

A Case of Plane Xanthoma Associated with Type IV Hyperlipoproteinemia

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A case of plane xanthoma arising in type IV hyperlipoproteinemia in a 53-year old woman is reported. The patient had well-defined yellowish to orange colored slightly elevated plaques on both upper eyelids, cheeks, neck, and upper chest. The laboratory findings showed increased serum triglyceride on lipid profile, and widening of pre-beta band on electrophoresis lipoprotein. She had a fatty liver, mild hepatomegaly, but other systemic diseases such as multiple myeloma, leukemia and lymphoma were not found.

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Plane xanthoma described by Lynch & Winkelmann¹, is yellow to yellowish-brown flat patches or slightly elevated plaques. When generalized, the disease is frequently associated with multiple myeloma^{1,2} or other reticulo-endothelial malignancies^{3,5}. Although assigned to the group of normolipemic xanthomata⁶⁻¹³, occasional cases with concomitant hyperlipoproteinemia type II or IV^{15,16} have been observed. We report herein a typical case of plane xanthoma on face, neck and anterior chest, associated with type IV hyperlipoproteinemia in a 53-year old woman, who had no systemic diseases such as multiple myeloma or other reticulo-endothelial malignancies.

REPORT OF A CASE

A 53-year-old woman visited our department because of xanthelasmata on both upper eyelids (Fig. 1) for 2 years and well-defined yellowish to orange discoloration of the skin for about 6

months. Her past medical and family histories were not contributory. Cutaneous examination showed well defined yellowish to orange colored slightly elevated plaques on both cheeks, neck and upper chest (Fig. 2). Laboratory studies, including complete blood cell count, LFT, RFT, chest PA, urinalysis, serologic test for syphilis, electrocardiogram, stool examination, fasting blood sugar and cryoglobulin were within normal limits or negative. But plasma triglyceride level measured after an overnight fast was 483 mg/dl (normal range, 76 to 172mg/dl), cholesterol 235mg/dl (normal range, 130 to 250mg/dl), HDL-cholesterol 34.5mg/dl (normal range, 42.3 to 70.7mg/dl). Electrophoresis of lipoproteins revealed that α -lipoprotein was 17.0% (normal range, 15 to 40%), pre- β lipoprotein 32.0% (normal range, 5 to 15%), β lipoprotein 28.0% (normal range, 40 to 60%), chylomicron 0% (normal range, 0 to 20%). The pattern of this lipoprotein is typical for hyperlipoproteinemia type IV according to the WHO classification¹⁷. Plasma stored overnight at 4°C showed a clear normal appearance. Quantitative analysis of the serum immunoglobulins and immunoelectrophoretic study were within normal limits. Abdominal ultrasonographic examination showed fatty liver and mild hepatomegaly. Biopsy specimens from upper chest

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Fig. 1. Well defined slightly elevated yellowish plaques on both eyelids.

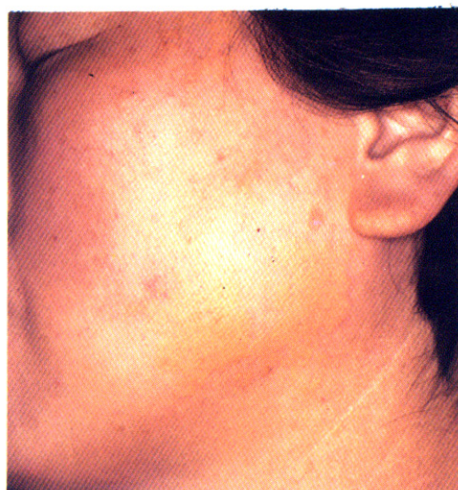


Fig. 2. Relatively well defined yellow to orange colored discoloration on left submandibular and temple regions.

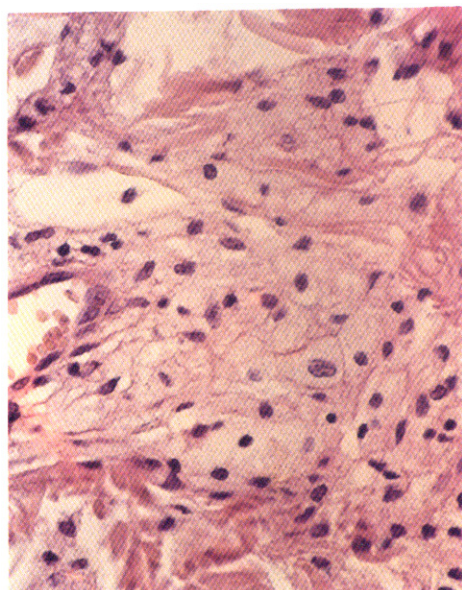


Fig. 3. Histologic section of biopsy specimen from region of upper chest shows large clusters of foam cells (H&E, ×400).

and left cheek showed large clusters of foam cells in papillary dermis but Touton giant cells were not observed (Fig. 3). Oil red O stain showed many lipid droplets in the dermis (Fig. 4). Xanthelasma lesions of both upper eyelids were treated with electrodesiccation, and oral niacin was started for the control of type IV hyperlipoproteinemia.

DISCUSSION

Plane xanthomas occur in several areas of the body, but the most commonly encountered lesion, x-

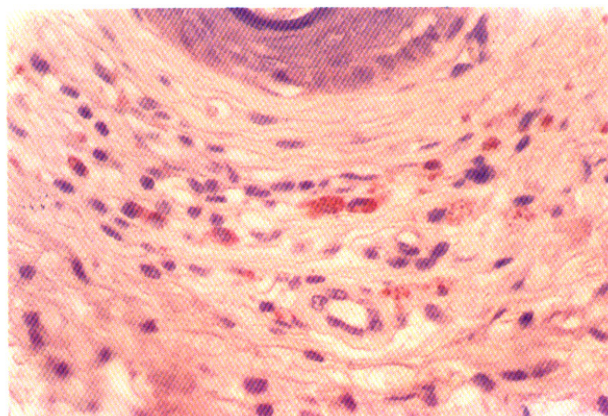


Fig. 4. Reddish lipid droplets are seen in the upper dermis (Oil red O stain, ×400).

anthelasma, occurs on the eyelids. A second type of plane xanthoma has been termed xanthoma striatum palmare, which appears as flat, yellow to orange, linear lesions in the crease of the palms and fingers. A third form of plane xanthoma, generalized plane xanthoma is extensive, yellow to orange infiltrative lesions that diffusely involve the face, neck and upper trunk and arms. In many instances, this phenomenon is a direct result of hyperlipoproteinemia¹⁴⁻¹⁶, which may be familial or acquired. However, some patients have no disorder of lipoprotein metabolism. Generalized plane xan-

thoma can be categorized into two large groups¹⁹. Group 1 consists of generalized plane xanthoma occurring with other xanthomatous processes. The serum is always hyperlipemic, and the increased lipids always can be explained on the basis of a familial pattern or of biliary cirrhosis. Group 2 includes generalized plane xanthoma occurring normolipemia, but lipid levels may be elevated, and there is neither familial pattern nor significant liver disease to explain the rise. This second group can be further subdivided into (a) xanthoma not associated with any overt systemic disease (essential), (b) xanthoma associated with disease of the reticuloendothelial system, and (c) xanthoma associated with miscellaneous disease (possible coincidental association).

Our case had type IV hyperlipoproteinemia, with no overt reticuloendothelial system disease, being compatible with the group 1 category mentioned above. In our case, some hepatopathy such as fatty liver and mild hepatomegaly were noted. But we did not observe biliary cirrhosis. Reticuloendothelial diseases associated with xanthoma planum are multiple myeloma^{1,2}, chronic granulocytic leukemia⁴, chronic myeloid leukemia⁵, cryoglobulinemia³, Waldenström's macroglobulinemia, lymphoma, and benign monoclonal gammopathy. There was no evidence of reticuloendothelial diseases in our case.

Much of the lipid constituent of xanthomas is derived from the plasma²⁰. In many instances the occurrence of skin xanthoma is related directly to the degree of elevation of lipoproteins. Further, correlative lipid analytic and electron microscopic studies on experimental and human xanthomas suggest that lipoprotein permeates the walls of dermal capillaries and is then phagocytized by dermal histiocytes that evolve into foamy cells²¹.

Type IV hyperlipoproteinemia shows an autosomal dominant inheritance pattern and elevation of triglycerides is present. Plasma stored cold overnight is usually turbid in appearance due to a high concentration of VLDL²². But Plasma showed a clear normal appearance in our case. This type represents diabetes mellitus, atherosclerosis and hepatomegaly. Mild hepatomegaly was noted in our case.

Plane xanthoma seems to lack a specific histopathologic picture. Bazex *et al.*²³ claimed that no giant cells of either the Touton or the foreign

body type occur in these lesions, but Winkelmann and Welborn¹², on the contrary, considered such giant cells to be a characteristic feature. Our case did not show giant cells.

There is no consistently effective treatment of plane xanthoma. We initially treated niacin, hypolipidemics for type IV hyperlipoproteinemia. Some xanthoma lesions were treated with electrodesiccation, and others with trichloroacetic acid peeling.

In conclusion, our case showed clinical and histopathological findings of plane xanthomas, which occurred on both upper eyelids, cheeks, neck and upper chest in a 53-year old female, who had no familial pattern or biliary cirrhosis, being nearly compatible with group 1 category mentioned above, associated with type IV hyperlipoproteinemia.

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