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Case Report

Diffuse Normolipemic Plane Xanthoma (DNPX) of the Neck without Xanthelasma Palpebrum

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Abstract

Diffuse normolipemic plane xanthoma (DNPX) is an uncommon subtype of non-Langerhans histiocytosis. DNPX is characterised by xanthelasma palpebrarum, diffuse plane xanthoma of the head, neck, trunk, or extremities, and normal plasma lipid levels. The neck is the most common site. We report about a 62-year-old female Caucasian patient, who developed an asymptomatic fine wrinkling and loose skin on the neck and décolleté about three years ago. The skin colour became yellowish. Xanthelasma was absent. Histopathology of a skin biopsy confirmed the diagnosis of DNPX. The patient had a medical history of chronic myeloblastic leukaemia. No other laboratory abnormalities were found. Laser treatment was offered but opposed by the patient.

Introduction

Diffuse normolipemic plane xanthoma (DNPX) was first described by Altman and Winkelmann in 1962 [1]. It is now considered as an uncommon subtype of non - Langerhans histiocytosis [2].

DNPX is characterized by xanthelasma palpebrarum, diffuse plane xanthoma of the head, neck, trunk, or extremities, and normal plasma lipid levels. The neck is the most common site [1][3]. Xanthelasma palpebrarum usually appears first [1].

The clinical presentation is characterised by the presence of symmetric, asymptomatic, yellowish-orange plaques [1][2]. Oral lesions are extremely rare [3].

In histology, foam cells (macrophages), and variable numbers of Touton giant cells, lymphocytes, and foamy histiocytes are present; sometimes only foam cells can be seen [4][5].

DNPX has been associated with systemic diseases, particularly multiple myeloma and monoclonal gammopathy [4][5][6]. In other cases, malignant haematological or lymphoproliferative disorders have been observed [7][8].

Case report

A 62-year-old female Caucasian patient developed an asymptomatic fine wrinkling and loose skin on the neck and décolleté about three years ago.

The skin colour became yellowish (Fig. 1).

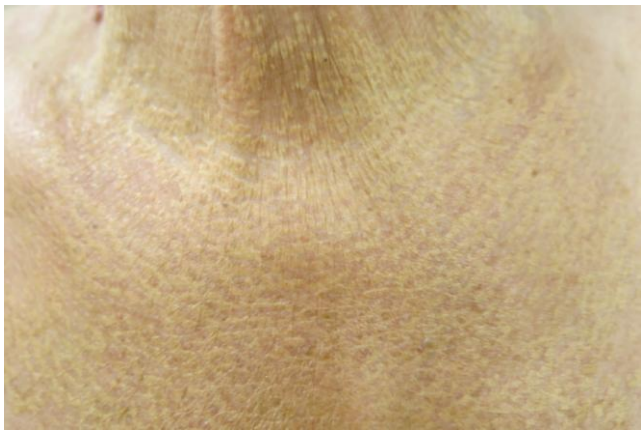

Figure 1: Diffuse plane yellowish plaques on neck and décolleté

No other body areas were involved. Her medical history was remarkable for chronic myeloblastic leukaemia without chemotherapy. She did not take any medical drugs. The family history was negative for skin diseases. Laboratory investigations of metabolic abnormalities remained unremarkable. A skin biopsy revealed an atrophic epidermis and a massive infiltration of the upper and mid-dermis by CD68 positive macrophages including foam cells. Here, elastic fibres were somewhat reduced. No calcifications were noted (Fig. 2).

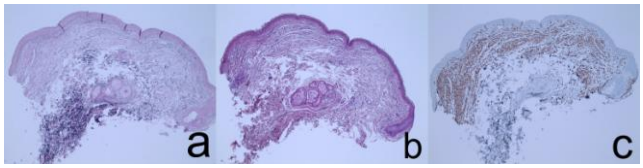

Figure 2: Histopathology of diffuse normolipemic plane xanthoma (x 4). (a) Elastica stain, (b) hematoxylin-eosin, and (c) immunoperoxidase for CD68

The diagnosis of DNPX was confirmed. Ablative laser therapy was mentioned, but treatment was not warranted.

Discussion

DNXP is part of the Langerhans cell histiocytes-spectrum [9]. No standardised treatment is available yet. However, cladribine (2-chlorodeoxyadenosine) is a candidate drug since it is particularly metabolised, phosphorylated and concentrated in lymphocytes, macrophages/ histocytes and Langerhans cells. The active compound is 2-chloroadenosine triphosphate. Cladribine has been used successfully in Langerhans cell histiocytosis of different types including plane xanthoma [10]. There are case reports on regression of DNPX during treatment (of associated disorders) with cyclosporine A [11] or bexarotene [12]. DNPX can be treated by ablative lasers such as erbium-YAG laser [13][14].

DNXP itself does not cause significant health problems although it can be esthetically annoying. Of greater importance is the fact, that DNXP has been observed in association with monoclonal gammopathy, monoclonal gammopathy of unknown significance (MGUS) and plasmacytoma [4][5][6][7][8][14][15][16]. In our case, chronic myeloblastic leukaemia was evident. Under this view, DNXP may have a marker function for unknown myeloproliferative disorders and dermatologists should be familiar with this uncommon entity.

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