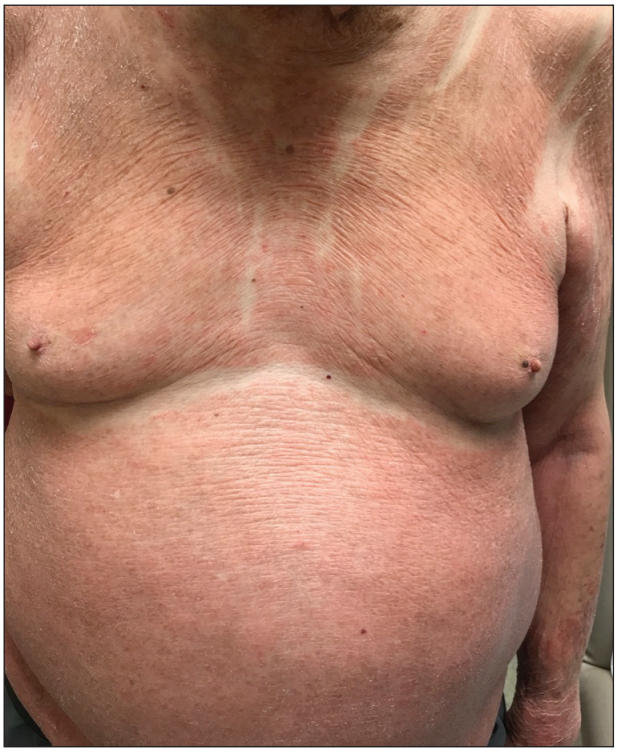
Pruritic Eruption With Skinfold Sparing

Taylor Novice, BS; Danielle Yeager, MD; David Oberlin, MD; Henry Lim, MD



An 89-year-old Asian man presented with a generalized pruritic eruption of 2 months' duration. The rash started on the flanks and later spread to the arms and legs, abdomen, and back; the face and palms were spared. Physical examination revealed numerous erythematous papules coalescing into large scaly plaques on the trunk, arms, and legs. There were noticeable areas of sparing of the skinfolds, especially the axillary, inframammary, and inguinal folds, as well as the midline of the back. A biopsy performed by an outside physician showed findings consistent with a possible pityriasiform drug eruption; however, there were no recent changes in medication history.

WHAT'S THE **DIAGNOSIS?**

a. cutaneous T-cell lymphoma
b. malignant acanthosis nigricans
c. papuloerythroderma of Ofuji
d. parthenium dermatitis
e. pityriasis rubra pilaris

PLEASE TURN TO PAGE E9 FOR THE DIAGNOSIS

Ms. Novice is from University of Michigan School of Medicine, Ann Arbor. Drs. Yeager, Oberlin, and Lim are from Department of Dermatology, Henry Ford Hospital, Detroit, Michigan.
The authors report no conflict of interest.
Correspondence: Taylor Novice, BS, 3031 W Grand Blvd, Ste 800, Detroit, MI 48202 (tnovice@med.umich.edu).

THE **DIAGNOSIS:** Papuloerythroderma of Ofuji

The patient presented with a characteristic finding of skinfold sparing, known as the "deck-chair sign" (Figure 1).[1] A repeat biopsy at our institution revealed a dermal perivascular and bandlike infiltrate with lymphocytes and occasional eosinophils (Figure 2). The epidermis showed mild spongiosis, lymphocytic exocytosis, and rare necrotic keratinocytes. A T-cell gene rearrangement assay was negative for a monoclonal population of T lymphocytes. Based on the clinical and histologic features, the diagnosis was most consistent with papuloerythroderma of Ofuji (PEO); however, a lymphoproliferative disorder needed to be excluded. Further workup included a peripheral smear, complete blood cell count with differential, comprehensive metabolic panel, IgE level, and hepatitis panel; all were normal, except for an elevated serum IgE level. Human immunodeficiency virus and age-appropriate malignancy screening were negative. The patient was prescribed betamethasone dipropionate cream 0.05% twice daily, which resulted in near-complete resolution of the rash and marked improvement in pruritus.

In 1984, PEO was described as an entity of generalized pruritic erythroderma characterized by flat-topped, red to brown, coalescing papules with sparing of the skinfolds, later coined the deck-chair sign.[1,2] Papuloerythroderma of Ofuji commonly presents in elderly Asian males with a male to female ratio of 4:1.[3] Papuloerythroderma of Ofuji is a T cell–mediated skin disease; however, the etiology of the signature rash remains unclear. One explanation includes circulating factors in the skin that elicit an inflammatory response, which does not occur in areas of external pressure.[3] The deck-chair sign may occur more frequently in elderly individuals due to increased skin laxity, which creates crease lines that are spared from rubbing and excoriations.[4]

Although Ofuji et al[2] initially reported 4 idiopathic cases, subsequent authors described PEO in association with other conditions, including cutaneous T-cell lymphoma (CTCL) and atopic diathesis, and infections, as well as secondary to medications. Some authors have suggested that PEO may be an early variant of mycosis fungoides; therefore, physicians should monitor patients closely.[5-7] Maher et al[6] commented that multiple causative factors including CTCL underlie the development of papuloerythroderma.

In a review of PEO, Torchia et al[3] proposed diagnostic criteria and an etiologic classification to address whether PEO represents an independent entity or an unusual manifestation of other dermatoses. They established 4 categories of papuloerythroderma—primary, secondary, papuloerythrodermalike CTCL, and

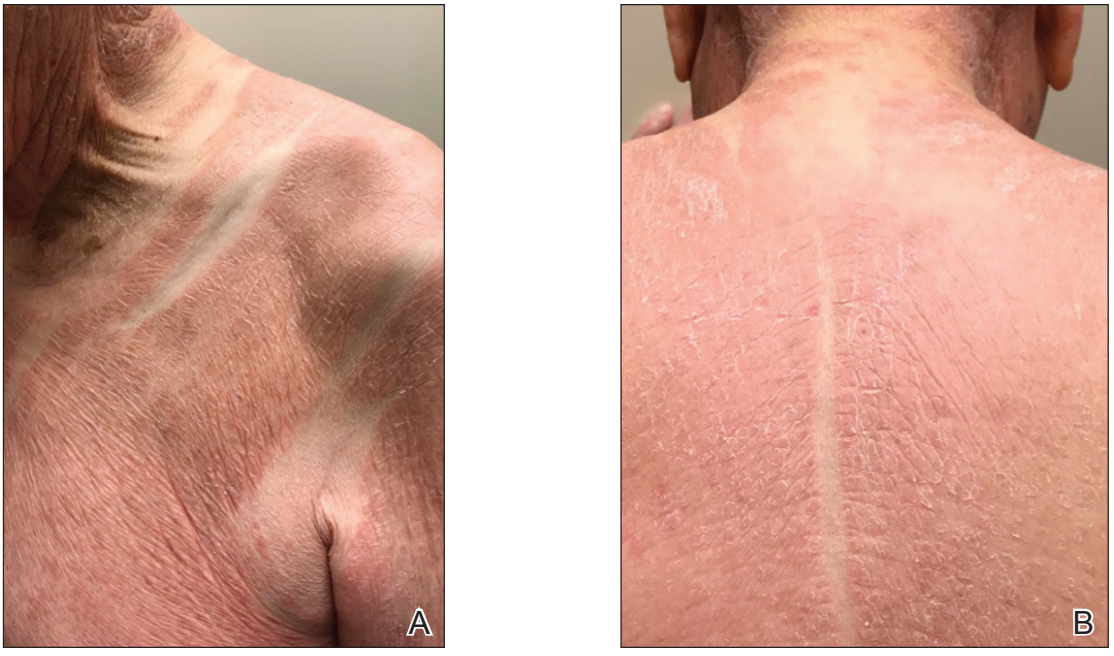


FIGURE 1. A and B, The deck-chair sign with sparing of the skinfolds along the neck lines and infra-axillary region as well as in the folds of the central mid back.

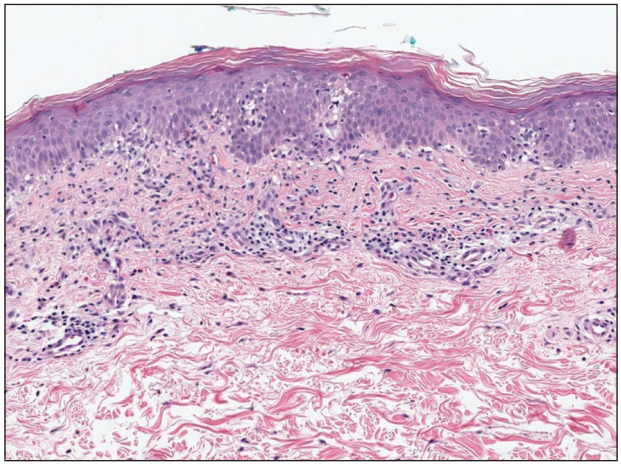


FIGURE 2. Punch biopsy revealed a perivascular bandlike infiltrate in the upper and mid dermis consisting of lymphocytes and occasional eosinophils (H&E, original magnification ×40). There were rare necrotic keratinocytes. In the epidermis there was mild spongiosis as well as lymphocytic exocytosis.

pseudopapuloerythroderma—and proposed that primary PEO is a diagnosis of exclusion. If no secondary association is found, they proposed 10 criteria for primary PEO: 5 major criteria include coalescing flat-topped papules, the deck-chair sign, pruritus, histopathologic exclusion of diseases such as CTCL, and a negative workup to exclude other causes.[3] In 2018, Maher et al[6] recommended workup to rule out cutaneous malignancy, including skin biopsy, flow cytometry, Sézary cell count, T-cell rearrangement, lactate dehydrogenase, and human T-cell lymphotropic virus 1. The 5 minor criteria proposed by Torchia et al[3] include age older than 55 years, male sex, eosinophilia, elevated IgE level, and lymphopenia. Our patient fulfilled all 5 major criteria and 3 minor criteria; eosinophilia and lymphopenia were absent.

Clinically, PEO has been associated with the deck-chair sign, a pattern of selective sparing of skinfolds, including axillary, inguinal, submammary, and other flexural areas. Although the deck-chair sign was originally considered pathognomonic for PEO, this clinical pattern also has been observed in other entities, such as angioimmunoblastic lymphoma, cutaneous Waldenström macroglobulinemia, and acanthosis nigricans.[5,8,9]

Specific characteristics of the rash and certain clinical symptoms may help to distinguish the deck-chair sign of PEO from its other causes. Although malignant acanthosis nigricans may spare the skinfolds, lesions have a classic velvety thickening and brownish hyperpigmentation, which is not characteristic of the reddish brown, flat-topped papules of PEO.[9] Pai et al[5] described a patient with parthenium dermatitis presenting with the deck-chair sign that developed years after repeated exposure to the allergen. Our patient did not have a history of repeated episodes of allergic contact dermatitis. In addition, areas of sparing may mimic the appearance of pityriasis rubra pilaris. As in our patient, those with PEO generally lack the follicular hyperkeratotic papules, palmoplantar keratoderma, widespread orange-red erythema, and characteristic histopathologic finding of hyperkeratosis with alternating orthokeratosis and parakeratosis, allowing these entities to be easily distinguished in most instances.[10]

Histopathologically, primary PEO shows a nonspecific spongiotic dermatitis–like pattern characterized by slight epidermal hyperplasia with spongiosis and a predominantly perivascular dermal infiltrate with lymphocytes, histiocytes, and eosinophils.[3] These histologic findings may at times show some overlap with CTCL, and therefore T-cell gene rearrangement and flow cytometry may be performed in those instances.[6]

Treatment includes the management of any underlying condition causing the papuloerythroderma.[3,6] There are no large clinical trials of treatment options for primary PEO due to its rarity. Topical or systemic corticosteroids remain the mainstay of treatment.[3] Alternative treatments used with variable success include phototherapy, interferon, etretinate, cyclosporine, and azathioprine.[11] Allegue et al[11] successfully used methotrexate to treat a patient with primary PEO and postulated that methotrexate may act through an immunosuppressive mechanism on activated T cells due to the involvement of helper T cells T_H2 and T_H22 in its pathogenesis.

Although the cutaneous manifestations of PEO may respond well to topical steroids, it is important to consider the possible presence of an underlying malignancy and other associated systemic conditions.

REFERENCES

1. Farthing CF, Staughton RC, Harper JI, et al. Papuloerythroderma—a further case with the 'deck chair sign.' *Dermatologica.* 1986;172:65-66.
2. Ofuji S, Furukawa F, Miyachi Y, et al. Papuloerythroderma. *Dermatologica.* 1984;169:125-130.
3. Torchia D, Miteva M, Hu S, et al. Papuloerythroderma 2009: two new cases and systematic review of the worldwide literature 25 years after its identification by Ofuji et al. *Dermatology.* 2010;220:311-320.
4. Ochi H, Ang CC. Novel association of a papuloerythroderma of Ofuji phenotype with dermatitis herpetiformis. *Int J Dermatol.* 2018;57:856-857.
5. Pai S, Shetty S, Rao R. Parthenium dermatitis with deck-chair sign. *JAMA Dermatol.* 2015;151:906-907.
6. Maher AM, Ward CE, Glassman S, et al. The importance of excluding cutaneous T-cell lymphomas in patients with a working diagnosis of papuloerythroderma of Ofuji: a case series. *Case Rep Dermatol.* 2018;10:46-54.
7. Grob JJ, Collet-Villette AM, Horchowski N, et al. Ofuji papuloerythroderma. report of a case with T cell skin lymphoma and discussion of the nature of this disease. *J Am Acad Dermatol.* 1989;20(5 pt 2):927-931.
8. Ferran M, Gallardo F, Baena V, et al. The 'deck chair sign' in specific cutaneous involvement by angioimmunoblastic T cell lymphoma. *Dermatology.* 2006;213:50-52.
9. Murao K, Sadamoto Y, Kubo Y, et al. Generalized malignant acanthosis nigricans with "deck-chair sign." *Int J Dermatol.* 2013;52:377-378.
10. Regina G, Paramita L, Radiono S, et al. Papuloerythroderma of Ofuji in Indonesia: the first case report. *JDVI.* 2016;1:93-98.
11. Allegue F, Fachal C, Gonzalez-Vilas D, et al. Papuloerythroderma of Ofuji successfully treated with methotrexate. *Dermatol Ther.* 2018;31:E12638.