Papuloerythroderma of Ofuji in Indonesia: the first case report

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Abstract

Papuloerythroderma (PE) is a rare skin disease which was first described by Ofuji et al. in 1984, with a typical sign that the lesions spare the large cutaneous folds, known as the deck chair sign. Due to its recent identification, this disease is still underrecognized and may lead to misdiagnosis. We reported the first case report of PE of Ofuji from Indonesia in which the diagnosis was delayed for two years. Besides the deck chair sign in the large cutaneous fold, we also found that the area between and above his eyebrows that was relatively spared in contrast to the sparing of the cutaneous folds, and it may be considered as pseudo-deck chair sign. The patient showed good response with combination therapy of phototherapy with Narrow-Band Ultraviolet B (NBUVB), oral methotrexate, and corticosteroids. The deck chair sign disappeared after six months therapy, but the patient’s skin was still xerotic.

Key words: Papuloerythroderma of Ofuji, deck chair sign, pseudo-deck chair sign

Introduction

Papuloerythroderma (PE) is a rare, well-characterized skin disease, first described by Ofuji et al. in 1984.1 PE mostly affects elderly men. Reports show association of PE with malignancy in 21.76% of patients, atopic dermatitis in 9.23% of patients, and drug-induced hypersensitivity in less than 5% of patients.2-4 It is characterized by a widespread eruption of intense pruritic papules which later coalesce to form plaques, and potentially evolve into erythroderma. The typical sign of PE is the free lesions of large cutaneous folds, which is known as the deck chair sign.5 Bech and Thomsen6 found an annual incidence of 1.5 per million over ten years observation.
However, poor awareness among dermatologists, may underlie the fact that PE is still underrecognized, underdiagnosed, and underreported. To the authors’ knowledge, this is the first case report of PE in Indonesia.

**Report**

A 62-year old male, diagnosed as exfoliative dermatitis caused by seborrheic dermatitis was referred to the outpatient Department of Dermatology and Venereology, dr. Sardjito General Hospital Yogyakarta. There was a 2-year history of itchy skin lesions. It started as multiple itchy red papules on both thighs, which then eventually distributed to cover almost full body surface. He then visited a dermatovenereologist. Initially, the diagnosis was exfoliative dermatitis. He received oral corticosteroid, antihistamines, topical corticosteroids, and emollients for 2 weeks. His symptoms were responsive to the therapy but recurred after the treatment finished. A skin biopsy was performed one year after the first visit. The pathology result suggested exfoliative dermatitis with seborrhic dermatitis. He then received the treatment as mentioned above for another two weeks.

A month before he came to our clinic his skin condition worsened. He was hospitalized in a district hospital for a week. At the time of hospital discharge, the treatment were oral antihistamines, topical corticosteroids, and olive oil. The itch severity decreased and his skin became hyperpigmented. He took this treatment for three weeks, but the symptoms recurred two days after the medication was stopped.

The patient denied any drug consumption prior to the skin eruption. There was no history of a prolonged cough, gastrointestinal tract complaints, or rapid weight loss. No risk for HIV infection was found.

On examination the patient’s vital signs were within normal limits. There were multiple brown, flat-topped scaly papules covering his trunk and extremities. The abdominal and cubital folds were free of lesions *(Figure 1).* There were flat-topped papules on the cheeks and infiltration in the forehead with prominent skin lines. The area between and above his eyebrows was relatively free of lesions *(Figure 2).* No lesions were found on his scalp. The distribution of the lesion did not correspond to the Langer’s lines. There were smooth, circumscribed lymph nodes in his left axilla and right inguinal fold.

Peripheral blood examination showed mild leucocytosis with absolute neutrophilia (9,060 cells/µl). Lymphocyte, eosinophil, erythrocyte, and platelet counts were normal. Haemoglobin, hematocrit, transaminase enzymes, blood urea nitrogen, and immunoglobulin E (IgE) were within normal limits. No Sezary cells were found.

Histopathological examination taken from patient’s left arm stained with hematoxylin eosin revealed orthokeratosis, flattening of rete ridges, and focal spongiosis in the epidermis with lymphocyte infiltration. In the upper dermis, there were lymphocytes and histiocytes patchy infiltrates with sparse eosinophils, and edema. *(Figure 3).* The conclusion was spongiotic dermatitis, compatible with PE. The immunohistochemistry staining showed CD 45+ and CD3+ cell infiltrates in the dermis and epidermis, which may indicate a T-cell origin of the infiltrates.

The patient was diagnosed with primary PE and treatment was started with a combined therapy of daily 16 mg oral methylprednisolone and weekly 10 mg methotrexate, and phototherapy with Narrowband Ultraviolet B (NBUVB). The NBUVB therapy was conducted twice a week, and was started from 500mj/cm² to increase 25% after every three sessions. After three weeks of the combined therapy, the itch intensity lessened and the papules flattened, but xerotic scaly skin and the deck chair sign was still visible. After six months of treatment, the scaly xerotic skin was still present but the deck chair sign had disappeared *(Figure 4).*

**Discussion**

Classical PE of Ofuji has typical clinical manifestations of generalized pruritic eruption mimicking erythroderma with itchy flat-topped papules which coalesce generally. The lesions spare large skin folds and pressure areas, and this is known as the deck chair sign. Besides the typical skin lesions appearance, in PE, lymphadenopathy, palmpoplantar keratoderma, and nail infarcts may also be present.

The patient had a history of chronic itch and redness covering his entire skin with multiple brown flat-topped papules which spared his abdominal and cubital folds. There was also lymphadenopathy found.

Torchia et al. in their review, mentioned that face and scalp are seldom involved. In our patient, the lesions were also found on his face.
Figure 1. Abdominal and cubital folds were lesions free, which is known as a *deck chair sign*. There was cobblestone appearance of the lesional skin.

Figure 2. The area between and above his eyebrows was relatively free of lesions. We propose to identify this as a pseudo-deck chair sign.
Figure 3. Histopathology examination with hematoxylin eosin staining (x400) showed spongiotic epidermis with lymphocytes and histiocytes infiltrate in the upper dermis, concluded as spongiotic dermatitis.

Figure 4. Patient's skin condition after six months of treatment. Xerotic and scaly skin, the deck chair sign on the abdominal skin disappeared.
Moreover, the area between and above his eyebrows was relatively free of lesions. We propose this phenomenon to be considered as a pseudo-deck chair sign (Figure 2), as it is in contrast to the sparing of the cutaneous folds.

Laboratory results of PE patients revealed eosinophilia in 85% of patients, low lymphocyte count in 29% of patient, and an increase in IgE serum level in 50% of patients.\(^2\)\(^,\)\(^3\) None of these was found in our patient.

Histopathologic examination from the skin lesion of PE is not characteristic. In the epidermis hyperkeratosis, acanthosis, and focal spongiosis, in the dermis perivascular infiltrates consisting mostly of lymphocytes, plasma cells, histiocytes, and eosinophils.\(^7\) In our patient, the histopathological result was compatible with PE. Torchia et al.\(^4\) classified PE according to its etiology. Primary PE is when the underlying cause is unknown. Secondary PE is referred to PE that is associated with atopic dermatitis, malignancy, infection, or drug-induced. In cutaneous T-cell lymphoma (CTCL) like PE, the patients have PE clinically, but their histopathology results fit CTCL. Pseudopapuloerythroderma is a condition of an erythroderma without papules but featuring a deck chair sign on the large skin folds, like those found in PE.\(^4\) Our patient fitted the primary PE, because we could not find any underlying disease related to PE, and there were no Pautrier's abscess nor cerebri form lymphocytes in histopathology examination.

Torchia, et al.\(^4\) also reported that the median duration of the skin rash at the time of diagnosis was seven months. Before the patient was referred to our hospital, he was diagnosed as having exfoliative dermatitis, a disease characterized by erythema scaly skin involving more than 90% of the body surface.\(^8\) Initial misdiagnosed by a dermatologist resulted in 2 years delayed diagnoses, longer than the average delayed time suggested by Torchia et al.\(^4\)

The skin areas without lesions may lead clinicians to make a differential diagnosis of pityriasis rubra pilaris (PRP). PRP is a papulosquamous disorder with follicular hyperkeratotic papules, palmoplantar keratoderma, and widespread orange-red erythema with islands of skin without lesions.\(^9\) The histopathology of PRP shows hyperkeratosis with alternating orthokeratosis and parakeratosis, hypergranulosis, and a perivascular lymphocytic dermal infiltrate.\(^9\)\(^,\)\(^10\) Our patient did not have follicular hyperkeratotic skin lesions, palmoplantar keratoderma, and the skin lesions were not orange-red in color. The histopathology did not show any alternating, orthokeratosis and parakeratosis; thus the diagnosis of PRP can be excluded.

Bech-Thomsen and Thomsen\(^6\) reported improvement of PE cases treated with systemic corticosteroids, phototherapy with UVA (PUVA), combination therapy of PUVA with systemic corticosteroids or phototherapy with UVB in combination with potent topical corticosteroids.\(^6\) The itch severity and papules redness of our patient improved after receiving combination therapy of NBUVB, oral methotrexate, and oral corticosteroids for three weeks. Six months after this treatment combination, the scaly xerotic skin was still present but the deck chair sign had disappeared.

**Conclusion**

Papulo-erythroderma of Ofuji is a rare skin disease with clinical characteristic known as the deck chair sign. In our case, there were two years of diagnosis delay since the onset of the disease. The skin condition showed a good response to combination therapy of NBUVB, oral methotrexate, and oral corticosteroid.

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**References**