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Papuloerytroderma of *Ofuji*: Case Report

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Abstract

Observation: Papuloerythroderma of *Ofuji* is a disease characterized by intensely pruritic papules which may progress to cutaneous lymphoma. Serum IgE levels are high and peripheral eosinophilia is present. Most of the patients are men. This paper reports a thirty years old Kyrgyz male having pruritus and erythema all over the body. From patient's history, it was learned that there had been erythema with pruritis and edema throughout the body lasting for 3 years and there had been no recovery despite various medical treatments. There was dermopathic lymphadenopathy, peripheral eosinophilia and high IgE levels. Skin biopsy revealed increases in Langerhans cells stained as S-100+ and thus, the case was diagnosed as *Ofuji*'s papuloerythroderma. By means of this case, data about this disorder will be able to reviewed.

Introduction

Ofuji's papuloerythroderma of is a rare disease with unkonown etiology, and sometimes it may progress to cutaneous lymphomas. It is characterized by intensely pruritic papules which is widespread, flat and reddish and tending to coalescense. Majority of patients are men. Deck-chair view is typical because the disease spares the body folds.Serum IgE levels ar elevated and peripheral eosinophilia present [**1, 2**].

Case Report

A 30-year-old male patient from Kyrgyz descend submitted May 2005 with complaints of intensely pruritic surfaces and edema all over the body surface. Patient's history revealed that he had pruritic erithemas and swellings particularly in the trunk and that lasted for three years, and that in different centers, he had been investigated for erythroderma however, no result could have been obtained for clarification of etiology. At admission, patient had widespread erithematous papules throughout his body and had been using 20 mg of prednisolone for 8 months. He had been hospitalized for detailed investigation with potential diagnoses of PRP, parapsoriasis, mycosis fungoides, acquired ichtyosis, lichen scroflosorum and papular drug reaction. On examination, there was no pathologic findings except bilateral inguinal lymphadenopathies with a diameter of 0.5 cm. On dermatologic examination, there was an appearance in which erithematous papules located to the back and upper extremities with a diameter of 1 to 3 mm forming plaques by coalescing, and that especially folding areas were spared. In addition, there were erythema and desqumation over 80% of body surface (**Figure 1**). Laboratory investigations revealed that peripheral eosinophils of 8.9%, total IgE level 554, other laboratory tests were in

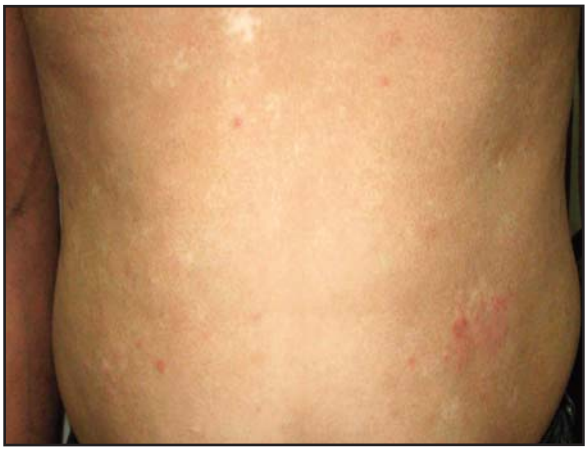

Figure 1. Erithematous papules located to the abdomen and upper extremities with a diameter of 1 to 3 mm forming plaques by coalescing

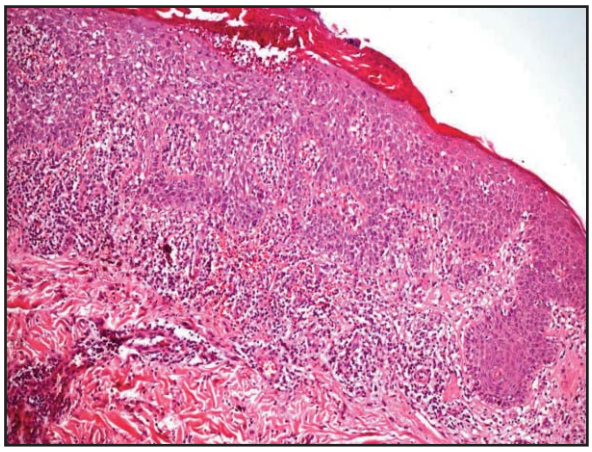

Figure 2. Histology parakeratose in which there was neutrophil and eosinophil clusters, mild spongiosis, irregular acanthosis, dense perivascular lymphocyte and neutroophil infiltration in upper dermis, localised eosinophil existence in upper and middle dermis and increse in langerhans cells stained with S-100

normal ranges. Abdominal ultrasound showed no pathologic findings except inguinal lymphadenopathy. Biopsy obtained from lymph nodes was diagnosed as dermopathic lymphadenopathy. Skin biopsy revealed parakeratose in which there was neutrophil and eosinophil clusters, mild spongiosis, irregular acanthosis, dense perivascular lymphocyte and neutroophil infiltration in upper dermis, localised eosinophil existence in upper and middle dermis and increse in langerhans cells stained with S-100 (**Figure 2**).

Discussion

Ofuji's papuloerythroderma is a rare entity first described by *Ofuji* et al. at 1984 in four elderly Japanese male patients [**1**]. It is characterized with intensely pruritic red papules having widespread distributiony, a flat surface. Body folds and creases are less involved (so-called 'deck-chair' sign) [**1, 2, 3**]. It is a chronic disease refractory to treatment, reducing quality of life. In a retrospective study from Bech-Thomsen and Thomsen, estimated annual incidence is one and a half per million, and average age of diagnosis is 72 years [**1**]. Our patient was 30 years old. This disorder occurs particularly in elderly men and frequently related with lymphadenopathy, eosinophilia and high IgE levels in peripheral blood [**1, 2, 3, 4, 5**]. These findings were present in our patient.

Ethiopathogenesis is not known, however it is hypothetised that it is a rare variant of several inflammatory dermatoses, particularly atopic dermatitis in elderly patients. It may also occur as a finding of internal malignancy, AIDS, hypersensitivity to drugs or skin lymphomas [**1**]. Skin lymphomas can be arised as Papuloerythroderma of *Ofuji* (PEO) or it can be seen as an early form of skin T lymphomas. Therefore, these patients need long term follow up. It is observed that mucosa is definitely not involved [**3**]. Lympadenopathy, infarcts in nailfolds and buttocks, palmoplantar keratoderma, visceral malignancies and circulating Sezary cells may be seen [**1**].

Histology of PEO is non-specific. Epidermis is usually normal however mild acanthosis, spongiosis, parakeratosis and rarely exocytosis may be seen. There is a perivascular inflammatory infiltrates in which mainly T lymphocyte, histiocyte, eosinophils and plasma cells in upper and middle dermis [**1, 2**]. Systemic steroids, cyclosporine, azathioprine, interferon alpha, PUVA, systemic retinoids, retinoid+PUVA (RePUVA) are all among treatment ooptions [**2**]. Systemic corticosteroids are generally effective however, relapses may be seen when dose is lowered and adverse effects can occur due to steroids. Bech-Thomsen and Thomsen reported that PUVA and oral prednisolone in combination or alone was effective for treatment [**2**]. We used systemic steroid and asitretin in our case. We observed new lesion upon steroid withdraval despite continuing asitretin treatment. We report this case to contribute to the literature since this disease is a rare entity and there is still no established information for the diagnosis and treatment.

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