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Case Report

Eruptive xanthomas in secondary hyperlipidemia with retinitis pigmentosa – A rare case report

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Abstract

A thirteen years old male patient of insulin dependent diabetes mellitus from Medicine Department was referred to our Dermatology Department with multiple eruptive xanthomas over the buttocks, knees and hands. Patient was blind since birth and diagnosis of "retinitis pigmentosa" was made by Ophthalmology Department. He was investigated and found raised levels of blood sugar, triglycerides, VLDL, TSH, SGOT/PT and kidney functions tests. He was diagnosed as a case of eruptive xanthomas with retinitis pigmentosa in secondary hyperlipidemia. We have reported this case because of the rare associations.

Key words

Eruptive xanthomas, Hyperlipidemia, Retinitis pigmentosa.

Introduction

Eruptive xanthomas may occur suddenly and at any site, but most commonly favor the buttocks, flexors of arms, thighs, knees or may be localized to pressure points. Lesions usually appear as yellow papules. Eruptive xanthomas are usually associated with pure or mixed hyper-triglyceridemia and high concentration of very low density lipoproteins (VLDL) or chylomicrons of Fredrickson's classification (**Table - 1**). They are therefore associated with Frederickson type 1, 5 and type-4 hyperlipidemia [1]. They may also be seen in secondary hyperlipidemia, generally together with insulin dependent diabetes-mellitus, obesity, pancreatitis, chronic renal failure, hypothyroidism and treatment with steroids or retinoids.

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Table – 1: WHO/Fredrickson's classification of hyperlipoproteinemia/hyperlipidimia	
Type-I	Hyper-chylomicronemia
Type-IIa	Elevated LDL (Familial hypercholesterolemia)
Type-IIb	Elevated LDL and VLDL (Familial combined hypercholesterolemia)
Type-III	Broad β-VLDL (Familial dysbetalipoproteinemia)
Type-IV	Elevated VLDL (Familial hyper-triglyceridemia)
Type-V	Elevated chylomicrons and VLDL (mixed hyperlipidemia)

Case report

A 13 years old male patient presented with sudden eruption of yellow colored papules over bilateral buttocks, knees, palms and dorsum of hands over a period of 2-4 weeks. The lesions were asymptomatic and progressive in nature. The patient was blind since birth and born out of a non-consanguineous marriage. The child was developmentally normal for his age. The patient was referred to ophthalmology department and was diagnosed as a case of retinitis pigmentosa. He was a known case of diabetes mellitus and was on Insulin therapy for 3-4 years. There was no family history of such illness.

Cutaneous examination revealed multiple skin to yellowish colored flat topped discrete papular lesions over the bilateral buttocks, knees, palms and dorsum of hands. **(Figure – 1)** On general physical examination, patient was blind, moderately built and well nourished. His pulse rate was 90/min regular; BP was 124/90mm of Hg, slightly higher for his age. Routine laboratory investigations were as per **Table - 2**.

Histopathological examination revealed infiltration of the dermis with neutrophils, lymphoid cells, histiocytes and few macrophages filled with lipid droplets (foam cells) along with extra vascular lipid deposits in the form of lace like eosinophilic material between the collagen bundles suggestive of eruptive xanthomas. Chest Radiography and Ultrasound abdomen were normal. A diagnosis of eruptive xanthomas in secondary hyperlipidemia with retinitis pigmentosa was made and the patient was treated with Atrovastatin, Clofibrate Gemfibrozil, Insulin therapy, L-thyroxin and Antibiotics.

There was moderate flattening of lesions over the limbs after one month of treatment. Dietary modifications were done.

Figure - 1: Bilateral knees and hands showings multiple eruptive xanthomas.

Discussion

Xanthomatosis is a cutaneous manifestation of lipidosis in which plasma lipoproteins and free fatty acids are qualitatively altered, resulting in morphologic change as lipids accumulate in foam cells in the tissues. Lipoproteins are soluble compounds formed by the combination of insoluble circulating lipids (cholesterol, cholesterol esters, triglycerides and phospholipids) and proteins. Any disorder of lipoprotein metabolism confers on an individual, an increased risk of cardiovascular disease, pancreatitis or xanthomas [2].

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In eruptive xanthomas, small yellow papules arising from a slightly wider base develop over buttocks, thighs, knees and extensor aspects of arms and legs. They are usually associated with WHO type-4, type-5 hyperlipidemia but also can be seen in patients with obesity, uncontrolled diabetes, lipase deficiency, pregnancy, and hypothyroidism etc.

Xanthomas develop either through increased uptake of lipids transported through the capillaries or increased lipid synthesis in the dermal macrophages which result in the production of foam cells [3].

The presence of increased number of E- selectin positive endothelial cells and a decrease in the intracellular cell adhesion molecule cells promote macrophage migration into xanthomas lesions [4].

Conclusion

Though the disease is associated with uncontrolled diabetes mellitus and thyroid disorder, these factors contribute to hyper-triglyceridemia which may lead to eruptive xanthomas. So it is important to treat all the conditions simultaneously and family members should also be investigated for hyperlipidemia. Patient should be on regular follow-up to avoid cardiovascular complications. Retinitis pigmentosa is a rare association with hyper-lipidemia and eruptive xanthomas. So we are reporting this case for its rarity.

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Table – 2: Laboratory investigations.

Blood/Serum/Plasma	**Result**	**Normal Range**
Glucose F	320 mg/dl	60-110 mg/dl
Glucose PP	502 mg/dl	110-140 mg/dl
HbA1C	12.9%	4.0-5.6%
Serum triglyceride level	3170	60-160 mg/dl
Serum cholesterol	48	150-220 mg/dl
LDL	199	40-60 mg/dl
VLDL	634	30-52 mg/dl
Blood Urea	88 gm%	15-45 mg/dl
S-creatinine	5.3 mg/dl	0.8-1.2 mg/dl
S. TSH level	12.03 mIU/L	**0.4 - 4.0** mIU/L
SGOT	120	Up to 50IU/L
SGPT	142	Up to 50IU/L
Hb	11 gm/dl	**13.5 to 17.5** gm/dl
Total leukocyte count	14,800/mm^3	4000-11000 /cumm
Differential Leukocyte Count		
Polymorph	76%	40-75%
Lymphocyte	20%	20-40 %
Monocyte	2%	02-10%
Eosinophil	2%	0-8 %
Urine Examination		
Microscopic Examination	presence of Pus cells	Nil
Albumin	2+	Nil
Sugar	3+	Nil