

CASE REPORT

Palmar xanthoma: Mirror to pernicious underlying condition

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ABSTRACT

Background: Xanthomas are disorders of lipid metabolism, characterized by the deposition of lipid-laden macrophages in the subcutaneous tissue. Clinically these present as yellowish, asymptomatic plaques. Various types with underlying defect in one or more types of lipoprotein metabolism have been described. Palmar Xanthoma, also known as Xanthoma Striatum Palmare (XSP), is a rare type of Xanthomas that is characterized by the development of orange-yellow linear plaques along the creases. The basic underlying metabolic defect is type III hyperlipoproteinemias, which is associated with high risk of cardiovascular diseases.

Main Observation: A female patient in her thirties presented to the Dermatology department with development asymptomatic linear yellowish, slightly elevated, plaques running along the palmar creases of both the hands. Patient did not have any history of hypertension, hyperlipidemia or cardiac disease in the past. Investigations revealed dyslipidemias i.e. high cholesterol and triglyceride levels supporting the clinical diagnosis of palmar xanthoma.

Conclusion: Skin manifestations as for many other metabolic diseases are a key feature in case of dyslipidemias as well. Dermatologists are generally the first to diagnose dyslipidemias in apparently healthy individuals especially in case of Palmar Xanthomas. Therefore, the dermatologists should be vigilant in such cases. Besides managing the cutaneous lesions, they must get these patients thoroughly investigated and refer them to physicians for further management. Timely intervention and treatment can thus prevent serious morbidity, as there is potential risk of cardiovascular disease in these subjects.

KEY WORDS: Hypercholesterolemia, Hyperlipidemia, Palmar Xanthoma, Xanthomas

INTRODUCTION

Xanthomas are disorders of lipid metabolism, characterized by the deposition of lipid-laden macrophages in the subcutaneous tissue, imparting a yellowish hue to the affected skin. Seven types of xanthomas have been identified. Xanthomas at times are the presenting features, which help diagnose the basic underlying pathology and lead patient to seek proper treatment. This is especially true for Palmar Xanthoma, where the lesions are generally the only clinical feature of an underlying type III

hyperlipoproteinemia, which is associated with a high risk of cardiovascular diseases.

CASE REPORT

A 36 year old, North-Indian female presented with the complaint of development of asymptomatic, yellowish, mildly elevated linear plaques on bilateral palms, running along the palmar creases, especially on the digits. These were present for last eight months. (Fig. 1) The lesions were gradually increasing in number and size. The patient did not any seek consultation

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Fig. 1 Clinical picture showing linear, yellowish plaques along the palmar creases of the patient.

prior to this. The main concern of the patient for consultation was cosmetic disfigurement. The patient was examined to look for similar lesions elsewhere on the body but none was found. Even the eyelids did not have any yellowish plaques. The history did not reveal any medical ailment in the past, which included no history of hypertension, hyperlipidemia or cardiac disease. The patient was moderately built. The patient however, gave a family history of coronary artery disease in the family on paternal side. Based on these findings patient was therefore clinically diagnosed as a case of palmar xanthoma.

Patient was then evaluated to look for the underlying etiology. Blood tests including lipid profile, thyroid profile and liver function tests were done initially. Fasting lipid profile of the patient showed hypercholesterolemia (447mg/dl) and hypertriglyceridemia (562mg/dl). These findings strongly supported the clinical diagnosis of palmar xanthoma. Thyroids profile and the liver function tests were however within normal limits. The patient was then referred to a physician for further investigations such as high-density lipoproteins and cardiovascular evaluation. The initial treatment in the form of

lipid lowering drugs was started. The patient is currently under the follow-up of her physician.

DISCUSSION

The term Xanthoma is derived from the Greek word “ xanthos” which means yellow. Xanthomas are clinically evident as yellowish asymptomatic plaques and histologically these are characterized by presence of abundant foamy macrophages i.e. the accumulation of the lipids in the macrophages. Xanthomas develop mainly due to leakage of lipids from the vessels into the surrounding tissue. Macrophages in the tissues phagocytize these lipids and hence form the “foamy macrophages”.¹

Practically all types of hyperlipidemias lead to development of xanthomas. Seven types of xanthomas have been described so far namely, tendon xanthoma, xanthelasma palpebrarum, eruptive xanthomata, tuberous xanthomata, planar xanthomata, xanthoma disseminatum and verruciform xanthoma.² Different types of hyperlipidemias are responsible for each variety and these are outlined in table 1.

Table 1 Etiology of different types of Xanthomas

Type of Xanthoma	Type of Hyperlipoproteinemia
Tendon xanthomata	Type II
Xanthelasma palpebrarum	Any type
Tuberous xanthomata	Type II and III
Eruptive xanthomata	Type I, IV, V
Planar xanthomata	Type III

Out of these, xanthelasma palpebrarum is the most common type. Xanthelasma palpebrarum, xanthoma disseminatum and verruciform xanthoma are known to occur even in individuals with a normal lipid profile. Planar xanthomas present as either macular or slightly raised le-

sions. These can occur anywhere on the body. Even generalized xanthomas covering large areas affecting the face and neck have been described. Another variety known is corneal arcus. In this type, there is deposition of cholesterol esters in the peripheral cornea. It is also known to occur with normal lipid levels. When associated with hypercholesterolaemia, it does not respond to lipid lowering drugs.

Palmar Xanthomas typically appear as orange-yellow plaques along the palmar creases. These are commonly referred to as Xanthoma Striatum Palmare (XSP). These can also be present along the wrist creases. Though a rare form, it is pathognomic of type III hyperlipoproteinemia and has propensity for development of atherosclerosis.

Type III hyperlipoproteinemia is also known as “dysbetalipoproteinemia”, “remnant hyperlipidemia” and “broad beta disease”. In this type, there is deposition of beta-VLDL (very low density lipoproteins), clinically evident as increased plasma triglycerides and cholesterol levels.³ This is a genetically influenced form of hyperlipidemia, predisposing the affected individuals to a higher risk of cardiovascular diseases. Mutations in the apoprotein E are responsible for its defective binding to beta-VLDL resulting in the broad beta disease. Especially those homozygous for apoprotein E2 are affected.²

The suggested hypothesis for the development of XSP as proposed by Walton *et al.*, is that minor trauma to palms due to pressure, leads to increased vascular permeability, which in turn causes increased leakage of lipids and their deposition in the skin resulting in yellowish plaques.⁴

Apart from type III hyperlipoproteinemia, palmar xanthomas can be associated with pancreatic insufficiency and multiple myeloma, and primary biliary cirrhosis. In case of deranged liver function tests, it is advised to keep primary biliary cirrhosis as a differential diagnosis. XSP is known to be associated with diabetes mellitus and coronary heart disease in young individuals.

Management mainly relies on life style modifications and administration of anti-hyperlipidemic drugs, to which XSP responds well. If the lesions are large hindering patients' daily routine activity or are a cosmetic concern for the patient, these can be treated with either electrical cautery or chemical cautery. Biopsy can be undertaken to confirm the diagnosis where numerous foamy macrophages can be seen.

XSP can be an initial manifestation and at times the only sign of a pernicious underlying condition;⁵ leading these patients to seek dermatological consultation. Thus, the role of a dermatologist in preventing the associated morbidity cannot be understated. Once the diagnosis is confirmed, a detailed evaluation by the physician is advisable; so as to accurately assess the cardiovascular status of the patient. Proper treatment, life style modifications and early intervention can help in preventing serious morbidity.

CONCLUSION

At the end, it is apt to label XSP as a mirror of morbid underlying disease. Dermatologists need to be more vigilant in such cases. Those patients presenting with XSP must be thoroughly investigated and followed up.

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