

Auto Immune Pancreatitis Presenting As Obstructive Jaundice

Nurul Amin Miah¹, Tania Mahbub², Ferdous Jahan³, Mehruba Alam Ananna⁴, Shohael Mahmud Arafat⁵, Salma Jahan⁶

¹Medical officer, Dept of Medicine, SSMC&MH, Dhaka, ²Medical officer, Kurmitola General Hospital Dhaka, ³Medical officer, Department of Medicine, Bangabandhu Sheikh Mujib Medical University, Dhaka, ⁴Assistant Professor, Department of Nephrology, BIRDEM, ⁵Associate Professor, Department of Medicine, ⁶Consultant, Paediatric Nephrology, Bangabandhu Sheikh Mujib Medical University, Dhaka;

Abstract:

A 71 years cachexic male presented with sign symptoms of biliary tract obstruction. There was a large mass in the head of pancreas with raised CA 19.9. But elevated serum lipase raised the suspicion of pancreatitis. Elevated immunoglobulin IgG₄ confirmed this case as a auto immune aetiology. Initially there was much difficulty to differentiate auto immune pancreatitis from pancreatic carcinoma but after successful stenting and a course of corticosteroids, patient improved dramatically along with normalisation of all the radiological, bio-chemical and immunological parameters.

Key word: Auto Immune Pancreatitis, Obstructive Jaundice, Pancreatic Carcinoma.

[BSMMU J 2014 ; 7 (1) : 74-76]

Introduction:

The term autoimmune pancreatitis is used to describe a heterogeneous set of pancreatic conditions which are associated with characteristic laboratory and histologic findings. The prevalence of autoimmune pancreatitis has been estimated to constitute approximately 5-10% of patients diagnosed with chronic pancreatitis. The mean age at diagnosis is 55 year, and while autoimmune pancreatitis occurs in both sexes, it is twice as common in men as in women.¹ Imaging of patients with autoimmune pancreatitis commonly reveals an enlarged homogeneously disorganized pancreas and regional lymphadenopathy. When confirmed with serum IgG₄ levels or response to treatment, the diagnosis of autoimmune pancreatitis can be made.³

Autoimmune pancreatitis can mimic pancreatic adenocarcinoma clinically and radiographically. One must differentiate between the two disorders to prevent unnecessary surgery or delay in corticosteroid therapy. Treatment of autoimmune pancreatitis with corticosteroids leads to the rapid and sustained resolution of pancreatic mass lesions,

biliary obstruction, and pancreatic-duct strictures, which has stimulated widespread interest in this condition from gastroenterologists, endoscopists, pathologists, and surgeons.²

Case report:

A 71-years married muslim, non diabetic, normotensive, non alcoholic retired school teacher from Tangail presented to the Dept of Medicine, SSMC&MH, Dhaka on 14 March 2010 with the complaint of pain in right upper abdomen for 2 months. Pain was continuous, mild to moderate in intensity, radiates towards back which was not burning in nature but increased was after taking meal and also associated with nausea. He lost almost 5 % of his total body weight within this short period. With this complaint he was treated by local physician and improved partially but 20 days later, the patient developed jaundice which was quickly increasing in intensity. On query, he gave history of dark urine, pale stool, itching but no history of previous jaundice, family history of liver disease, intravenous drug abuse, recent use of hepatotoxic medication. There was no risk factors for acute pancreatitis, no sign symptoms of lymphoma, no history of gall stone and abdominal surgery, family history of GI cancer or any other gastro intestinal disorder. All other systemic

Address for Correspondence: Md. Shoheal Mahmud Arafat
Associate Professor, Department of Medicine, BSMMU
E-mail: arafatdr@yahoo.com

query were insignificant with unremarkable past medical history. On investigations, Hb% was 9.4g/dl, ESR-88mm in first hr, RBS 6mmol/l, serum bilirubin 14.7mg/dl[13.4.10], all viral markers were negative, prothrombin time 12 sec, serum lipase 1033u/l, serum amylase 203u/l, SGPT 71u/l, alkaline phosphatase 176u/l, CA-19.9-7231.30u/ml[normal up to 37u/l], USG whole abdomen showed a large ovoid hypo echoic mass of 54X29mm seen along the pancreatic head region. Mass showed some flow within it and compressing the CBD. ERCP showed that distal CBD was narrow and obstructed. Proximal CBD and distal CHD were dilated. (Fig-1)

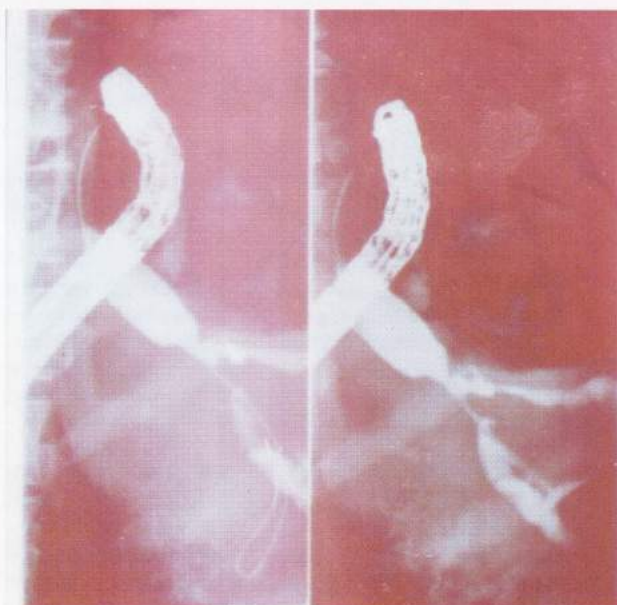


Fig-2: ERCP Report:Duodenum and papilla: Papilla normal. No bile flow seen. Pancreatogram: Normal main

duct and 2nd generation ducts.

Cholangiogram: Distal CBD is narrow and obstructed. Proximal CBD and distal CHD dilated. Cystic duct and gallbladder opacified,look normal. Proximal CHD is narrow and obstructed extending upto hilum. Right hepatic duct opacified. After papillotomy,guide wire passed and endoprosthesis implanted into the right hepatic duct. Bile flow established.

Comment: Cholangio Carcinoma at CHD and distal CBD.

Therapeutic stenting was done to relieve the obstruction. After stenting, the patient improved dramatically. As there was very high rise serum lipase, auto immune pancreatitis is an important differential diagnosis. We investigated the patient for immunoglobulin IgG, which was very high, i.e. 2912.68mg/dl[normal up to 700-1600mg/dl]. ANA was positive and serum immunoglobulin IgG was also raised - 9.47g/l[normal-0.03 to 2g/l]. We have treated the case with corticosteroid [40mg/day with gradual tapering of doses. All his symptoms disappeared within 2 weeks. On repeated investigations, LFT returned to normal. USG show normal pancreas with stent in CBD, CA 19.9-128u/ml. Despite high suspicion of Ca-head of pancreas, the case was diagnosed as a case of auto immune pancreatitis.

Discussion:

In this case, an elderly patient who presented with features of obstructive jaundice having a mass occupying in pancreas on ultrasonographically pancreatic carcinoma was highly suspected but simultaneous presence of high CA19-9 and very high serum lipase made the case more conflicting. Moreover there was report that auto immune pancreatitis is associated with high serum CA 19-9¹³ and in some cases pancreatic carcinoma may lead to raised serum lipase.¹⁴ So the diagnosis remained beyond our reach but as the patient completely improved after stenting and a course of steroid, thereby fulfilled the criteria for diagnosis of autoimmune pancreatitis.⁴ Since pathological confirmation of autoimmune pancreatitis is difficult, corticosteroid therapy can be used as a diagnostic tool in

patients whose clinical and laboratory findings are strongly suggestive of autoimmune pancreatitis. In these patients, short-interval imaging at 2 to 4 weeks after the initiation of therapy must be used to confirm whether there is resolution of the mass. Since malignant masses can also respond to corticosteroids, complete resolution of the pancreatic mass is critical for the diagnosis of autoimmune pancreatitis.²

Once a diagnosis of autoimmune pancreatitis has been made, a treatment course with corticosteroids should be considered. However, the use of corticosteroid therapy is not mandatory since there have been reports of the spontaneous resolution of a pancreatic mass, stricture, and jaundice.^{5,6} The initial dose of corticosteroids (prednisone) should be 40 mg daily for 1 week, followed by a tapering of the daily dose by 5 mg per week. The response to corticosteroids is often dramatic.^{7,8,9} During the first 2 to 4 weeks of therapy, CT should be used to monitor the response.^{10,11} Laboratory abnormalities (such as elevated IgG4 levels, hypergammaglobulinemia, and presence of auto antibodies) may also show improvement with corticosteroid therapy. Extra hepatic biliary obstruction has also proved to be responsive to corticosteroid therapy.^{10, 12} A poor response to corticosteroids should raise the question of pancreatic cancer or other forms of chronic pancreatitis. Although the vast majority of patients with autoimmune pancreatitis will readily respond to corticosteroids within a few weeks, a small subgroup may require maintenance therapy with prednisone at a dose of 5 to 10 mg per day.²

So, in conclusion we want to say that autoimmune pancreatitis is an important differential diagnosis of pancreatic carcinoma. This benign disease resembles pancreatic carcinoma clinically, radiographically and serologically making the diagnosis challenging. However suspicion of auto immune pancreatitis in case of obstructive jaundice might escape the patient from unnecessary surgery.

Reference:

1. Sarles H, Sarles JC, Muratore R, Guien C. Chronic inflammatory

sclerosis of pancreas – an auto immune disease ? *Am J Dig Dis*, 1961; Vol 6:688-698.

2. Dimitry L, Finkelberg, Dushyont Sahoni, Vikram D, Millium R, Brugge. Autoimmune Pancreatitis:A mimic of pancreatic carcinoma.*J Hepatobiliary Pancreatic Surg*, 2006;4:45.
3. Amber A, Fronk, Michael D, Morse, Benjamin R, Smith, Kill S. Auto immune pancreatitis presenting with mass and biliary obstruction; Frank; *Radiology Case Report*, 2008, Vol 3:1.
4. Ryan Law, Mary B, David Vogt. Auto immune pancreatitis : A mimic of pancreatic cancer. *Cleveland Clinic Journal of Medicine*,2009, Vol 76:607-613..
5. Wakabayashi T, Kawaura Y, Satomura Y, Et al. Clinical study of chronic pancreatitis with focal irregular narrowing of the main pancreatic duct and mass formation: comparison with chronic pancreatitis showing diffuse irregular narrowing,*Pancrease*,2002;25:283-9.
6. Ozden I, Dizdaroglu F, Poyanli A, Emre A. Spontaneous regression of a pancreatic head mass and biliary obstruction due to autoimmune pancreatitis. *P a n c r e a t o l o g y* 2005;5:300-303.
7. Ito T, Nakano I, Koyanagi S, et al. Autoimmune pancreatitis as a new clinical entity: three cases of autoimmune pancreatitis with effective steroid therapy. *Dig Dis Sci* 1997;42:1458-1468.
8. Saito T, Tanaka S, Yoshida H, et al. A case of autoimmune pancreatitis responding to steroid therapy: evidence of histologic recovery. *Pancreatology* 2002;2:550-556.
9. Kojima E, Kimura K, Noda Y, Kobayashi G, Itoh K, Fujita N. Autoimmune pancreatitis and multiple bile duct strictures treated effectively with steroid. *J Gastroenterol* 2003;38:603-607.
10. Kamisawa T, Yoshiike M, Egawa N, Nakajima H, Tsuruta K, Okamoto A. Treating patients with autoimmune pancreatitis: results from a long-term follow-up study. *Pancreatology* 2005;5:234-238.
11. Fernandez-del Castillo CF, Sahani DV, Lauwers GY. A 36-year-old man with recurrent epigastric pain and elevated amylase levels. *N Engl J Med* 2003;349:893-901.
12. Hirano K, Shiratori Y, Komatsu Y, et al. Involvement of the biliary system in autoimmune pancreatitis: a follow-up study. *Clin Gastroenterol Hepatol* 2003;1:453-464.
13. Raffaele Pezzilli, Riccardo Casadei, Lucia Calculli, Donatella Santini Autoimmune Pancreatitis. A Case Mimicking Carcinoma departments of Internal Medicine, Surgery, Radiology, and Pathology, Sant'Orsola Malpighi Hospital. Bologna, Autoimmune Pancreatitis. A Case Mimicking Carcinoma *JOP. J Pancreas (Online)* 2004; 5(5):527-530.
14. R J van Klaveren, P H de Mulder, A M Boerbooms, C A van de Kaa, U J van Haelst, D J Wagener, and J C Hafkenscheid. Department of Internal Medicine, Nijmegen University Hospital, The Netherlands; Pancreatic carcinoma with polyarthritis, fat necrosis, and high serum lipase and trypsin activity; *Gut*. 1990 August; 31(8): 953–955.