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Diffuse normolipemic plane xanthoma associated with monoclonal gammopathy

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ABSTRACT Diffuse normolipemic plane xanthoma (DNPX) was first described by Altman and Winkelmann in 1962. It is a rare and non-inherited form of xanthomatosis. Clinically, the dermatosis is characterized by the presence of symmetric yellowish-orange plaques that favor the neck, upper trunk, flexural folds and periorbital region. It has been recognized to be associated with hematological diseases, especially with multiple myeloma and monoclonal gammopathy. We present a patient with diffuse plane xanthoma, normal lipid level, and monoclonal gammopathy.

Case report

An 85-year-old man presented with an 8-month history of slowly increasing diffuse yellow skin lesions on the torso and upper arms. He had been unaware of the discoloration until his internist noted it and referred him to dermatology. The patient had been followed by a hematologist for paraproteinemia that had not been treated.

The examination showed well-demarcated yellow patches and plaques covering large portions of his upper trunk with some islands of sparing (Figure 1, 2). The dermoscopic evaluation showed a reticular pattern of yellow amorphous homogeneous structures with overlying branched and linear vessels (Figure 3). A skin biopsy showed scattered foamy histiocytes within the reticular dermis, which were consistent

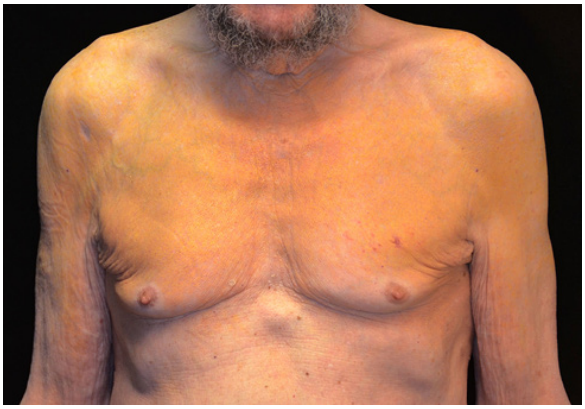


Figure 1. Clinical view shows well-demarcated yellow patches and plaques covering large portions of the upper chest and shoulder with some islands of sparing. [Copyright: ©2015 Cohen et al.]

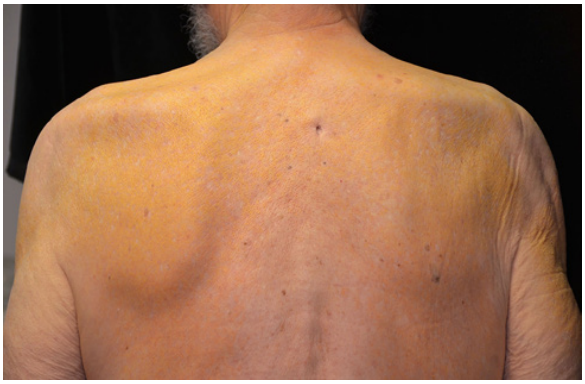
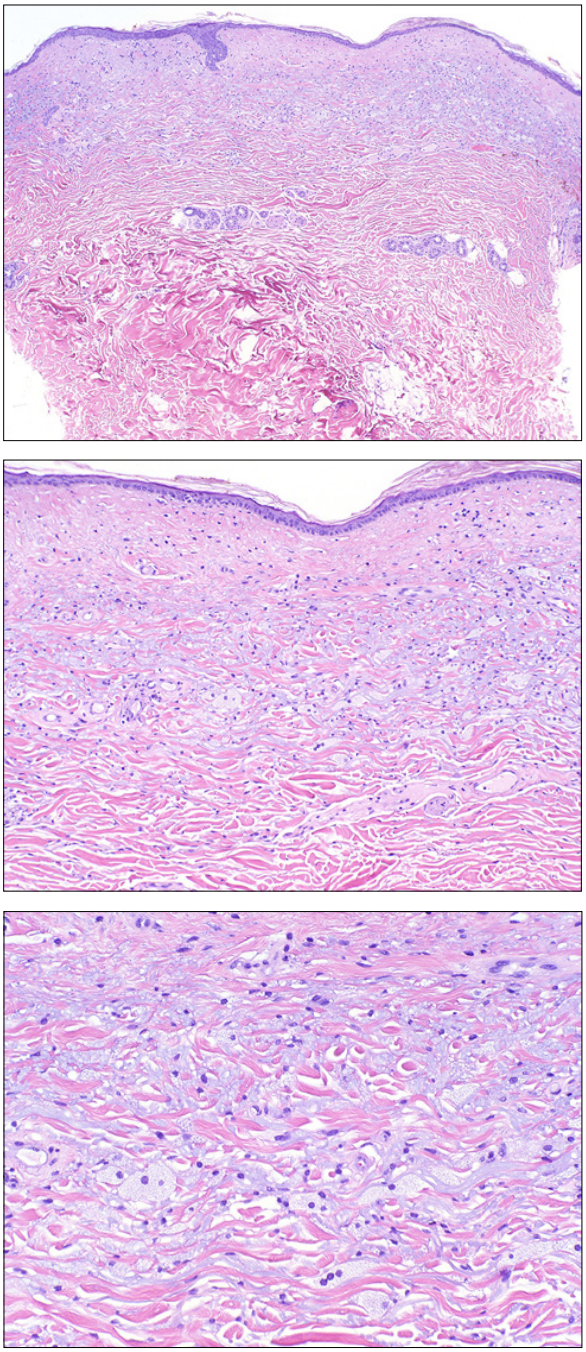


Figure 2. Well-demarcated yellow plaques on the upper back. [Copyright: ©2015 Cohen et al.]

Figure 3. Dermoscopic view shows a reticular pattern of yellow amorphous uniformed granules with overlying vascular structures. [Copyright: ©2015 Cohen et al.]



Figures 4, 5, 6. Histology views show scattered foamy histiocytes within the papillary dermis with a normal overlying epidermis, which were consistent with a plane xanthoma. [Copyright: ©2015 Cohen et al.]

with a plane xanthoma (Figures 4, 5, 6). A complete blood count, comprehensive metabolic panel, fasting lipid panel were within normal range. The serum protein electrophoresis showed elevated IgG at 3780. There was an M-band present in the gamma region.

Discussion

Diffuse normolipemic plane xanthoma (DNPX) was first described by Altman and Winkelmann in 1962 [1]. It is an uncommon subtype of non-Langerhans histiocytosis [15]. DNPX is characterized by xanthelasma palpebrarum; diffuse plane xanthoma of the head, neck, trunk, and extremities; and normal plasma lipid levels [2,4]. Xanthelasma typically appears first, followed by involvement of the lateral parts of the neck and upper trunk [1]. Clinically, the dermatosis is characterized by the presence of symmetric yellowish-orange plaques that favor the neck, upper trunk, flexural folds and periorbital region [1,2]. Histologically, foam cells (macrophages that have engulfed lipid droplets) and variable numbers of Touton giant cells, lymphocytes, and foamy histiocytes can be seen [3].

While not all cutaneous xanthomas are associated with systemic diseases, DNPX has been associated with systemic

diseases, particularly multiple myeloma and monoclonal gammopathy [2,3]. However, other malignant hematological or lymphoproliferative disorders associated with DNPX include acute monoblastic leukemia, chronic myelomonocytic leukemia, chronic myloid leukemia, chronic lymphatic leukemia, non-Hodgkin's lymphoma, adult T-cell lymphoma/leukemia, Sezary syndrome, Waldenstrom's macroglobulinemia, cryoglobulineimia and Castleman's disease [2,3,5-12].

The pathogenesis of DNPX has not been fully elucidated, however, in gammopathy-associated DNPX, monoclonal IgG is thought to bind to circulating LDL, rendering the antibody–LDL complex more susceptible to phagocytosis by macrophages [13]. DNPX can precede such disorders by several years; therefore close follow-up with periodic laboratory tests for myeloproliferative disorders should be performed [4].

There are currently several treatment options available. In patients with limited involvement, the individual lesions can be excised. Other options include chemabrasion, dermabrasion, and ablative laser therapy. The erbium:YAG laser has been used successfully to treat facial xanthomas in one patient [14].

In this patient, currently there are no associated systemic symptoms. His hematologist was informed of the diagnosis. The patient will be evaluated for potential underlying malignancies. Moreover, this case shows that dermatological lesions can be the first manifestation of important hematological diseases and so physicians should be familiarized with this rare entity.

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