Case study

Eosinophilic pustular folliculitis in a 90-year-old patient

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Abstract

Introduction: Eosinophilic pustular folliculitis (EPF), sometimes called Ofuji disease, is a chronic skin disorder of unknown etiology which develops predominantly in elderly man. Clinically it usually presents as itching centrifugally extending papulopustular rash.

Aim: To present a case of EPF and to raise dermatologists’ awareness of this dermatosis when dealing with patients who are in higher risk of EPF occurrence.

Case study: A 90-year-old man presented to the dermatology clinic due to the itching rash that had been present for 3 months. Physical examination of the patient revealed red-brown papulopustules covering trunk and extremities, most severe on the frontal part of thighs. Other possible causative agents of the symptoms were excluded. Histopathological results together with clinical symptoms led us to the final diagnosis of classic EPF.

Results and discussion: EPF is challenging to diagnose. The disease is very rare, especially in Europe, and clinical and/or histological appearance may mimic other dermatological conditions. The most commonly affected areas are face, back and trunk. EPF is usually classified into three main subtypes: classic, infantile and immunosuppression-associated. There are many viable therapeutic algorithms for the treatment of EPF.

Conclusions: Eosinophilic pustular folliculitis should be considered in differential diagnosis of pruritic follicular papulopustular rash affecting elderly men. Histopathological results play a crucial role in the making of final diagnosis.

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1. INTRODUCTION

Eosinophilic pustular folliculitis (EPF) is a chronic, inflammatory dermatosis of unknown etiology characterized by annular, centrifugally extending sterile papulopustules. Although anyone can be affected, EPF is predominantly described in elderly men, with greater occurrence in Japan. It was originally described in 1965 by Ise and Ofuji as ‘subcorneal pustular dermatosis.’ The most commonly affected areas are face, back and trunk. It does not affect mucous membranes.

2. AIM

The aim of this report is to describe a case of EPF. We would like to increase the awareness of this skin condition among doctors and, hence, to facilitate the diagnostic procedure.

3. CASE STUDY

A 90-year-old patient with Parkinson disease attended the dermatology clinic due to a rash that had been present for the past 3 months. The patient’s history of atopy was negative, he denied drug allergies or newly induced drugs. He did not recall any clear trigger factors. In the past, patient was treated with topical permethrin with no improvement. Neither his wife nor people in the close surroundings had similar symptoms. He received a first dose of the COVID-19 vaccine after the eruption of skin changes.

Physical examination showed 5 mm-sized erythematous red-to-brown papules and pustules on the trunk and frontal parts of thighs (Figure 1). Face was spared from disease; mucous membranes were not affected either. Patient provided a result of previous histopathological skin biopsy. However, the overall result was not determining and did not correlate with the clinical symptoms, hence the second skin biopsy was performed. Histological examination revealed intense eosinophilic infiltration (Figure 2).

Laboratory tests results showed increased total immunoglobulin E (3581 IU/mL, reference range <100 IU/mL) and eosinophilia (0.88 × 10^9/µL, reference range 0.12 to 0.24 × 10^9/µL). Tumor blood markers (carcinoembryonic antigen – CEA, alpha fetoprotein – AFP, cancer antigen 19-9 – CA19-9, prostate specific antigen – PSA) as well as inflammatory markers (erythrocyte sedimentation rate, C-reactive protein – CRP) were negative. Veneral diseases research laboratory test (VDRL), human immunodeficiency virus (HIV) screening test, hepatitis C antibodies (anti-HCV) and hepatitis B antigen (HBs antigen) were also negative. Possibility of COVID-19 infection was eliminated with negative PCR result. Diagnostic imaging investigation did not reveal any ongoing neoplastic proliferation. The patient was prescribed oral cyclosporine-A (300 mg daily) and oral prednisone (30 mg daily) with good response to reduction of rash and itching.

Malignance, atopy and drugs were excluded as causative agents of the symptoms. Since scabies in the elderly is often misdiagnosed and can mimic many pruritis dermatoses, infection with Sarcoptes scabiei var. hominis was excluded as well. Use of clinical follow-up together with histopathological results led us to the final diagnosis of classic EPF.

4. RESULTS AND DISCUSSION

EPF is predominantly seen among elderly man with trunk, extremities and face being most commonly affected skin areas. While our patient did not have facial lesion, the extremities and trunk were severely affected. Apart from leukocytosis and eosinophilia observed in about 30% patients, there are no specific laboratory abnormalities. EPF is classified into three main subtypes: classic, infantile and

Figure 1. Clinical examination showed red-brown papulopustural lesion on legs (a), trunk (b) and back (c) of the patient.
immunosuppression-associated.\textsuperscript{4,5} However, drug-induced EPF is associated with administration of medications such as allopurinol, carbamazepine, minocycline and indeloxazine hydrochloride.\textsuperscript{8} As EPF occurs rarely especially in Europe, it proves difficult to diagnose. Its clinical and/or histological appearance may mimic other dermatological conditions like scabies, bacterial folliculitis, cutaneous T-cell lymphoma, dermatomycosis, follicular mucinosis, or reaction to foreign body. Therefore, biopsy and thorough histological examination are crucial for the final diagnosis. The treatment of EPF is challenging. The course of the disease is usually chronic, skin lesions tend to wax and wane causing a slight hyperpigmentation upon subsiding. The main treatment goal is to resolve skin lesions and control the recurrence. Nomura et al. described many viable therapeutical algorithms indicating oral and topical indomethacin as the first-line drug in classic-EPF.\textsuperscript{9} Interestingly, phototherapy (UVB or narrowband UVB or UVA) and photochemotherapy (psoralen plus UVA – PUVA) are successful treatment options for maintenance therapy. Other effective treatments reported so far is either low-dose isotretinoin alone or a combination of low-dose indomethacin and topical tacrolimus.\textsuperscript{10,11}

5. CONCLUSIONS

While cases of EPF occur scarcely, dermatologists should be aware of this dermatosis when dealing with patients who are in higher risk of EPF occurrence.

Conflict of interest

None declared.

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None declared.

Ethic

The patient gave consent for publication of case details and images.

References


