Recurrent Pancreatitis Induced by Uncontrolled Type V Hyperlipoproteinemia

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Abstract

Background: Type V Hyperlipoproteinemia is a rare disease characterized by increased chylomicron and very low-density lipoprotein (VLDL) plasma levels. No official guidelines for treatment or monitoring of this disorder have been established due to the scarcity of cases. We present a patient with recurrent episodes of pancreatitis and diffuse eruptive xanthomas associated with uncontrolled type V hyperlipoproteinemia requiring recurrent Intensive Care Unit (ICU) admissions for management of his condition. Clinical Case: A 28 y/o male with Type V Hyperlipoproteinemia, Hypertension, uncontrolled Type 2 Diabetes Mellitus (DM2) and history of non-compliance, presented with epigastric pain and increased xanthoma formation. Family history was remarkable for hypertriglyceridemia and Coronary Artery Disease in his father and grandfather. On examination, he had moderate epigastric tenderness along with diffuse and extensive xanthomas in both hands, knees, and thighs. Outpatient regimen for the management of the hyperlipoproteinemia consisted of Gemfibrozil 600mg twice daily, Niacin 2000mg daily, and Omega-3 2,000mg twice daily. He was also on Lantus and Novolog for the management of DM2, last A1C being 12.4 %. Laboratories for this admission showed triglycerides >5,680 mg/dl (n<150 mg/dl), Cholesterol >2,820 mg/dl, HDL 30 mg/dl, LDL 29 mg/dl, Lipase 17 u/L (n <78 u/L), Amylase 54 u/L (n <125 u/L). MRCP showed an atrophic
pancreas. The patient was admitted to the ICU for the management of recurrent pancreatitis secondary to severe hypertriglyceridemia. He was started on intravenous insulin drip, restarted on home medications, low-fat diet, and Pravastatin. After 8 days on the insulin drip, his triglycerides had decreased to 566 mg/dl and abdominal pain had resolved. He subsequently underwent a celiac plexus nerve block due to recurrent admissions for abdominal pain. **Conclusion:** Type V Hyperlipoproteinemia is a complex lipid disorder that is likely due to triglyceride overproduction and clearance defect of VLDL. Environmental factors such as uncontrolled DM2, obesity, and increased alcohol intake can worsen the disease and lead to hypertriglyceridemia associated pancreatitis. Treatment involves lifestyle modifications, and medications, such as fibrates, nicotinic acid, fish oil if indicated to prevent pancreatitis and to lower cardiovascular risk. We present a case of hereditary hyperlipoproteinemia causing significant morbidity and recurrent ICU admissions for the treatment of severe hypertriglyceridemia. Due to the severity of hypertriglyceridemia and acute pancreatitis, intravenous insulin was used to enhance lipoprotein lipase activity to accelerate chylomicron and VLDL metabolism, therefore decreasing triglycerides. Although rare, this genetic disorder can cause significant and recurrent pancreatitis that may increase mortality if not appropriately treated.

**Issue Section:** Endocrine Regulation of Cardiovascular Disease: Hormones and Lipids