



Diffuse Normolipemic Plane Xanthomatosis with Immunoglobulin-Lambda Light-Chain Deposition in a Patient with Multiple Myeloma

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Dear Editor:

Altman and Winkelmann¹ first described diffuse normolipemic plane xanthomatosis (DNPX) in 1962. Patients with DNPX usually present with yellowish plaques that characteristically appear on the periorbital region, neck, and flexural folds; plasma lipid levels are normal^{1,2}. Relationships with underlying hematological diseases, particularly multiple myeloma and other types of monoclonal gammopathy, have frequently been suggested^{3,4}.

A 69-year-old woman presented with a 10-year history of slowly increasing yellow skin lesions on the whole body. She showed symmetrically distributed well-demarcated yellow plaques in both periorbital areas and multiple yellow

low patches and papules in the flexural area, including the axilla and the inguinal region (Fig. 1). The patient did not complain of any systemic symptoms. Laboratory examinations revealed iron deficiency anemia and hypoalbuminemia. Plasma lipid and triglyceride levels were within normal ranges. Urinalysis revealed overt proteinuria and hematuria. Serum electrophoresis showed a monoclonal peak in the gamma region and immunoglobulin G-lambda monoclonal gammopathy. An echocardiogram showed congestive heart failure and a supracardinal lymph-node biopsy revealed amyloid deposition. Renal biopsy demonstrated amyloid light chain-related amyloidosis. Bone marrow biopsy revealed aggregates of plasma cells with lambda

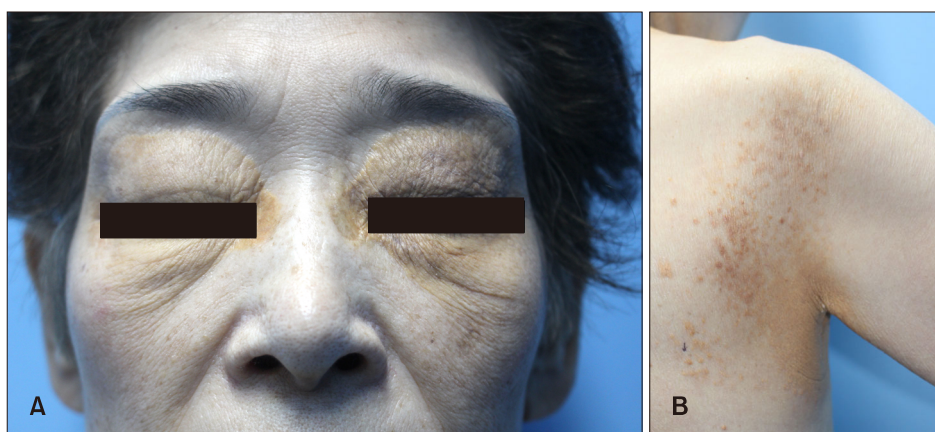


Fig. 1. (A) Symmetrically distributed yellow plaques on both periorbital areas. (B) Yellow patches, papules on the axilla. We received the patient's consent form about publishing all photographic materials.

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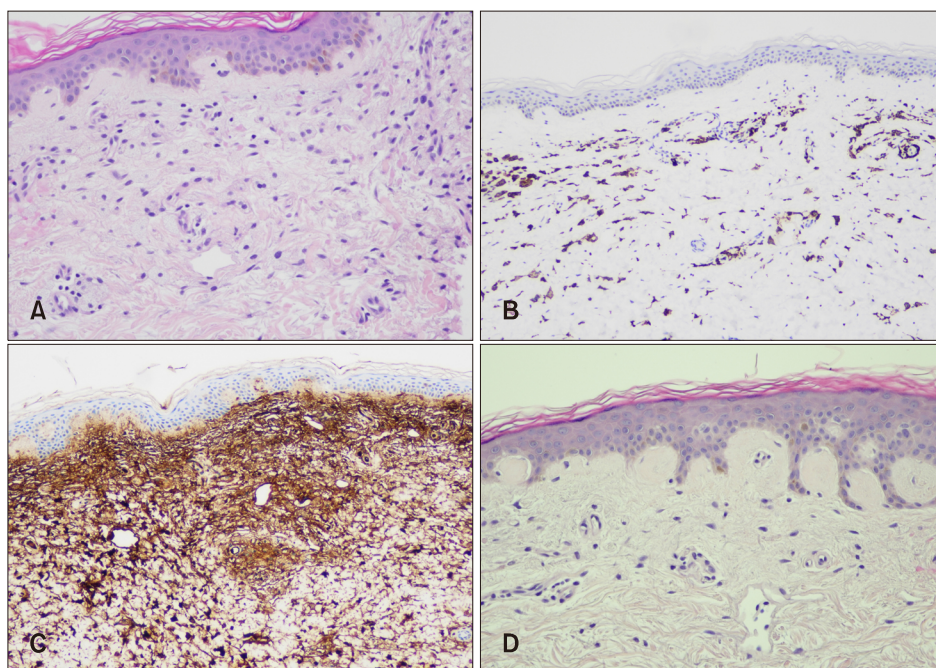


Fig. 2. (A) Scattered foamy histiocytes are observed in the upper and mid dermis, with an admixture of histiocytes and lymphocytes (H&E, $\times 200$). (B) Immunohistochemical staining was diffusely positive for CD68 (CD68, $\times 100$). (C) Dense staining throughout the entire dermis for the immunoglobulin-lambda light chain is noted (lambda chain, $\times 100$). (D) Globular deposits of homogenous, faintly eosinophilic materials in the papillary dermis (H&E, $\times 200$).

da monoclonality. Based on these findings, the patient was diagnosed with multiple myeloma. A skin biopsy from her axilla showed scattered foamy histiocytes in the upper and mid dermis, with an admixture of histiocytes and lymphocytes (Fig. 2A). Immunohistochemical staining was diffusely positive for CD68 (Fig. 2B). Dense staining throughout the entire dermis for the immunoglobulin-lambda light chain was also observed (Fig. 2C). These findings were consistent with DNPX. The papillary dermis contained globular deposits of homogenous, faintly eosinophilic materials (Fig. 2D). However, they showed no definite affinity to congo red.

The pathogenesis of DNPX remains unclear. The previously suggested pathogenesis of DNPX is that monoclonal immunoglobulins bind to circulating low-density lipoproteins in plasma and form complexes around vessels in the skin, where they are phagocytosed by macrophages, which finally appear as foam cells^{3,4}. Several cases of DNPX associated with monoclonal gammopathy have been reported to date³⁻⁵, but this report is the first to describe dense lambda immunoglobulin deposition with infiltration of histiocytes in the entire dermis, as revealed by skin biopsy. Furthermore, we cautiously suggest the coexistence of cutaneous amyloidosis, which is another unique finding of this report, based on the observation of globular deposits of eosinophilic materials in the papillary dermis on histological examination. Cutaneous amyloid deposition may exist in this patient, considering that it is one of the symptoms of multiple myeloma, and the patient also had accompanying cardiac and renal amyloidosis. Although the

globular deposits show no definite affinity to congo red, inadequate tissue quantity and staining might have caused a false negative result or decreased sensitivity.

Although not all xanthomas are associated with systemic diseases, DNPX may appear as the first symptom of underlying malignant hematological or lymphoproliferative diseases⁵. Therefore, in patients with DNPX, skin biopsy and hematological evaluation should be performed, and close follow-up with periodic laboratory testing is recommended.

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CONFLICTS OF INTEREST

The authors have nothing to disclose.

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Docetaxel-Aggravated Psoriasis

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Dear Editor:

A 78-year-old man with a 50-year history of psoriasis presented with generalized erythematous scaly plaques involving the scalp and face (Fig. 1). Over 5 years, the psoriasis had been confined to the hands and feet and controlled using topical agents alone. Three days prior to emergence of the rash, he reported being administered the first dose of docetaxel 90 mg as treatment for prostate cancer. No aggravation of the psoriasis had been observed throughout past 4 years of treatment with antiandrogens and radiotherapy.

At the present visit, the psoriasis area severity index score was 10.4, and the percentage of body surface area involved was 20%. Given his medical history and clinical features, we concluded that the preexisting psoriasis had exacerbated due to the newly introduced docetaxel treatment. The patient was treated with acitretin at a dose of 20 mg daily and topical calcipotriol/betamethasone dipro-

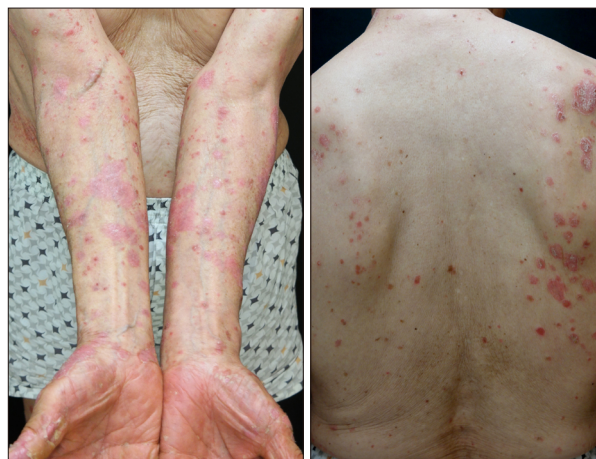


Fig. 1. Scaly erythematous plaques that appeared 3 days after the administration of docetaxel. We received the patient's consent form about publishing all photographic materials.

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