alterations, which produce a secretion of eosinophilic chemotactic and activation factors [2]. The probable mechanism by which eosinophils infiltrate the pilosebaceous units has not yet been established. The PGD_2/PGF_2- peroxisome proliferator-activated receptor gamma pathway may be involved in the pathogenesis. It induces eotaxin production from sebocytes, which may explain the massive eosinophil infiltrates observed around the pilosebaceous units in EPF [3].

Case Report

In early October 2012, a 66-year-old man presented with a 5-day-old itchy rash on his lower limbs, which progressively involved most of his body surface. No mucosal involvement or other systemic manifestations were reported. His medical history revealed hypertension controlled with irbesartan. Physical examination showed scaly annular erythematous-violaceous maculopapules involving the palms of his hands and soles of his feet (fig. 1). On pressure areas, the lesions showed a palpable purpuric macular morphology (fig. 2). Histopathology revealed eosinophilic exocytosis and infundibular spongiosis with eosinophilic and neutrophilic micropustules (fig. 3). In the dermis, an inflammatory infil-
trate of lymphocytes, eosinophils and neutrophils with perivasu-
lar and interstitial distribution was observed. In order to exclude a
condition of immunosuppression, a complete blood cell count was
performed which showed neutrophilia without eosinophilia. Sero-
logical tests were negative for human immunodeficiency virus,
hepatitis B and C, syphilis, antinuclear antibodies, extractable nu-
clear antibodies and tumor markers. Immunoelectrophoresis of
proteins, a systematic urine test and a peripheral blood smear were
normal. Chest computerized tomography and thoracoabdominal
magnetic nuclear resonance imaging showed no signs of adenopa-
thies or solid neoplasms. Based on both the positive and the nega-
tive findings, a diagnosis of EPF or Ofuji’s disease was made. Treat-
ment with indomethacin was initiated and this led to a rapid im-
provement and the disappearance of the lesions. The patient has
remained in good health with no signs of recurrence after 2 years
of follow-up.

Discussion

This is a case of EPF with typical lesions in an other-
wise healthy patient, where we were unable to determine
a clear etiological factor despite performing several tests
and his excellent response to indomethacin. EPF is a rare
disease characterized by the presence of itchy papules and
pustules in a circinate configuration. Three different sub-
types of EPF have been described: classic EPF, immuno-
suppression-associated EPF (mostly HIV-related or oc-
curring in patients who have received a bone marrow
transplant) and childhood EPF. Our case is one of many
of EPF in immunocompetent individuals which have
been described [4].

In classic EPF, the lesions are characterized by recur-
rent outbreaks of sterile follicular pustules and papules
forming circinate plaques with centrifugal progression
which show a predilection for seborrheic areas. Pruritus
is present in about 40–50% of cases, and leukocytosis and
eosinophilia have been reported in up to 35% [2]. In our
case, typical circinate lesions were observed, also on the
palms of his hands and the soles of his feet, even though
these areas have no hair follicles. The lesions displayed
neutrophilia without eosinophilia.

Fig. 1. Scaly annular erythematous-violaceous maculopapules.
Fig. 2. Lesions show a palpable purpuric macular morphology in the lower limbs.

Fig. 3. A histological image shows micropustules with a mixed in-
flammatory infiltrate consisting predominantly of eosinophils
with lymphocytes and neutrophils.
Immunosuppression-associated EPF displays severe itch and discrete erythematous papules with pustules. In childhood, the lesions are on the scalp [5]. In some cases, an association of EPF exacerbation with pregnancy has been found [6].

The skin biopsy of our patient showed eosinophilic exocytosis, infundibular spongiosis and eosinophilic and neutrophilic micropustules. In the dermis, an inflammatory infiltrate of lymphocytes, eosinophils and neutrophils was observed. The epidermis was intact. These histological features characterize EPF [2].

In our patient, we made a differential diagnosis by examining the characteristics of the lesions and the findings of the skin biopsy compared to other conditions. Differential diagnosis should be considered with fungal and viral folliculitis, dermatophyte infections, papular urticaria, graft-versus-host disease (in appropriate settings) and seborrheic dermatitis. Histologically, a differential diagnosis includes various conditions classified under the term 'eosinophilic dermatoses', i.e. arthropod bite reactions, drug-related skin reactions or eosinophilic cellulitis (Wells syndrome) [7].

Different options for EPF management and treatment have been described with variable outcomes [8]. The first choice includes topical corticosteroids. Topical tacrolimus has also been used successfully [9]. Indomethacin is the most effective treatment for typical EPF and it shows clinical improvement in the majority of cases including ours [10]. Our patient remained in good health after achieving resolution with indomethacin. The absence of immunosuppression probably led to a more benign clinical course. Indomethacin is an inhibitor of cyclooxygenases and a potent agonist of the PGD$_2$ receptor CRTH2. It may exert its therapeutic effect via a reduction of CRTH2 expression as well as inhibiting PGD$_2$ synthesis [10]. Other oral treatments with variable outcomes include drugs such as prednisolone, retinoids, colchicine, dapsone, itraconazole, cyclosporine or minocycline [8].

**Conclusions**

Our patient with typical EPF had a good clinical response to indomethacin therapy. This report highlights the importance of considering this entity in the workup of pustular and circinate dermatosis.

**References**