

Case Report

A man with classic eosinophilic pustular folliculitis presenting with bilateral facial rash

男患者雙頰皮疹 / 嗜酸性膿疱性毛囊炎

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A 26-year-old Chinese man presented with annular plaques with pustular borders over both cheeks for 6 months. He was treated empirically with topical steroid and systemic anti-fungals without improvement. Incisional biopsy of the right cheek lesion showed features compatible with eosinophilic pustular folliculitis. This patient has no evidence of immunodeficiency. Clinically, this is a case of Ofuji's disease. He responded well to non-steroidal anti-inflammatory drug.

26 歲華人男患者 6 個月來於其兩頰出現環型丘疹，其四週有膿疱性邊緣。曾接受局部皮質激素及口服抗真菌藥治療無效。於其右面頰作活組織病理學檢查，結果符合嗜酸性膿疱性毛囊炎的特徵。本患者無免疫缺陷的表現。臨床表現為太藤氏病。患者對口服非皮質激素抗炎藥治療效果良好。

Keywords: Eosinophilic pustular folliculitis, Ofuji's disease

關鍵詞：嗜酸性膿疱性毛囊炎，太藤氏病

Introduction

There are 3 major types of eosinophilic pustular folliculitis (EPF). They are, in the order

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of prevalence, the HIV infection or other immunosuppression-related EPF, classic EPF and infantile EPF. They all share similar histopathological changes but carry variable clinical features and prognosis. We report a patient with the classic EPF, which is also known as Ofuji's disease.

Case report

A 26-year-old Chinese man presented with a six month history of an itchy pustular rash over left face which gradually increased in size and progressed to involve the other side of the face. He enjoyed good past health with no previous history of drug allergy. He worked as a sales representative in a fashion boutique without

obvious exposure to chemicals or irritants. He had a history of contact with a dog with tinea infection after the onset of rash. He had not taken any drugs or herbs before the onset of the rash. Prior to our assessment, he attended a private general practitioner and was given topical steroid, oral griseofulvin and oral antihistamine. However, his condition did not improve, prompting a consultation with a private dermatologist. Skin biopsy was offered but the patient declined. He was then treated empirically with oral terbinafine, topical fusidic acid and topical isoconazole. However, the rash progressed despite treatment and he was referred to our clinic.

On examination, large annular plaques with active erythematous pustular borders, central crusting and hyperpigmentation were found over both cheeks of the patient (Figure 1). Examination under Wood's light showed no enhancement. Other body areas were not affected. Differential diagnoses included contact dermatitis, photodermatitis, tinea incognito, impetigo, eosinophilic pustular folliculitis and pustular psoriasis.

Blood tests showed peripheral eosinophilia with an eosinophil count of $0.9 \times 10^9/L$ (normal $<0.5 \times 10^9/L$). Haemoglobin, platelet and total white

cell counts were normal. Other tests including autoimmune markers, liver and renal biochemistry were also normal. HIV antibody was negative. An incisional biopsy performed over the right cheek showed epidermal hyperplasia with hyperkeratosis and parakeratosis. Heavy infiltrates of lymphocytes, polymorphs and eosinophils were found around and within pilosebaceous units as well as around dermal vessels. An eosinophilic microabscess was seen within a dilated hair infundibulum (Figures 2 & 3). There was minimal inflammatory infiltration



Figure 1. Erythematous plaque with an annular pustular rim on right cheek.

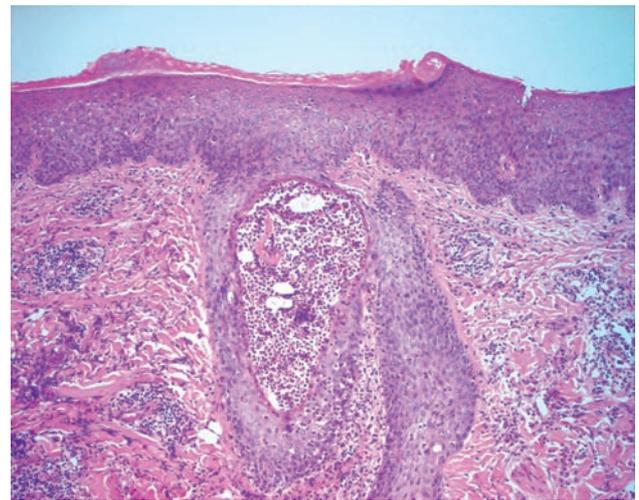


Figure 2. Hyperkeratotic epidermis and microabscess within a dilated hair infundibulum (H&E, Original magnification x 40).

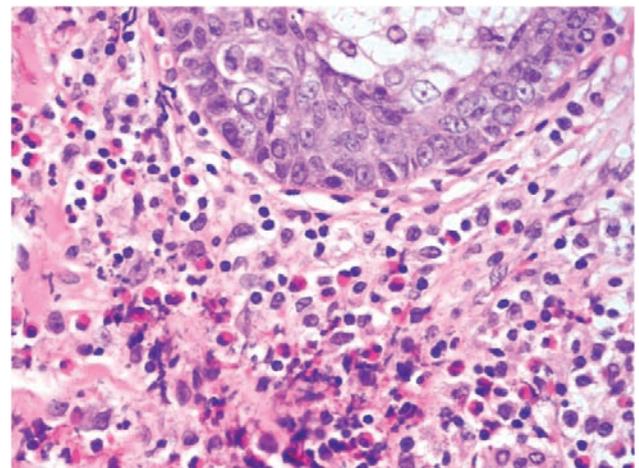


Figure 3. Mixed inflammatory infiltrates around pilosebaceous units with notable presence of eosinophils (H&E, Original magnification x 400).

in the epidermis. Special stains did not demonstrate any fungal elements. Direct immunofluorescence studies for IgA, IgG, IgM, C3 and C1q were all negative. In constellation, the clinical findings and investigations indicated that the patient got eosinophilic pustular folliculitis which was also known as Ofuji's disease.

He was treated with metronidazole 400 mg bid for 1 week together with oral indomethacin 25 mg tds and ranitidine 150 mg bid. Marked improvement was noted after 2 weeks of treatment. Clinical remission was maintained by oral indomethacin and topical steroids.

Discussion

Eosinophilic pustular folliculitis (EPF) was first described by Ise and Ofuji in 1965 in a 42-year-old Japanese woman who had follicular pustulosis on her torso, arms, and face, with blood tests demonstrating mild to moderate leucocytosis with eosinophilia. The pustules consisted of non-infectious eosinophilic infiltration of the hair follicles.¹ In 1970, Ofuji described 3 additional patients and proposed the name 'eosinophilic pustular folliculitis' for this entity.² This type of EPF was then referred to as classic EPF, or Ofuji's disease. Two additional types, the immunosuppression-associated eosinophilic pustular folliculitis and infancy-associated eosinophilic folliculitis were described later. They had similar histological findings but different clinical manifestations.¹

Ofuji's disease is a dermatosis of unknown aetiology. It occurs most commonly in Asians and is relatively rare in Caucasians. Most patients are adults, with an average age of 30 years. The male to female ratio is approximately 5:1. There have been occasional reports in children, often with clinical features similar to the infantile form.

Clinically, Ofuji's disease manifests as chronic and recurrent annular clusters of sterile follicular papules and pustules superimposed on plaques

with central clearing and peripheral extension. The lesions typically affect the face (85%), back and trunk (59%), and the extensor surfaces of the arms. However, areas without follicles such as the digits, palms and soles may also be affected. Generally, there is no systemic involvement. Individual cluster lasts for 7-10 days but relapses every 3-4 weeks. They usually heal without scarring but often result in post-inflammatory hyperpigmentation. There is mild to moderate peripheral blood leucocytosis with eosinophilia in 35% of patients.

The exact pathogenesis of classic EPF has not yet been elucidated. Proposed mechanisms include hypersensitivity reaction to various infections or medications, and autoimmune disorders possibly facilitated by skin surface lipid-derived eosinophilic chemotactic and activation factors.¹ EPF has been associated with immune dysfunction. Other than HIV infection, immunosuppression-related EPF has been associated with lymphoma, leukaemia, myelodysplastic syndrome and bone marrow transplantation.¹ Buchness et al reported that biopsy specimens showed degranulation of both eosinophils and mast cells.³ Ishiguro et al found moderately increased mast cells around follicles and sebaceous glands.⁴ There are two types of mast cells identified. One type contains the neutral proteases, tryptase and chymase, and is termed the TC mast cell, whereas the second type contains only tryptase and is termed the T mast cell. TC mast cells are predominant in skin and small intestinal mucosa whereas T mast cells are the predominant type seen in alveoli of the lung and small intestinal mucosa. The majority of infiltrating mast cells are T mast cells. Although differences in biologic activities of TC mast cells and T mast cells remain unclear, they might play a role in the pathogenesis of Ofuji's disease.

Various treatments have been tried but no definitively effective therapy has been established. First line treatment options include topical steroid, topical tacrolimus, antihistamines and oral indomethacin. Indomethacin 50-75 mg daily is the most frequently used treatment. Clinical improvement accompanied by a decrease of

peripheral blood eosinophils can usually be observed within one to two weeks after initiation of treatment.⁵ Other treatment options include dapsone 100-200 mg per day for more than two weeks, minocycline 100 mg bid, UVB phototherapy, systemic steroid, colchicine, itraconazole 100-400 mg per day, metronidazole 250 mg tds, permethrin 5% cream, PUVA, acitretin 0.5 mg per kg per day, interferon- α 3-9 x 10⁶ units per day and interferon- γ 0.5-2 x 10⁶ Japan Reference Units per day.⁶

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