EOSINOPHILIC PUSTULAR FOLLICULITIS – CASE REPORT

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Summary

Introduction: Eosinophilic pustular folliculitis (EPF) is a benign, chronic, recurrent condition most commonly described in people of Asian descent. It is found in three clinical forms: classic, associated with immunosuppression especially with HIV infection and infantile form.

Clinical case: A 38-year-old patient is consulted for papules and pustules, with a tendency to group and form plaques with central healing and peripheral extension, slightly pruriginous located on the face. The histo-pathological examination revealed multiple follicular lesions, infundibular spongiosis, eosinophilic follicular pustules in the infundibular segmentd, dermal and perivascular perifollicular infiltrate rich in eosinophils. Eosinophilia. HIV negative reaction. Treatment was prescribed with Indomethacin 2x25 mg/day for 40 days with wound healing.

Discussions: The etiopathogenesis of EPF is unknown and may be associated with HIV infection, other infections, drugs, other conditions (eg. lymphomas, leukemia), bone marrow or organ transplantation. Clinically is characterized by sterile localized papules and pustules, which meet on the face and trunk, and currently, there are subcornous pustules with eosinophils. Treatment includes several options such as topical with dermatocorticoids and oral indomethacin.

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Introduction

Eosinophilic pustular folliculitis (EPF) is a benign condition that consists of non-infectious eosinophilic infiltrate of the hair follicle. Three clinical forms of EPF have been described: the classic form, the form associated with immunosuppression (especially HIV-related) and the infantile eosinophilic pustular folliculitis.

The etiopathogenesis of the disease remains unknown, EPF being associated with HIV infection, drugs, hematological diseases, infections or bone marrow and organ transplantation. EPF is more common among Asian population. Clinically EPF has a chronic recurrent evolution with the appearance of itchy papules and sterile follicular pustules located on the face and trunk. Histologically, the subcorneal pustule with eosinophils is highlighted. The treatment includes several options, the main one being topical with corticosteroids and indomethacin. We present the case of a patient with EPF, the classic form located on the face.

Clinical Presentation

A 38-year-old patient is consulted for papules and pustules located on the face for aproximately four months. The patient underwent treatments with oral doxycycline 100 mg/day, various anti-acne topics and dermocorticoids without any improvement. No systemic involvement is evident. The patient has no other pathological conditions and denies the administration of other drugs before the lesions appear.

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Dermatological examination reveals papules and pustules gradually become confluent located on the cheeks and temporal region bilaterally, slightly itchy (Fig. 1, 2).

Histologically findings reveals multiple follicular lesions, follicular hyperkeratosis, infundibular spongiosis, eosinophilic pustules in the infundibular segment. Perifollicular and perivascular dermal infiltrate consisting of lymphocytes, numerous eosinophils and rare neutrophils (Fig 3,4,5,6). PAS staining did not show fungal elements.

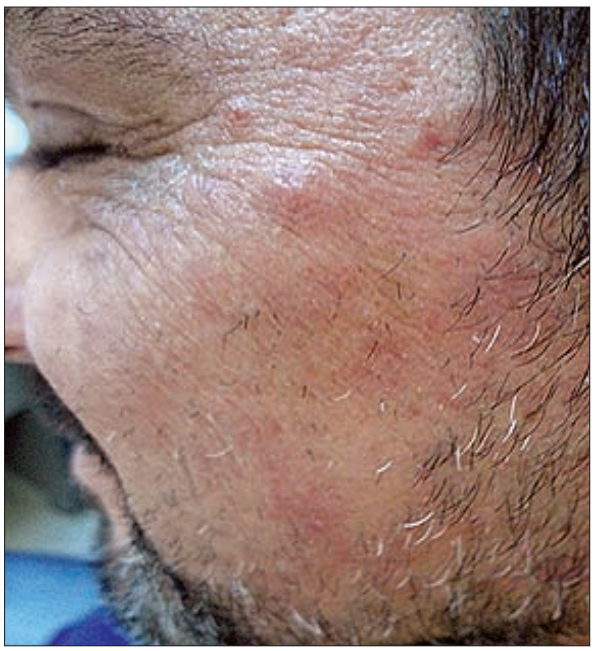

Figure 1. Papules and pustules located on the face

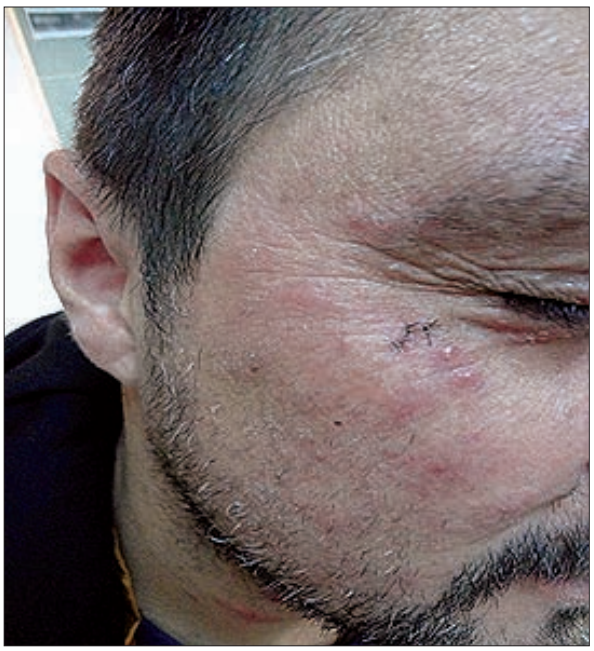

Figure 2. Papules and pustules grouped in the form of a plate

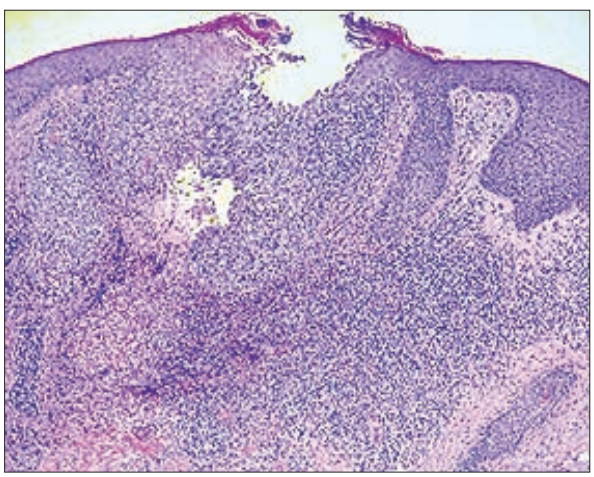

Figure 3. Histology: lesion of folliculitis and perifolliculitis with the formation of a microabscess at the level of the follicular ostium and an infundibular pustule with eosinophils (H.E 40x)

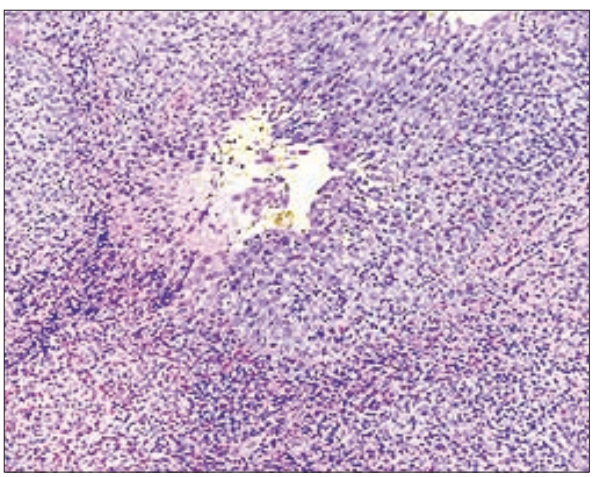

Figure 4. Histology: infundibular pustule with numerous eosinophils and neutrophils in the follicular epithelium, remnant of intrainfundibular pillar stem and abundant perifollicular infiltrates with eosinophils, neutrophils, lymphocytes and histiocytes (H.E. 100x)

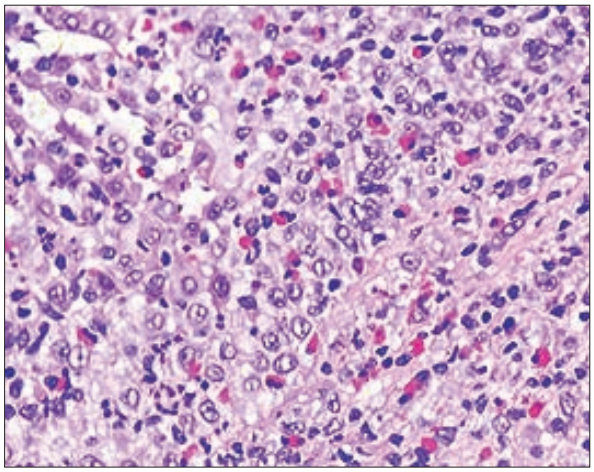

Figure 5. Histology: detail on the follicular epithelium at the level of the infundibular segment of the hair follicle, with eosinophilic and neutrophilic infiltrates (H.E. 400x)

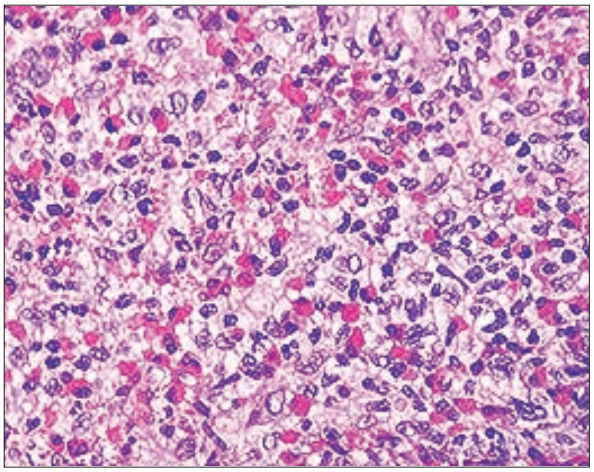

Figure 6. Histology: follicular pustule composed of numerous eosinophils and low neutrophil count (H.E. 400x)

Laboratory tests show increased eosinophilia and an HIV-negative reaction.

Treatment with Indomethacin 2x25 mg/day was administered for 40 days with the disappearance of the lesions. No recurrence of lesions was observed 3 months after discontinuation of treatment.

Discussions

Eosinophilic pustular folliculitis (EPF) is a recurrent condition of unknown etiology. In 1965, Ofuji and Ise published a case of recurrent follicular pustules and eosinophilia in a Japanese woman. Five years later after the publication of three additional cases Ofuji named this skin condition follicular pustulosis with eosinophils. [1] Other names have been proposed: sterile eosinophilic pustulosis, the classic form of folliculitis with eosinophils, Ofuji disease, eosinophilic pustular dermatosis.

Eosinophilic folliculitis is characterized by noninfectious eosinophil infiltration of the hair follicle. However, the term folliculitis has been challenged for this disorder due to the damage including palms and soles despite the absence of hair follicles in these regions. Three forms of eosinophilic pustular folliculitis have been described: the classic form of EPF, eosinophilic pustular folliculitis associated with immunosuppression, most often with HIV infection and infantile eosinophilic pustular folliculitis. A possible fourth subtype is that which occurs in adults beginning 2-3 months after hematopoietic stem cell transplantation.

Regarding the pathogenesis, the similarity between EPF and fungal folliculitis has led to speculation that EPF is due to a hyperreactivity of dermatophytes or saprophytic fungi such as Pityrosporum ovale, in association with immune system dysfunction, a theory supported by the favorable action of itraconazole in some cases. An aberrant helper T- cell type 2 immune response to a follicular antigen such as Demodex species may be responsible in patients with eosinophilic folliculitis associated with HIV infection.[3] Other authors consider that the form associated with HIV infection is due to an autoimmune disorder in which cells of the sebaceous glands or constituents of the sebum play the role of autoantigens. EPF has also been described in atopic children with hypersensitivity to Dermatophagoides pteronyssinus. [3] Another theory involved the appearance of the disease is eosinophilic chemotactic factors on the lipid surface of the skin.[4]

The etiology of EPF remains unknown, although immune processes almost certainly play a key role in the pathogenesis of the disease. EPF has been associated with HIV infection, with drugs such as carbamazepine, allopurinol,

foscarnet, capecitabine, [5,6,7,16] with other conditions such as lymphomas, [14] Sezary syndrome, [8] leukemia, myelodysplastic syndrome, atopy, cutaneous angiosarcoma, polycythemia vera.[9,11,12,13,10] EPF can also occur after bone marrow and solid organ transplants, in infections with pseudomonas, dermatophytes, cutaneous larva migrans, pityrosporum, retroviruses, hepatitis C virus [2] but also the possibility of appearance in pregnancy.

The frequency of EPF remains unknown. It is more common in Asians (eg Japanese) but also occurs in people of Hispanic descent, whites or blacks. The sex ratio of men and women is 5:1 on all three forms of EPF. In the classic form of EPF there is no sexual predilection. Instead, the form associated with HIV infection is more common in homosexual or bisexual men. EPF most frequently affects the age group 20-40 years, and in the pediatric patients aged between 5 and 10 months.

EPF the classic form has a chronic, recurrent evolution with the appearance of sterile follicular papules and pustules. In evolution, the papules tend to widen and may converge and form large polycyclic plates with central clearing and peripheral extension. Sometimes they may be acneiform. EPF is located mainly on the face (85% of cases) and trunk, and can be found on the back, on the extension surface of the upper limbs and rarely on the palms and soles. In children, the scalp and particularly the vertex can be affected. The itching is present in half of the patients and is sometimes of considerable intensity. Atypical forms of EPF such as those with follicular papules and urticarial plaques may occur in HIV-infected patients and children. EPF is a benign condition, with individual lesions healing spontaneously within months or years, usually leaving a residual postinflammatory hyperpigmentation. There is no systemic involment in EPF, but the patients with HIV-disease have a more severe and lasting course.

Patients with EPF have mild to moderate leukocytosis and eosinophilia present in half of cases. Immunoelectrophoresis shows a high level of immunoglobulin E [15] and low immunoglobulin G3, and in infantile EPF the level of immunoglobulin A is low. No bacteria or fungi developed on the cultures made from the skin pustules. In HIV-positive patients the CD4 level is less than 250-300 cells/μl.[22]

The histologically diagnosis of EPF requires multiple vertical and cross sections of the skin. In the epidermis and in the outer sheath of the hair follicle root, the subcorneal pustule with the predominance of eosinophils is evident. The structure of the hair and the sebaceous gland are infiltrated by eosinophils and a few neutrophils and mononuclear cells, and the presence of a moderately perivascular and perifollicular inflammatory infiltrate formed by eosinophils can be observed. Sometimes mucin deposits can be observed in the hair follicle or small areas of acantolysis or spongiosis with eosinophils.[21,22]

The differential diagnosis of EPF is made with acne, infantile acropustulosis, dermatitis herpetiform, erythema multiforme, toxic erythema of the newborn, impetigo, Langerhans cell histiocytosis, urticaria, atopic dermatitis, pemphigus foliaceous, perifolliculitis capitis abscedens et suffodiens, pustular psoriasis, scabies, seborrheic dermatitis, subcorneal pustular dermatosis, tinea corporis, transient neonatal pustular melanosis.

Regarding the treatment there are several options with varied results. Topics with corticosteroids represent the main treatment of EPF because of its anti-inflammatory and immunosuppressive action. The potency of the prescribed steroid depends on the location of the lesions, the applications being made twice a day, being continued even after the disappearance of the lesions. Severe rashes can be treated with short courses of Prednisone. Indomethacin (oral or topical) is another treatment option. The dose of Indomethacin administered orally to adults is 50-75 mg/day and is considered the highest cure. [17] Naproxen has also been used with some success. [18] Ointment with tacrolimus 0.03% may lead to rapid healing in children.[19.20]

Other drugs used in EPF are dapsone that can be combined with indomethacin, cyclosporine, oral or topical retinoids, itraconazole, interferon alfa-2b, antibiotics (eg doxycycline), permethrin. Narrowband, wideband UVB, UVA or PUVA phototherapy (psoralen and UVA) can also be effective. In case of pruritus, cetirizine has been shown to be most effective.[23] In HIV-infected

patients, antiviral therapy leads to a decrease or disappearance of EPF.

In conclusion, EPF is a rare condition whose diagnosis is established by correlating the clinical appearance of papules and pustules with the histological appearance of subcorneal pustule with eosinophils and infiltration of hair and sebaceous gland with eosinophils.

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Conflict of interest
NONE DECLARED

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