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Case Report

Papuloerythroderma of Ofuji in a young adult

Mary Mathew[1], Bhavna Nayal[1], Raghunath Narayanan Unni[1], C. Balachandaran[2]

[1]*Departments of Pathology, KMC, Manipal University, Manipal, India*
[2]*Departments of Dermatology, KMC, Manipal University, Manipal, India*

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Corresponding Author:
Bhavna Nayal,
Kasturba Medical College,
Manipal University, Manipal, India
bhavnayal@yahoo.com

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Abstract

Papuloerythroderma of Ofuji (PEO) is an inflammatory skin condition with typical clinical presentation including generalized itchy papular eruptions, sparing the major skin folds in the body. They usually have peripheral eosinophilia and elevated serum IgE levels. This condition may be associated with an internal malignancy, infection or drug therapy. It usually affects the elderly and very few cases have been reported in young adults. We report a 31 year old male with PEO in the absence of an underlying inciting factor.

INTRODUCTION

Papuloerythroderma of Ofuji (PEO) is a rare skin disease described first by Ofuji in 1984. This disease is characterized by extensively pruritic erythematous eruptions involving the entire body which spares the body folds and is associated with peripheral eosinophilia [1]. It is often associated with an underlying malignancy and infectious disease [2, 3]. This disease occurs exclusively in the elderly [3]. However, two cases have been reported in patients <50 years of age [4, 5]. We report an additional case of PEO occurring in a 31-year old male with no evidence of underlying malignancy or infection. The patient showed a dramatic response to steroid therapy.

CASE REPORT

A 31-year- old male presented with complaints of generalized itchy scaly lesions since 3 months. These lesions developed over the abdomen and gradually spread to the upper limbs followed by lower limbs and later progressed to involve the whole body. The face, scalp, palms, soles and regions of skin creases were spared. He did not have similar episode in the past. There was no history of weight loss, systemic symptoms, drug intake prior to the onset of the lesions or allergy. On examination, the lesions were erythematous scaly papules. His hair and nails were normal. In addition, he had cervical, left axillary and bilateral lymphadenopathy. There were no signs and symptoms of any internal malignancy. Routine hematological investigations revealed an absolute eosinophil count of 3.3x10, 00/µl. The peripheral smear revealed eosinophilia (35%) and mild lymphocytopenia (11%). No circulating abnormal lymphoid cells or Sezary cells were seen. Serum immunoglobulin E level was normal (128.6 IU/ml). Skin patch test with Indian standard series and parthenium was negative. The patient was referred to the department of medicine to rule out coexisting systemic disease. A detailed general physical examination was normal. Serological tests for malaria, scrub typhus, leptospira, orienta tsutsugamushi, human immunodeficiency virus and Hepatitis B antigen were negative. Urine and stool examination were normal. Liver and renal function tests detected no abnormalities. A clinical diagnosis of papuloerythroderma of Ofuji was proffered. The lymph node biopsy showed dermatopathic lymphadenitis.

Lesional biopsy from the trunk revealed a hyperkeratotic and irregularly acanthotic epidermis with mildly elongated and thickened rete ridges (Figure 1). The dermis showed pigment incontinence, ectatic blood vessels with perivascular lymphohistiocytic mononuclear inflammatory and eosinophilic infiltrate (Figure 2 and 3). There was no evidence of infiltration by abnormal lymphoid cells. Based on the clinical and histological features, a final diagnosis of papuloerythroderma of Ofuji/ primary papuloerythroderma was rendered. The patient was started on tapering doses of oral methylprednisolone, antihistamines along with two sittings of narrow band ultraviolet (UV) B therapy, to which he responded dramatically.

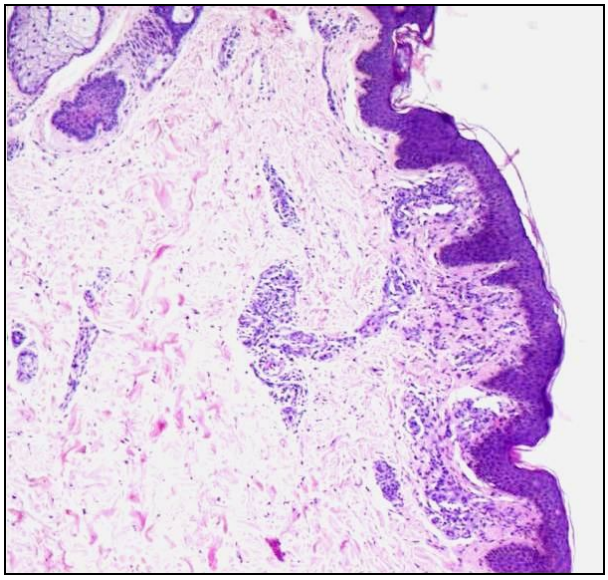

Figure 1. Hyperkeratotic and acanthotic epidermis with elongated rete ridges (H&E, x100).

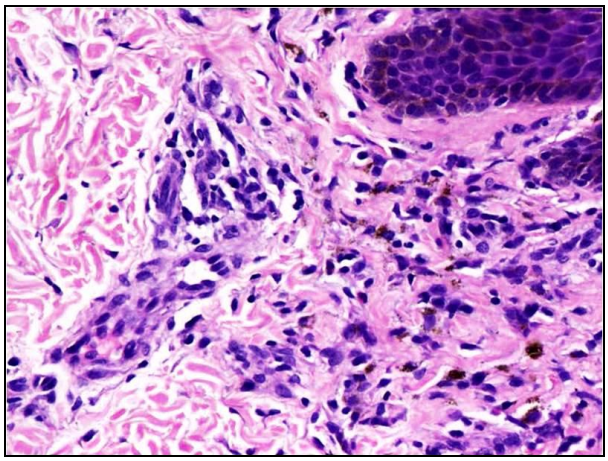

Figure 2. Dermis shows melanin incontinence and perivascular inflammatory infiltrate (H&E, x200).

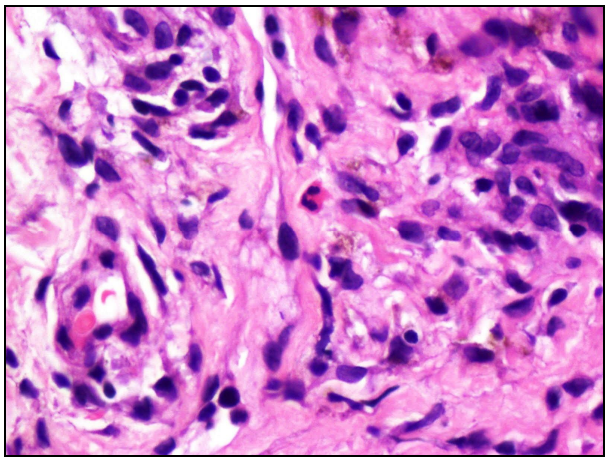

Figure 3. Perivascular eosionophilic and lymphocytic infiltrate in the dermis (H&E, x400).

DISCUSSION

Papuloerythroderma of Ofuji is an uncommon dermatosis with typical clinical features. It was first reported by Ofuji in 4 normal healthy adults [1]. These patients present with generalized pruritic confluent papules with flat tops. The pressure zones and normal body folds were characteristically spared. This was called the “deck-chair sign” by Farthing et al [6]. The face is usually spared in this condition [2].

The mean age of occurrence of PEO is approximately 73.42 years and it is known to affect the elderly. Only two cases of PEO have been described in the younger age group. The first documented case was in a 31 year old HIV infected male with lesions induced by dideoxyinosine therapy [4]. The second was a 41 year old female with underlying Hodgkin lymphoma [5]. The present case neither had evidence of any underlying malignancy nor history of exposure to any triggering factor.

PEO can coexist, precede or follow or may be a cutaneous manifestation of many malignancies, commonly gastric cancer. Other malignancies associated with PEO are hematological neoplasms such as B cell lymphoma, T cell lymphoma and Mycosis fungoides, lung and hepatocellular carcinomas. Such patients show poor response to treatment [2, 3]. Recently, a case of coexisting Bazex syndrome and PEO preceding hepatocellular carcinoma was reported [7]. Infections such as HIV and drugs have also been inciting agents for PEO [3, 5]. However, many cases of PEO with absence of any identifiable etiology have been reported [3, 8, 9].

In addition to the typical skin lesions, PEO may also present with dermatopathic lymphadenopathy, palmoplantar keratoderma, nail infarcts, peripheral

eosinophilia, lymphocytopenia, circulating Sezary cells and elevated serum IgE levels [1-3]. Our patient had dermatopathic lymphadenitis, peripheral eosinophilia and lymphocytopenia.

Histologically, the skin lesions show features of eczema or chronic dermatitis with epidermal hyperkeratosis, acanthosis, elongated rete ridges, focal spongiosis and exocytosis. The dermis shows perivascular inflammatory infiltrate composed of lymphocytes, histiocytes, eosinophils and Langerhans cells along with melanophages [2, 3]. Unusual features such as giant cells and granulomas have also been described [2].

Torchia et al laid down the diagnostic criteria for classifying papuloerythroderma which consists of necessary and minor criterias. The necessary criteria (from 1 to 5) includes erythoderma-like eruptions, 'deck-chair sign', itching, exclusion of cutaneous T cell lymphoma or other skin diseases and exclusion of malignancy, infections, drugs or atopy. The minor criteria are age >55 years, males, eosinophilia, elevated IgE levels and peripheral lymphocytopenias [3].

Based on these criteria, an etiological classification for papuloerythroderma was proposed: PEO (primary/idiopathic papuloerythroderma) (criteria 1-5), secondary PEO incorporating atopic, paraneoplastic, infection and drug induced PEO (criteria 1-4) and papuloerythroderma-like cutaneous T cell lymphoma (CTCL) (criteria 1, 2, 3, and 5). Erythroderma with the typical deck-chair sign but lacking papules was termed as pseudopapuloerythroderma [3]. The present case fulfilled all 5 necessary criteria described by Torchia et al.

The differential diagnosis of PEO includes exfoliative erythroderma, generalized lichen planus or lichen planus like drug eruption. However, sparing of body creases and histopathology aids in confirming the diagnosis of PEO [2].

The treatment modalities for PEO include topical or oral corticosteroids, antihistamines, PUVA and UV therapy in various combinations. Majority of the patients show prompt relief from symptoms following therapy has been reported [2, 3, 8, 9]. Our patient showed a prompt response to combination therapy.

In conclusion, Papuloerythroderma of Ofuji is a heterogeneous disease with typical skin lesions. Although PEO is a disease of the elderly, young adults may also be affected. In addition, association of this condition with internal malignancies emphasizes the importance of careful systemic examination and long term follow up.

CONFLICTS OF INTEREST

The authors declare that they have no conflict of interest.

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