The Clue to his Pancreatitis!

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Introduction

Xanthomas are lesions characterized by accumulations of lipid-laden macrophages. They are a common manifestation of lipid metabolism disorders, including hyperliproteinemia (HLP). HLPs have been classified according to 5 major phenotypes described in table 1. These phenotypes are based on specific electrophoretic patterns seen in patients with elevated lipoprotein levels. We present a case of a patient presenting with gallstones and pancreatitis, initially misdiagnosed as gallstone pancreatitis. His skin manifestations in combination with an abnormal lab specimen clued us in to the correct diagnosis.

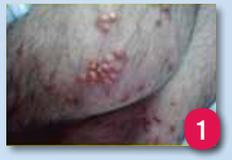
Case

A 28-year-old male presented to the Emergency department with abdominal pain. He had a CAT scan which showed extensive peripancreatic edema and cholelithiasis. His amylase and lipase levels were markedly elevated, but his LFTs were normal. A diagnosis of gallstone pancreatitis was made and the surgical service was consulted. Prior to entering the room, we were cautioned that the patient had a, “nasty” rash. On examination, he was found to have the lesions shown on images 1 and 2. Subsequently, we were called by the lab stating that the laboratory specimen was lipemic (Image 3), and his triglyceride level was >5000 mg/dL. This sent us to the textbooks, and we were quick to realize that he had tuberoeruptive xanthomas, most likely related to a form of HLP. He was later diagnosed with type III HLP, which was considered the cause of his pancreatitis. Both medicine and endocrine were consulted and the patient was transferred to the ICU.

Discussion

The patient was managed in the ICU with IV fluids, pain control, gemfibrozil/statin, and tight glycemic control for approximately 3 weeks. He recovered from his pancreatitis and was discharged home.
The plan is for an eventual cholecystectomy, but for now, he requires tight control of his HLP.

He was diagnosed with Type III HLP, a serious disorder, with lifelong consequences of premature vascular disease and recurrent pancreatitis. Drug treatment (such as HMG-CoA reductase inhibitors/other lipid-lowering drugs) is indicated since dietary modifications alone are insufficient in managing these patients.



Types of Hyperlipoproteinemia

Types	Causes and incidence	Diagnostic findings
I		
(Frederickson’s hyperlipoproteinemia, fat-induced hyperlipemia, idiopathic familial)	• Deficient or abnormal lipoprotein lipase, resulting in decreased or absent post heparin lipolytic activity • Relatively rare • Present at birth	• Chylomicrons (very low density lipoprotein (VLDL), low density lipotein (LDL), high density lipopro plasma 14 hours or more after last meal • Highly elevated serum chylomicrons and triglyceride levels, slightly elevated serum cholesterol levels • Lower serum lipoprotein lipase levels • Leukocytosis
II		
(Familial hyperbetalipoproteinemia, essential familial hypercholesterolemia)	• Deficient cell surface receptors that regulate LDL, degradation and cholesterol synthesis, resulting in increased levels of plasma LDL over joints and pressure points • Onset between ages 10 and 30	• Increased plasma concentrations of LDL • Increased serum LDL and cholesterol levels • Amniocentesis shows increased LDL levels
III		
(Familial broad beta disease, dysbetalipoproteinemia, remnant removal disease, xanthomia tuberosum)	• Unknown underlying defect results in deficient conversion of triglyceride rich VLDL to LDL • Uncommon: usually occurs after age 20 but can occur earlier in men	• Abnormal serum beta lipoprotein • Elevated cholesterol and tryglyeride • Slightly elevated glucose tolerance • Hyperuricemia
IV		
(Endogenous hypertriglyceridemia, hyperbetalipoproteinemia)	• Usually occurs secondary to obesity, alcoholism, diabetes, or emotional disorders • Relatively common, especially in middle-aged men	• Elevated VLDL levels • Abnormal levels of triglycerides in plasma: variable increase in serum • Normal or slightly elevated serum cholesterol levels • Mildly abnormal glucose tolerance • Family history • Early coronary artery disease
V		
(Mixed hypertriglyceridemia mixed hyperlipidemia)	• Defective triglyceride clearance causes pancreatitis usually secondary to another disorder, such as obesity or nephrosis • Uncommon: onset usually occurs in late adolescence or early adulthood	• Chylomicrons in plasma • Elevated plasma VLDV levels • Elevated serum cholesterol and triglyceride levels

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