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Case Study

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PLANE NORMOLIPEMIC XANTHOMA LOCALISED OVER THE NECK: A CASE REPORT

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ABSTRACT

Diffuse plane xanthomas manifest as symmetrical, asymptomatic, yellow-to-brown patches or plaques involving the head, eyelids, neck, chest, trunk, extremities, flexural areas and scars. Diffuse normolipaemic plane xanthomatosis represents an uncommon cutaneous disease consisting of xanthelasma, diffuse plane xanthomas and normal plasma lipid levels. Here we present a case of diffuse normolipemic plane xanthoma localized to the neck.

KEYWORDS: Diffuse plane xanthoma, normolipemic, foamy histiocytes, systemic disease.

INTRODUCTION

Diffuse plane xanthoma (DPX) was first described in 1962.[1] It manifests as symmetrically distributed, asymptomatic, yellow-to-brown patches or plaques which may involve the head, eyelids, neck, chest, trunk, extremities, flexural areas as well as scars.[2,3] Diffuse normolipaemic plane xanthomatosis represents an uncommon cutaneous disease consisting of xanthelasma, diffuse plane xanthomas and normal plasma lipid levels. Xanthelasma typically appear first followed by involvement of the lateral parts of the neck and upper trunk.[4,5]

Here we present a case of diffuse plane xanthoma localized to the neck associated with normal serum lipid levels.

CASE REPORT

A 60-year-old woman presented with a 2-month history of yellowish colored skin lesions over the neck gradually increasing in size and number. The lesions were asymptomatic except for the occasional itching. The examination showed involvement of the lateral and posterior aspect of the neck in the form of multiple, discrete to coalescent yellowish papules and plaques arranged randomly as well as in linear and reticulate patterns at places. The rest of the physical examination was normal except for xanthelasma palpabrarum. Skin biopsy showed thin and flat epidermis with upper reticular dermis showing dense, diffuse infiltrate of sheets of foamy histiocytes without any accompanying inflammatory infiltrate. The patient was a known type II diabetic under treatment with controlled blood sugar levels. There was no significant family history. A complete blood count, comprehensive metabolic panel and fasting lipid panel were within normal range.

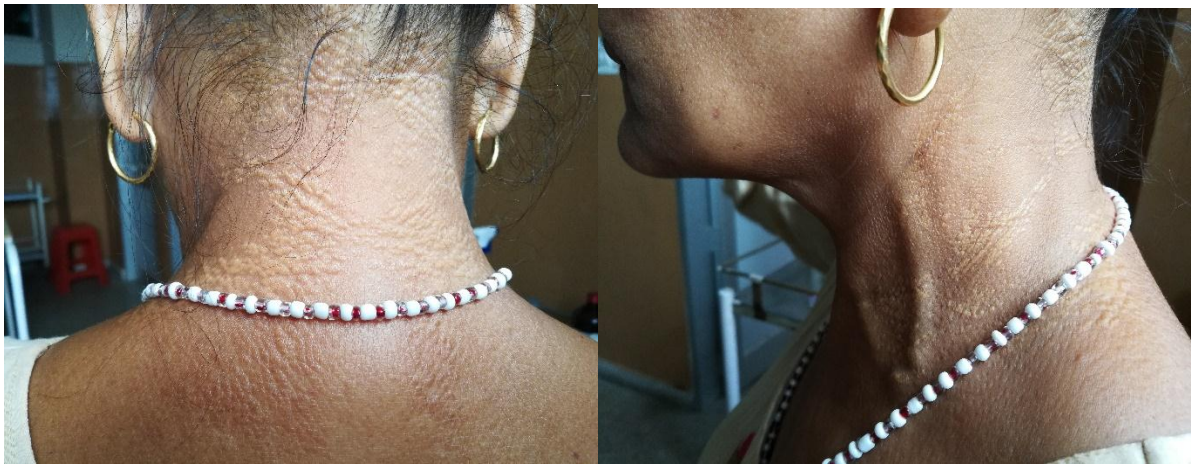

Figure 1,2: Showing yellowish papules and plaques over the neck.

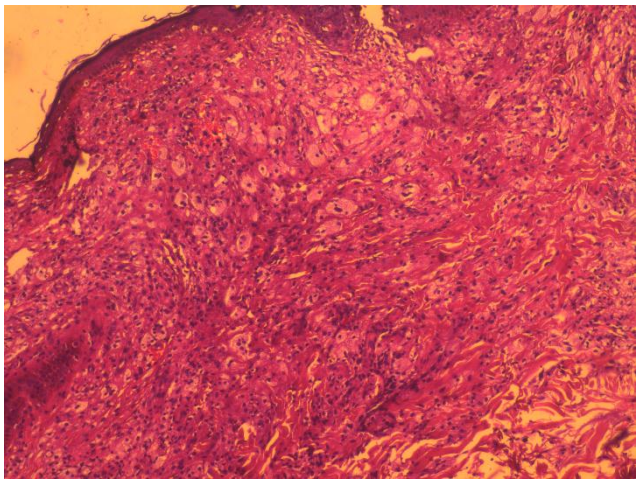

Figure 3: HPE showing xanthoma cells with vacuolated cytoplasm in the dermis (10x).

DISCUSSION

Rayer first introduced cutaneous xanthomas in the medical field in 1835. Various forms of xanthomas were first described by Addison & Gull in 1851.[2] Diffuse normolipemic plane xanthoma (DPX) was described by Altman and Winkelmann in 1962.[1,3,4] This disorder mostly affects the adults, although a few cases have been noted in children.[2] Diffuse plane xanthomatosis exhibits itself as symmetrical patches and plaque of yellowish orange lesions involving the eyelids, neck, upper trunk, buttocks and flexures.[2,3,4] On histopathological examination, foam cells (macrophages that have engulfed lipid droplets) and variable numbers of Touton giant cells, lymphocytes, and foamy histiocytes can be seen.[4,5,6]

Plane xanthomas have been divided into two groups. Group 1 is associated with increased serum lipid levels due to familial hyperlipidemia. Group 2 has either normal or slightly increased lipid levels without any associated family history. Group 2 can further be sub-divided into idiopathic, associated with some underlying disease and associated with abnormalities of the structure or content of lipoproteins. In patients without any associated systemic disease, the course is chronic and benign.[6,7] In a study conducted by Marcoval *et al* to determine the incidence of associated disorders in patients with diffuse plane xanthomas, it was found that out of eight patients, only three had an underlying disease (benign monoclonal gammopathy in 2 and chronic myelomonocytic leukemia in 1). It was suggested that the incidence of underlying disease association with diffuse plane xanthomas is lower than expected.[8] However, the xanthoma may precede the clinical symptoms of the systemic disease by years therefore, periodic follow up is suggested.[4,5,6,8] In our patient, no underlying disease could be identified. However, the patient was lost to follow-up.

The pathogenesis of diffuse plane xanthomas has not been clearly elucidated. Several theories have been suggested. Some authors suggest that paraprotein-lipoprotein complexes get deposited in the skin which are subsequently phagocytosed by the macrophages. Others suggest that leukemic cells infiltrate the skin with subsequent xanthomatization. Some consider diffuse plane xanthomata to be a form of non-X histiocytosis.[4,5,6] Abnormalities of the complement have also been described in a few cases.[3]

The plaque-type morphology and distribution of plane xanthomas create a differential diagnosis that includes

infiltrating disorders like amylodiosis, sarcoidosis and histiocytosis. In our patient, the lesions were localized to the neck. So the possibility of solar elastosis, age- related dermal fibroelastolysis and sporadic late onset pseudoxanthoma elasticum were also considered. In this case, the short history of two months suggest that the disease might be in the evolving stage with localization of the early lesions to the neck.

There are several treatment options available. In patients with limited involvement, the lesions can be excised. Other options include chemabrasion, dermabrasion and ablative laser therapy.[4,5] The erbium: YAG laser has been used successfully to treat facial xanthomas in one patient.[7]

To conclude, diffuse plane normolipemic xanthomas are rare and non-inherited disease which may be associated with systemic diseases. The skin lesions can precede the systemic disease by several years. So follow up is necessary even if the patient seems to have no underlying disease.

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