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CASE REPORT

Familial hypertriglyceridemia-induced acute necrotizing pancreatitis: A case report

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Abstract

Acutepancreatitis. can infrequently be a life-threatening complication of hypertriglyceridemia. Rarely, hypertriglyceridemia can originate from an inborn genetic error in lipoproteins metabolism. This condition can be manifested by very high serum triglyceride levels (>1000 mg/dl) and a more severe and lethal form of pancreatitis. Here, we present a case of acute pancreatic which was found out to be a complication of an undiagnosed familial hypertriglyceridemia who died in spite of receiving conventional care and treatment. Focus is on the importance of considering infrequent etiologies of acute pancreatitis and application of more effective treatments.

Key Words: Hypertriglyceridemia; Pancreatitis; Acute Necrotizing

Introduction

Hypertriglyceridemia (HTG) is a rare cause of acute pancreatitis accounting for only 1–4% of cases(1-3). The risk of developing acute pancreatitis is about 5% with triglyceride levelshigher than 1,000 mg/dl and 10-20% with TG levels higher than 2,000 mg/dl(4). It has been reported that hypertriglyceridemia-induced acute pancreatitis is associated with increased severity and complications when compared with pancreatitis induced by common causes such as gallstone or alcohol(5). HTG can infrequently be an autosomal dominant genetic disorder and is associated with higher TG levels in comparison to secondary HTG(6). Here, we describe a rare case of acute necrotizing pancreatitis caused by familial HTG to highlight the importance of considering familial HTG as an uncommon but life-threatening etiology when facing acute pancreatitis.

Cases

A 30-year-old woman presented to our emergency department with severe epigastric pain and vomiting. The pain, which had started in the same morning, was intense, unremitting, and radiating to her back and left arm. The patient was trying to keep a sitting and leaning forward position because it helped to decrease pain.

She had no previous history of similar symptoms. She had no history of fever, trauma or

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poisoning. Her past medical history was significant for diabetes for which she was on daily NPH insulin. She was taking no other medications. She did not smoke or drink. Regarding her familial history, hyperlipidemia in her father, sister and uncle and stroke in her uncle was remarkable.

On admission, she was alert, arterial blood pressure was 135/90 mm Hg, heart rate was 122 beats/min, and respiratory rate was 15 breaths/min. Physical examination of the abdomen was normal except for epigastric tenderness. No masses or organomegaly was noted on abdominal examination. Nothing was found in favor of abdominal free fluid. Other examinations did not reveal significant findings.

The initial relevant laboratory testing revealed total WBC count 19000/mm^3 with neutrophilic predominance (%80);red blood cells count was 4700000/mm^3 with a MCH of 33Pgand aMCHC of 39.2%. Serum amylase was 547 mg/dl. SGOT and SGPT were reported as 95U/L and 112U/L. Serum creatinine was 0.3 mg/dl. Reported LDH was 620U/L.The patient had a blood glucose level of 390 mg/dl. Serum triglyceride was 6140 mg/dl;total cholesterol was 1017mg/dl; HDL cholesterol and LDL cholesterol were 30 mg/dl and 572 mg/dl, respectively. We noticed that the serum looked milky and was full of chylomicrons. Other lab findings were in normal range and did not reveal anything significant.

An enhanced CT scan of the abdomen was performed, which showed decreased liver density with fatty liver changes, highly enlarged pancreatic parenchyma with necrotic areas especially in its tail, peripheral fluid accumulation,left lateral pleural effusion, and fluid in the pelvic cavity. These finding indicatedsevere acute pancreatitis with CT severity index (CTSI) of 8.

Acute pancreatitis was diagnosed for the patientwith aRanson's score of 3 and an APACHE II (acute physiology and chronic health evaluation) score of 15. The patient was transferred to the internal ward and underwent treatment with fluids, antibiotics, insulin, and analgesics. In the next hours a general surgery consultation was performed and, due to exacerbation of her condition,she wasadmitted to the intensive care unit (ICU). Statins, heparin,broad spectrum antibiotics, and octreotidewere added to the initial treatment.

On the second day, Respiratory rate increased to 50 breath/min and, due to respiratory insufficiency, she wasintubated and underwent mechanical ventilation.Lab results of the second day showed no remarkable improvement. On the same day, her blood pressure decreased and shedeveloped tachycardia and other signs of hypovolumic shock. To correct the condition, with the help of a central venous catheter (CV line), we monitoredCVP and administered fluid accordingly. Because of her high level of TG with no response to treatment, she became a candidate Forplasmapheresis although it could not be Performed because of insufficient facilities in our center. Instead, we started infusion of glucose and insulin. The last lab data showed cholesterol of 340, TGof 1363, and HDL andLDL of13and 77, respectively. On the third day, her condition exacerbated which did not respond to our efforts and led to her death.

Discussion

Acute pancreatitis is a condition with severe complications and high mortality(7). Acute pancreatitis is diagnosed if the patient meets at least two of the three criteria that follow: severe acute epigastric pain, serum amylase or lipase level over 3 times higher than the normal level,and manifestation of acute pancreatitis in imaging examinations. Our patient was admitted with acute abdominal pain, elevated amylase, and imaging that approved thediagnosis. For the etiology of acute pancreatitis, the imaging detected no structural causes such as gallstones, microlithiasis, pancreas divisum, sphincter of Oddi disease, etc.

The only identifiable etiology behind our patient's condition was HTG, which had been undiagnosed and seemed to be primary and familial.HTG accounts for approximately 3% of all cases ofacute pancreatitis, main causes being gallstones and alcohol(2). A highly elevated serum triglyceride level predisposes patients to pancreatitis development, occasionally leading to total necrosis of pancreas and death(8).

Although the exact mechanism is not clear,it seems that an elevated cholesterol level alone may not develop pancreatitis(9). The exposure of circulating TGs to lipase in the pancreatic capillaries could result in the hydrolytic formation of free fatty acids, which causes local tissue injury, the release of pancreatic enzymes and consequent auto-digestion(10).Typically, HTG-induced pancreatitis occurs in a patient with a pre-existing lipid abnormality, along with the presence of a secondary precipitating factor (e.g., poorly controlled diabetes, alcohol or medication). We suppose that in our patient, poorly controlled diabetes was the predisposing factor.

HTG can be primary in less than 5% of the cases, which is due to genetic causes.It is more often secondary to other causes such as diabetes,

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obesity, pregnancy, excess carbohydrate intake, hypothyroidism, alcohol, hepatitis, sepsis, renal failure, and drugs.TG levels higher than 1000 to 2000 mg/dl, similar tothe case with our patient, is expected to exist in patients with primary HTG especially in Type I, III, IV, and V hyperlipoproteinemia (Fredrickson's classification)(6). Her positive familial history was also supporting the diagnosis.

It has been reported that acute pancreatitis caused by HTG (especially familial types) is more severe and lethal in comparison to acute pancreatitis with other etiologies(5). It was clearly demonstrated in our patient as a severe, totally necrotizing pancreatitis with poor response to treatment.

Acute pancreatitis can be treated with fasting, gastric decompression by nasogastric tube, analgesia,and fluid therapy(11). These were administered for our patient although no amelioration were found, which was probably due to the ongoing etiology underlying the disease.For HTG, insulin-glucose perfusion can be used to control the extremely high triglyceride levels(12). Itlowered TG in our patient but could not stop the rapid disease progression. In patients who are in critical condition like ours, plasmapheresis can also be considered as a simple and safe optionfor TG removal. There have recently been few reports on the use of this method in such cases(3, 13-19). In these reports, indications of plasmapheresis were (a) patient not responding to pharmacological therapy, (b) serum triglyceride>1000 mg/dl, and (c) organ dysfunction. Our patient had at least 2 out of the three criteria.

in an observational study, Gubensek et al reported that plasmapheresis could dramatically improve the patients' outcomes (14). Another recent study has noted that patients with TG levels > 5000 mg/dl (similar to our case) who received plasmapheresis experienced shorter hospital courses, better outcomes, and decreased chances of readmission (19). The beneficial effect of plasmapheresis seems to be because of the rapid decrease in TG levels and also removal of plasma proteases, which are key factors in inflammation (13). On the other side, in a recent review on application of plasmapheresis as a treatment for HTG induced acute pancreatitis, it was reported that although this method can reduce TG levels up to 80%, there is not enough evidence to confirm its efficacy and safety (20). Furthermore some studies claim that there is no evidence to support mortality reduction by this method (21).

At any rate, in this case, plasmapheresis was not accessible at the moment and could not be provided in the proper time due the rapid progressionof the disease.

Conclusions

A severe, lethal and poorly-responding acute pancreatitis can complicate an asymptomatic, undiagnosed familial HTG.We would suggest screening with a fasting full lipid profile in individuals who are potentially at a risk of severe hypertriglyceridemia, especially individuals with considerable familial history or those with diabetes. Furthermore, in known HTG cases, preventive care with dietary fat restriction and lipid-lowering agents must be instituted. In the situation of acute pancreatitis development, early plasmapheresis should be considered in combination with first-line treatments and should not be postponed to failure of other treatments, although we need more studies with larger sample sizes in order to be able to confirm the applicability of this method.

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Conflict of Interest

The authors have no competing interests.

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