

An Unusual Presentation of Autoimmune Pancreatitis in a Patient with Acute Pancreatitis and Obstructive Jaundice

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ABSTRACT

Autoimmune pancreatitis is a rare fibroinflammatory disorder of the pancreas with immunoglobulin G 4(IgG4) associated systemic disease which is steroid responsive.^{1,2} It has a variable clinical presentation ranging from obstructive jaundice, abdominal pain, steatorrhea and diabetes mellitus. In general presentation of autoimmune pancreatitis as acute pancreatitis is considered to be uncommon. The article presents a case of a 30 years old female newly diagnosed as diabetes mellitus presented with obstructive jaundice with raised lipase and amylase treated as acute pancreatitis. Due to persistent elevation of amylase and lipase and deranged liver function test she was further evaluated her CT findings were typical of autoimmune pancreatitis with raised serum IgG4 levels diagnosed as autoimmune pancreatitis. She was treated with steroids and responded well and after few months presented with relapse.

KEY WORDS: *Autoimmune Pancreatitis, Obstructive Jaundice, Immunoglobulin IgG4, Acute Pancreatitis.*

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INTRODUCTION

Autoimmune pancreatitis is a form of pancreatitis in which there is systemic involvement of different organs with clinical, serological and histological features of autoimmune pancreatitis. It is also known as lymphoplasmacytic sclerosing pancreatitis featuring multiorgan immunoglobulin IgG4 rich lymphoplasmacytic infiltration.^{3,4,1} There are 2 types of autoimmune pancreatitis. Type 1 disease is most common associated with extrapancreatic manifestations and elevated levels of IgG4 positive cells. Type 2 is characterized by paucity of IgG4 positive cells which is more difficult to diagnose.^{9,10}

CASE

A 30 years old female known case of hypertension presented with a short history of epigastric pain associated with nausea and vomits. She had no symptoms of dysphagia, haemetemesis, diarrhea, and pain on defecation, hematochezia or melena. There was no history of alcohol or illicit drug use. On examination she was a young female of average height and built. Her pulse was 76 beats/min normal volumes with regular rhythm, BP was 134/86, temperature was 96.8 Fahrenheit, and she had sclera icterus with yellowish discoloration of her skin and mucous membrane. Her abdominal examination revealed slight distension and there was tenderness in right hypochondriac n epigastric region without rebound, gut sounds were audible. Her respiratory examination, cardiovascular system examination and central nervous system examination were unremarkable. The laboratory investigations revealed Hb 13.8gm/dl (11.5-15.4 gm/dl), PCV 42 (35-47%), platelet count 286(150-440*10⁹/L), total leukocyte count 21.8(4.0-10*10⁹/L), Serum Na 143MEq/L(136-139 MEq/L), K 3.4 MEq/L(3.8-5.2 MEq/L), HCO₃ 19MEq/L(22-29 MEq/L) Cl 102 MEq/L(98-107 MEq/L), Urea 23(10-50)mg/dl, creatinine 0.52mg/dl(0.6-1.5 mg/dl), serum albumin 3.9 (3.63-4.92gm%), Ca 8.12(8.1-10.4 mg/dl), Mg 2.53mg/dl(1.58-2.55mg/dl), total bilirubin 4.81mg/dl (less than 1.3mg/dl) direct bilirubin 3.25 (less than 0.3 mg/dl) alkaline phosphatase 112 IU/L (adults 39-117IU/L) SGPT 460 IU/L (upto 31 IU/L), gamma GT 131IU/L (11-50

IU/L), amylase 2112IU/L(28-100IU/L), lipase 8064 IU/L(13-60IU/L), Mg 2.04 mg/dl (1.58-2.55mg/dl). Her fasting lipid profile was within normal limits, tests for hepatitis A, B and C were negative. On further evaluation antinuclear antibody was negative in contrast immunoglobulin IgG4 leveled was found to be raised 322mg/dl. Ultrasound abdomen showed hypoechoic body and tail and bulky pancreas. Abdominal CT scan results revealed that her pancreas was diffusely enlarged, especially at the body and tail; no focal pancreatic lesions were seen CT scan index 2/3. A diagnosis of autoimmune pancreatitis presenting with acute pancreatitis was made^{5,6} and treated with antibiotics and I/V fluids initially and latter on given steroids to which she responded well and then discharged. She followed up in outpatient clinic after 2 weeks with a decline in lipase amylase and correction of liver function tests and then again followed up in outpatient clinic after 3 months with a relapse.

DISCUSSION

Autoimmune pancreatitis is a heterogeneous disorder with important variations in pathophysiology, genetic predisposition and extrapancreatic manifestations. Autoimmune pancreatitis can present as a primary disorder of pancreas or it can occur as a part of systemic disease associated with elevations in levels of IgG4 producing cells.^{7,8} The peak age of onset of type 1 disease is 6th decade; here the patient is young female of 30 years. Usually autoimmune processes have female preponderance. The most common form of autoimmune pancreatitis occurs frequently in male's particularly elderly males with an overall ratio of 2:1.¹¹ Autoimmune pancreatitis is diagnosed on the HISORT criteria published from MAYO CLINIC which takes into account all aspects of imaging pathology, laboratory values and response to steroids. It has 5 characteristic features:

1. Diagnostic histology, periductal lymphoplasmacytic infiltrate with storiform fibrosis and obliterate phlebitis.
2. Characteristic imaging, diffuse pancreatic enlargement rim enhancement with diffusely irregular narrow pancreatic duct.
3. Serology, elevated levels of IgG4.

4. Other organ involvement, like bile duct, retroperitoneal, salivary glands or mediastinum.¹⁰
5. Response to steroids, marked improvement.

The diagnosis of autoimmune pancreatitis was made in those with >1 or 1 of the following:

1. Diagnostic histology.
2. Characteristic pancreatic imaging.
3. Pancreatic disease with abnormal serology and/or other organ involvement that shows response to steroids. The case patient had characteristic pancreatic imaging and abnormal serology.

Acute pancreatitis is diagnosed when at least 2 of the following are present: severe upper abdominal pain, elevated serum lipase >3 times

normal with or without amylase and CT showing features of pancreatic inflammation (per pancreatic stranding, edema and swelling). Relapses occur in autoimmune pancreatitis. Diabetes mellitus is a common complication of autoimmune pancreatitis. The case patient was also a newly diagnosed case of diabetes mellitus.

In conclusion timely diagnosis and an appropriate index of suspicion require awareness of the disease. Presence of obstructive jaundice, elevated serum IgG4, abnormal liver enzymes and CT findings may provide important clues for the diagnosis of autoimmune pancreatitis in patients presenting with pancreatitis. Its good response to steroids long term follow-up is essential as it has relapse and recurrence.

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