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Case Report

Eosinophilic pustular folliculitis of Ofuji in an immunocompetent (non-HIV) Pakistani patient

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Abstract Eosinophilic pustular folliculitis is a rare condition characterized by recurrent, pruritic peripherally expanding, sterile papulopustules and plaques. Hair bearing areas such as face, chest, back and extensor surfaces of the upper arms are usually involved. More than 90% of cases reported are from Japan. We report a young male who had eosinophilic pustular folliculitis. He was diagnosed on clinical as well as on histological grounds.

Key words
Ofuji disease, eosinophilic pustular folliculitis.

Introduction

Eosinophilic pustular folliculitis (EPF) is an idiopathic, extremely pruritic, papulopustular follicular eruption of the upper trunk, face, neck and proximal extremities. Palms and soles can be effected, as well.[1] It is more common in immunocompromised and HIV patients.[2,3] The original cases were described by Ofuji in immunocompetent Japanese patients.[4] The peak incidence of the classic disease is in the second to fourth decades. The peak incidence is usually in the first year of life for the infantile form. EPF may be congenital in infantile cases. It may be seen at any age with HIV disease, with an incidence of almost 10% in one survey. EPF is most frequent in association with a low CD4 count.[5] The condition may be confused clinically with follicular eczema, subcorneal pustular dermatosis or pustular psoriasis but the histology is characteristic with perifollicular and perivascular eosinophilic infiltrate. When the plaques have reached a certain size, they tend to subside, leaving slight pigmentation. Many patients have either absolute or relative peripheral eosinophilia.[6] Treatment is with prednisolone, dapsone,[7] minocycline, oxyphenbutazone, INF-gamma and indomethacin.[8] Some patients have responded to topical glucocorticoids. Itraconazole and tacrolimus ointment is also reported to be effective.[9]

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Case report

A 23-year-old man presented with erythematous pruritic plaques and pustules on face, neck, ears, trunk, arms and both palms. Initial lesions started on trunk 4-5 months ago as small discrete pruritic papules and pustules. These lesions coalesced to form plaques. The lesions remained for some days then healed and new lesions were formed. On clinical examination there were

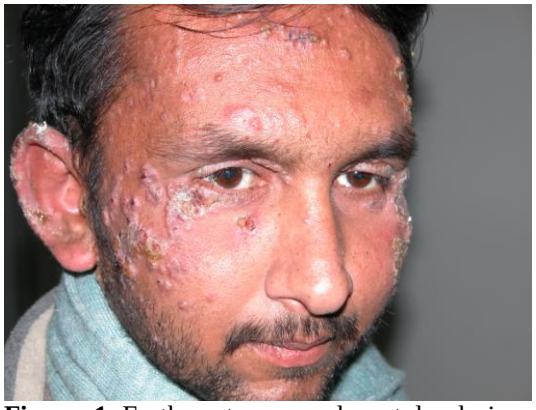

Figure 1 Erythematous papulopustular lesions over face.

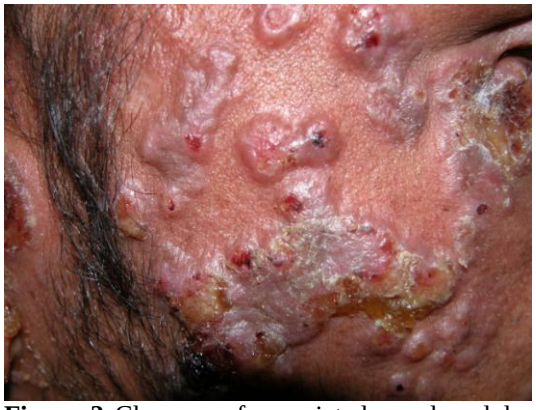

Figure 2 Close up of excoriated papulonodular lesions over right cheek.

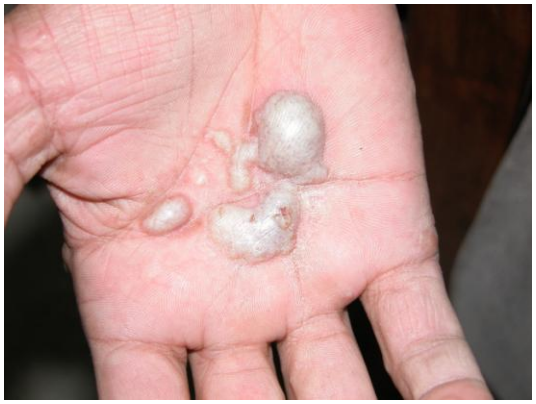

Figure 3 Pustular lesions involving the palm.

erythematous plaques studded with pustules present on face (**Figure 1** and **2)**, arms, neck and trunk. Some individual papulopustular lesions were also present on palms, as shown in **Figure** 3. In laboratory workup: ESR, 62mm; WBCs, 9900/mm^3; monocytes, 7%; lymphocytes, 49%; eosinophils, 33%;

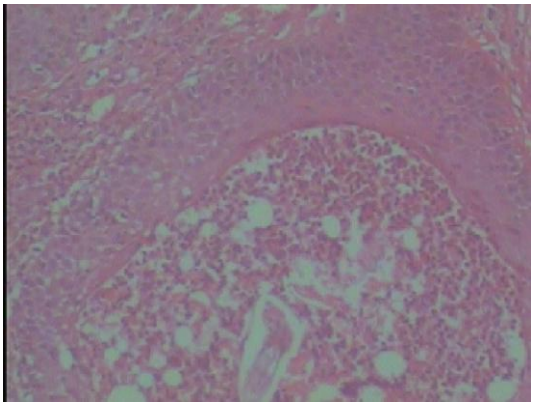

Figure 4 Dense perivascular eosinophilic infiltrate in the dermal papilla.

and platelets, 322000/mm^3. ASO titer was raised whereas rheumatoid factor, VDRL test and HIV screening were negative.

Skin biopsy revealed perifollicular and perivascular eosinophilic dermal infiltrate as shown in **Figure 4**. Epidermis was normal with some hyperkeratosis. Special stains did not reveal any evidence of fungal infection.

Clinical examination, laboratory investigations and results of histopathology confirmed the diagnosis of eosinophilic pustular folliculitis. Patient was treated with tablet prednisolone 60 mg and tablet dapsone 100mg per day. Within 20 days he improved.

Discussion

Eosinophilic pustular folliculitis is a papulopustular follicular eruption of the upper trunk, face, neck and proximal extremities. Palms and soles can be affected, as well. This disease has three variants. classic, HIV-associated and infantile EPF.[2,3] It is more common in immunocompromised and HIV patients. The peak incidence of the classic disease is in the second to fourth decades and in the first year of life for the

infantile form. EPF may be congenital in infantile cases. It may be seen at any age with HIV disease, with an incidence of almost 10% in one survey. EPF is most frequent in association with a low CD4 count. Although the histopathological hallmark of EPF is perifollicular eosinophilic infiltrate, its pathophysiology remains unclear. However, numerous studies have implicated chemotactic substances ICAM-1 and cyclo-oxygenase generated metabolites. Cytokines production e.g. interferon gamma is also increased. Immunohistochemical analysis showed expression of adhesion molecules predominantly on follicular distribution. Chemotactic factors for eosinophils and neutrophils have been found in lesional extracts.[11] The condition may be confused clinically with follicular eczema, subcorneal pustular dermatosis or pustular psoriasis but the histology is characteristic with perifollicular and perivascular eosinophilic infilterate. Many patients have either absolute or relative peripheral eosinophilia. Our patient was a case of classic eosinophilic pustular folliculitis, as he was HIV negative and peripheral eosinophil count was 33%. In the classic form of eosinophilic pustular folliculitis, mild to moderate leukocytosis and eosinophilia were evident. Different drugs have proved to be effective. Isotretinoin leads to a dramatic improvement but withdrawal is followed by recurrence of lesions. Indomethacin is most frequently used drug and it has shown clinical improvement in a lot of cases. The effect is observed within 1-2 weeks. Interferon-α 2b is also reported to be effective. Other treatments include oral steroids, diaphenyl sulphone, tacrolimus, colchicine, minocycline and UVB phototherapy and radiotherapy.[8-11] There are different studies that show the effectiveness of dapsone in cases of follicular pathologies which do not respond to conventional therapies. Ofuji's disease is one of them.[7] We treated our patient with dapsone and topical steroid and he improved within one month.

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