

Three Cases of Eosinophilic Pustular Folliculitis Without Eosinophilia

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We present three cases of eosinophilic pustular folliculitis without eosinophilia in a 19-year-old female, a 27-year-old male, and a 51-year-old male. Multiple erythematous plaques with papules and pustules were observed on the face in two patients and on the face, trunk, and extremities in the third patient. Histopathologic findings showed hair follicles and the sebaceous glands infiltrated with mixed inflammatory cells composed of eosinophils and neutrophils, characteristic of eosinophilic pustular folliculitis. However, peripheral blood eosinophilia was not present in any of our three cases. (Ann Dermatol 3:(1) 80–83, 1991)

Key Words: Eosinophilia, Eosinophilic pustular folliculitis, Ofuji's disease

REPORT OF CASES

Case 1

A 19-year-old female presented with a 5 month history of erythematous plaques with papules and pustules localized to the face.

The cutaneous eruptions began on the left cheek as crops of small erythematous follicular papules and changed to pustules in a few days. As the papulopustules increased gradually in size and number, they expanded centrifugally to form annular plaques with new eruptions predominant at the periphery. The erythematous plaques were elevated with densely packed pustules at the periphery. The erythematous plaques were elevated with densely packed pustules at the periphery and had a tendency toward central clearing (Fig. 1). The lesions resolved with residual pigmentation and new lesions appeared in the contralateral area and chin. The patient was asymptomatic except for mild itching at the beginning of the severe exacerbation. She denied seasonal variation of the lesions or exacerbation by sunlight.

Laboratory studies showed an eosinophil count of $126/\text{mm}^3$ with white blood cell count of $6,800/\text{mm}^3$.

Histopathologic examination of a biopsy from

Eosinophilic pustular folliculitis (EPF), first described by Ofuji et al in 1970¹, is a dermatosis characterized by remissions and exacerbations of erythematous plaques with follicular papules and pustules generally affecting the face, the trunk and the upper extremities. The typical histopathologic picture is that of an infiltrate of eosinophils and neutrophils within hair follicles and sebaceous glands. Eosinophilia and moderate leukocytosis in the peripheral blood is characteristic. The diagnosis is made on the basis of the distinctive clinical, histopathologic and laboratory findings.

Most cases of EPF have been reported in Japan and a few cases have been reported in Europe² and the United States. In Korea, four cases have been reported up to present^{3, 4, 5}. We report here three cases of EPF which did not show the characteristic peripheral eosinophilia.

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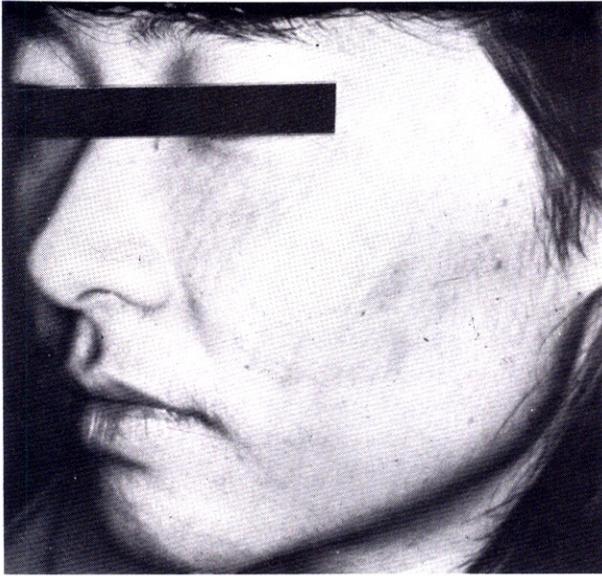


Fig. 1. (case 1). A fairly well defined indurated erythematous plaque studded with multiple papulopustules on the left cheek.

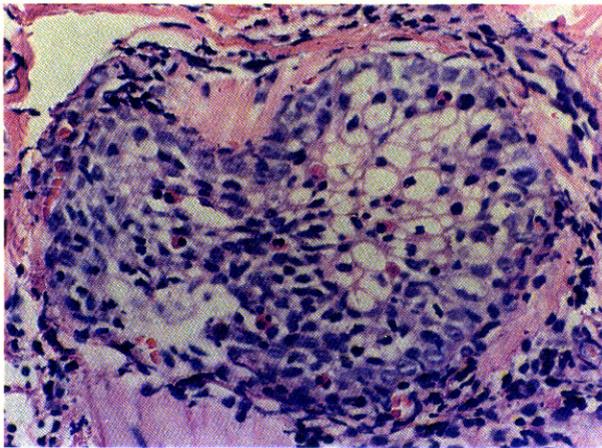


Fig. 2. (case 1). Sebaceous gland infiltrated with many eosinophils (H & E stain, $\times 200$).

the left cheek revealed sebaceous glands infiltrated with mixed inflammatory cells composed of mainly eosinophils, and a few mononuclear cells (Fig. 2). No fungal hyphae were demonstrated on PAS stained material.

Since the clinical examination and histopathologic findings were consistent with the diagnosis of EPF, antihistamines, dapsone (50mg/day) and topical corticosteroid were prescribed. The skin lesions showed some improvement, but exacer-

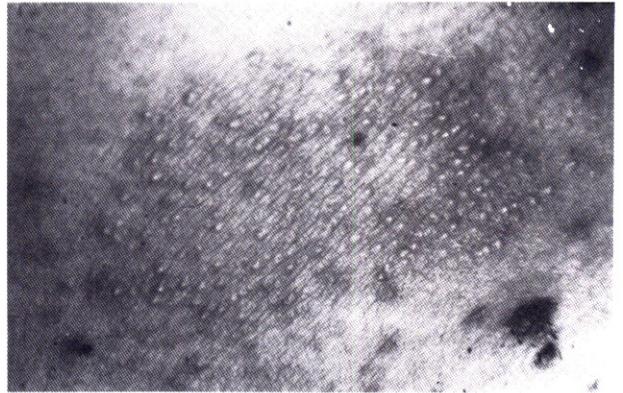


Fig. 3. (case 2). Erythematous, annular plaque with pustules on the trunk.

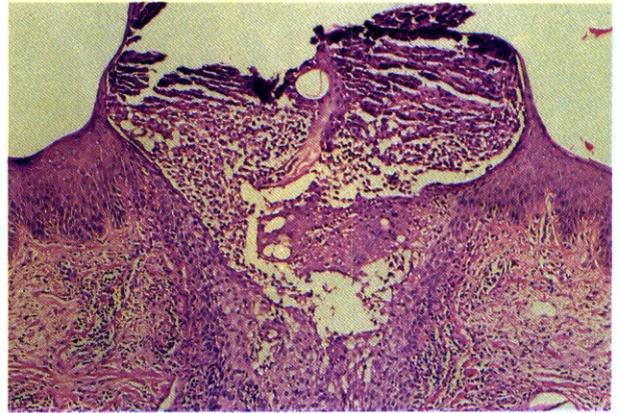


Fig. 4. (case 3). Intraepidermal abscess filled with predominantly eosinophils and a few neutrophils (H & E stain, $\times 100$).

bated after medication was discontinued.

Case 2.

A 27-year-old male presented with an 84 month history of a pruritic eruption on the face, trunk and extremities.

The grouped, reddish papulopustules were present on erythematous patches localized to the trunk and extremities. As the papulopustules spread gradually to the periphery, they became sharply demarcated erythematous plaques, which attained a diameter of 5cm in a few days (Fig. 3). The lesions resolved with residual pigmentation. Subsequently, the similar lesions developed on the both cheeks. The lesions underwent remissions and exacerbations in spite antihistamine therapy.

Laboratory studies disclosed a white blood cell count of 8,400/mm³ and an eosinophil count of 134/mm³.

The histopathology was similar to that of case 1.

There was no response to a multi drug regimen which included antihistamines, antibiotics, prednisolone (10-30mg/day), dapsone (100mg/day) and topical corticosteroid.

Case 3.

A 51-year-old male presented with a 5 month history of erythematous plaques on the face.

He had noticed pruritic, reddish follicular erythematous papules on both cheeks and eyelids. The lesions changed to pustules and became crusted over time. As the papulopustules increased gradually in size and number, they expanded peripherally to form sharply demarcated erythematous plaques with central clearing. They resolved with residual pigmentation and new papulopustules developed in the hyperpigmented areas aside from the skin lesions, the patient had been in good health and was on no medication.

Laboratory studies showed a white blood count of 8,100/mm³, with an eosinophil count of 45/mm³.

The histopathology was similar to that of case 1.

In addition an intraepidermal abscess containing predominantly eosinophils and a few neutrophils was noted (Fig. 4.).

Antihistamine, dapsone (100-150mg/day), prednisolone (5-10mg/day) and topical corticosteroids were prescribed, with little improvement in the skin lesions.

DISCUSSION

Eosinophilic pustular folliculitis (EPF) is a rare

condition. It was first described in 1965 as sub-corneal pustular dermatosis, a follicular variant? by Ise and Ofuji⁶. Subsequently, Ofuji *et al*⁷ reported three additional cases in 1970 and proposed "eosinophilic pustular folliculitis" as a new clinical entity. As the lesions were found not to be limited to the hair follicles, the terms "sterile eosinophile pustulosis" or "eosinophilic pustular dermatosis" were also proposed^{7,8}.

Although this dermatosis occurs from infancy to old age, the peak incidence is in the third decade⁹, with a male to female ratio of patients is 4.8:1⁹. The main features of this dermatosis are recurrent pruritic, follicular and sterile papulopustules, 1 to 2 mm in diameter, which become confluent to form plaques. New lesions tend to appear at the edge of the plaques, while the older more central lesions resolve with some hyperpigmentation. The distribution is usually asymmetric, with a predilection for the face, trunk and extensor aspect of the upper extremities. Rarely, lesions of the scalp, neck, axillae, groin and legs have been described. Pustular lesions on the palms and soles occur in one fourth of the patients⁸, while mucous membranes, and the dorsum of hands and feet are generally spared. In infants, the scalp is the primary focus of involvement unlike the disease in adults, the lesions often do not form annular or polycyclic rings¹⁰.

The clinical presentation of EPF varies. Although there are other dermatoses presenting with follicular pustules, it is not difficult to diagnose EPF because of the distinctive histopathologic findings.

Blood eosinophilia is present in most patients¹, however the eosinophil count may be within normal limits⁹. The peripheral eosinophil count in our cases, ranged from 45 to 134/mm³ during active disease (normal range: 0-450/mm³).

Table. Summary of the cases

	Case 1	Case 2	Case 3
Age/Sex	19/F	27/M	51/M
Site	face	face, trunk and extremities	face
Duration (month)	5	84	5
WBC count (/mm ³)	6,800	8,400	8,100
Eosinophil count (/mm ³)	126	134	45

The pathogenesis of EPF is still unknown. Takematsu et al¹¹ demonstrated that skin surface lipids from seborrheic areas of normal subjects and stratum corneum extracts from EPF lesions contained chemotactic factors for neutrophils and eosinophils. It is known that eosinophils release basic proteins which are cytotoxic to epithelium. However, the exact cause for the eosinophilic migration to hair follicles remains unclear. Cutler¹³ found a positive IgE RAST to the house dust mite *Dermatophagoides pteronyssimus* in a patient with EPF. Frentz et al¹⁴ suggested that EPF might occur as an early sign of infection with the Human Immunodeficiency Virus.

According to most authors the treatment of this dermatosis, with systemic or topical corticosteroids, dapsone¹⁵ or oxyphenbutazone, has varying results. It has been suggested that the effect of treatment may depend on the underlying cause of the disease in each patient. In our patients, these drug regimens resulted in some improvement or no response at all.

In summary, our 3 cases showed the typical skin manifestations and histopathologic findings of EPF without peripheral blood eosinophilia. We suggest, therefore, that the peripheral blood is not the source of the tissue eosinophilia in EPF and that an elevated eosinophil count in the peripheral blood is not essential in the diagnosis of EPF.

REFERENCES

1. Ofuji S, Ogaino A, Horio T et al: *Eosinophilic pustular folliculitis*. *Acta Dermatovenereol* 50:195, 1970.
2. Holst R: *Eosinophilic pustular folliculitis, report of a European case*. *Br J Dermatol* 95:661, 1976.
3. Kim JS, Kim YK, Rhim KJ et al: *Two cases of eosinophilic pustular folliculitis*. *Korean J Dermatol* 20:727, 1982.
4. Chun SI, Kang WH, Koh YJ: *A case of eosinophilic pustular folliculitis*. *Korean J Dermatol* 22:689, 1984.
5. Kim IH, Moon KC, Kim SN: *A case of eosinophilic pustular folliculitis*. *Korean J Dermatol* 23:662, 1985.
6. Ise S, Ofuji S: *Subcorneal pustular dermatosis, a follicular variant?* *Arch Dermatol* 92:169, 1965.
7. Orfanos CE, Sterry W: *Sterile Eosinophile Pustulose*. *Dermatologica* 157:193, 1978.
8. Saruta T, Nakamizo Y: *Eosinophilic pustular folliculitis, particularly with reference to lesions on the palm of the hand and sole of the foot*. *J Dermatol* 7:239, 1980.
9. Takematsu H, Nakamura K, Igarashi M et al: *Eosinophilic pustular folliculitis, report of two cases with a review of the Japanese literature*. *Arch Dermatol* 121:917, 1985.
10. Lucky AW, Esterly NB, Heskel N et al: *Eosinophilic pustular folliculitis in Infancy*. *Pediatr Dermatol* 1:202, 1984.
11. Takematsu H, Tagami H: *Eosinophilic pustular folliculitis. Studies on possible chemotactic factors involved in the formation of pustules*. *Br J Dermatol* 114:209, 1986.
12. Gleich GJ, Frigas E, Loegering DA et al: *Cytotoxic properties of the eosinophil major basic protein*. *J Immunol* 123:2925, 1979.
13. Cutler TP: *Eosinophilic pustular folliculitis*. *Clin Exp Dermatol* 6:327, 1981.
14. Frentz G, Niordson AM, Thomson K: *Eosinophilic pustular dermatosis: an early skin marker of infection with human immunodeficiency virus?* *Br J Dermatol* 121:271, 1989.
15. Steffen C: *Eosinophilic pustular folliculitis (Ofuji's disease) with response to dapsone therapy*. *Arch Dermatol* 121:921, 1985.