

XANTHOMA ERUPTIVUM (Case report)

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Summary

A case of Xanthoma eruptivum as the only manifestation of Type V hyperlipoproteinemia with diabetes mellitus is reported. Stepwise investigations to reach the exact diagnosis and rationale of treatment are discussed.

Introduction

The hyperlipidemic state usually constitutes 4 major types of xanthomatous lesions: plane, tuberous tendinous and eruptive. The eruptive xanthomas are seen in disorders that cause an elevation of the triglyceride fraction^{1,2}. These disorders may be either of a primary nature with genetic predisposition or acquired secondarily to some other diseases. Serum lipids are bound to proteins in the form of large molecular complexes. These lipoproteins and triglycerides are separable into four principal types by either ultracentrifugation or electrostatic techniques. These are: chylomicrons, betalipoproteins, prebeta lipoproteins and alpha-lipoproteins. Electrophoretic studies have further facilitated differentiation of familial hyperlipidemic states into 5 groups each with its specific clinical and laboratory findings. These respond differently to dietary restrictions and drugs³.

Eruptive xanthomata could be a manifestation of hyperlipoproteinemia of Type I, III, IV, and V but this type of eruption needs differentiation from

xanthoma disseminatum, which is normolipemic⁴. As mentioned earlier, it is important to know the type of hyperlipoproteinemia in order to institute the correct treatment and this is particularly so in cases of eruptive xanthomas which can be associated with four types of dyslipidoses. Disappearance of the eruptions with definite treatment instituted in the light of present knowledge of dyslipidoses has prompted us to record the following case.

Case Report

42 years old male, presented with multiple asymptomatic Papulonodular swellings on hands, forearms, elbows, knees and legs, of 4 years' duration. A detailed anamnestic investigation to detect the existence of such cutaneous abnormality or related disorders in his family proved negative. On examination patient was found to be of moderate build with body weight of 68 kgs and BP 130/76 mm of Hg. Systemic examination did not reveal any abnormal clinical findings. Numerous yellowish, round, hard, non tender papules and nodules were present on back of fingers, extensor aspect of elbows and forearms, front of knees extending down to legs and in the region of tendoachilles on both sides.

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Lesions were symmetrical in distribution. No secondary changes were seen in them. Movement of all the finger joints were full and free. There was no scarring anywhere. (Fig. 1 page No. 287).

Diagnosis

A clinical diagnosis of Xanthomatosis was made and the following investigations were done to decide the type of hyperlipoproteinemia which these represent.

Serum showed a creamy top layer and turbid infranate. Serum cholesterol was 600 mg/100 ml and Serum triglycerides 1100 mg/100 ml (Fat induced)

Glucose tolerance test showed a mild diabetic state, Hyperlipidemia Carbohydrate induced, Lipoprotein electrophoresis revealed the following features :

Chylomicrons	— Increased		Type V Hyperli- poprotei- nemia
Pre-beta	— Present, increased		
Beta	— Faint, decreased		
Alpha	— Present, normal		

Routine examination of blood, urine and stool as well as liver function tests were within normal limits. ECG did not reveal any abnormality. Biopsy of a nodule confirmed the diagnosis of Xanthoma. The case was diagnosed as Type V Hyperlipoproteinemia.

Treatment

Patient was treated with reduced carbohydrate and fat intake, weight reduction with a diet supplying 2000 calories per day and an oral hypoglycaemic agent-tolbutamide. Clofibrate was not used in this case as it is not as advantageous in Type V as in Type III hyperlipoproteinemia⁵. Most of the xanthomas completely disappeared

after about four months of therapy (Fig. 2 Page No. 287).

Comments

Xanthomas of skin can present as tendinous, planar, tuberous, papulo-eruptive, xanthelasma and xanthochromia. With rare exception the presence of xanthomas indicate a disorder of lipid metabolism. Xanthomatous lesions of skin of "normocholesterolemic" type are histiocytosis syndromes⁴. Cutaneous xanthomas may be seen in three groups of patients: (a) Those who have a basic genetic biochemical defect characterised by hypercholesterolemia, hyperlipidemia or both which may be induced either by carbohydrate, fat or both in the diet (b) those who have a systemic disease that causes a secondary overloading of the transport mechanism with lipids resulting in their metastatic deposition; and (c) those who have lymphoreticular or myeloproliferative disorders¹.

Inherited abnormalities in lipoprotein metabolism are classified into five types³. Type V hyperlipemia is a mixed type resulting from combined fat and carbohydrate induced exogenous hyperlipemia. It may occur primarily as a heritable syndrome or as complication of several diseases. It represents a combination of type I and IV; in that it has chylomicronemia as well as hyper-pre-beta lipoproteinemia. Mixed hyperlipemia is not unusual in poorly controlled diabetics. Some have a decrease in post-heparin lipolytic activity (PHLA). Insulin increases PHLA and improves fat tolerance.

Control of elevated cholesterol and phospholipids by dietary restrictions, weight reduction, decreased caloric intake and substitution of polyunsaturated fat for saturated, has been known although this is still a controversial subject^{6,7}. Little attention has been given to diagnosis and control

of the serum triglyceride level. Triglyceride is a product of fat induction, carbohydrate induction or a combination of both these mechanisms. Elevated triglyceride level falls significantly with restriction of fat intake⁸. Whether carbohydrate induced or fat induced, triglyceridemia is sensitive to appropriate restrictive diet⁹. Patients with fat induced hypertriglyceridemia should receive only 20 to 25% of their calories in the form of fat. The source of fat has no effect on the degree of lipemia¹⁰. Individuals suffering from carbohydrate induced hypertriglyceridemia should receive less than 1000 calories from carbohydrates per day¹¹.

Cases with associated diabetes mellitus may show quick improvement in glucose intolerance with treatment of their hyperlipemia, but reduction of elevated triglyceride level is not achieved by treatment with hypoglycaemic agents. This group of individuals requires to be distinguished from those having severe glucose intolerance with an associated hyperlipemia, because here the hypertriglyceridemia is secondary to the diabetes mellitus and treatment of diabetes brings it under control¹².

In view of the impaired clearance of dietary fat intake and abnormal glucose tolerance coupled with hypertriglyceridemia, satisfactory response was obtained in this case with the line of management mentioned earlier.

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