Chronic Calcific Pancreatitis with Concurrent Autoimmune Hemolytic Anemia: A Case Report

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Introduction

Chronic Pancreatitis is an irreversible inflammatory disease characterized by fibrosis and progressive destruction of exocrine and endocrine function. The common complications of chronic pancreatitis include bile duct obstruction with cholestasis, duodenal obstruction, portal/splenic vein hypertension, pancreatic pseudocysts, pancreatic fistula, ascites and rare complications like colonic stenosis. Autoimmune Hemolytic Anemia is an immune malfunction caused by autoantibodies which react with RBCs. It is further classified according to the temperature at which the autoantibodies react with the RBCs into warm antibody hemolytic anemia (>37°C) or cold agglutinin disease (, , ). Autoimmune Hemolytic Anemia has variable etiologies and can occur as a primary disorder where the cause cannot be determined or secondary to other autoimmune disorders such as systemic lupus erythematosus, various drugs, infections and hematological malignancies. Herein we report one rare case of a patient with chronic calcific pancreatitis with pseudocyst with concurrent autoimmune hemolytic anemia.

Case Report

A 36 year old male nonalcoholic with chronic pancreatitis and diabetes for 12 years was admitted to the hospital with complaints of continuous, dull aching pain in the epigastric region and constipation since the past ten days. There was no history of drug intake and no significant past medical/surgical illness. In the past, USG abdomen showed a cystic lesion measuring 5 ×3.6 cms in the head of pancreas. On examination, vitals were stable and tenderness was present in the epigastric region. There was no palpable abdominal mass or other specific findings noted on physical appearance. At the time of admission, Hemoglobin was 11.9 g/dl, Total Leukocyte count was 6800 and Platelet count was 441,000. Amylase, Lipase, CEA, CA-125, CA 19-9, Liver Function tests were within normal range. Abdominal contrast computed tomography revealed an atrophic pancreatic parenchyma with marked ductal dilatation and multiple intraductal calculi with well defined 4cms cystic lesion in the body of pancreas. On endosonography, a 4.2 ×3.4 cms thin walled cyst with debris inside in the region of neck apposing D1 and body of stomach was noted suggestive of a communicating Pseudocyst. FNAC of the cystic lesion showed no malignant cells. Endoscopic retrograde cholangiopancreatography confirmed the findings with multiple calculi in the head region along with pseudocyst with fibrosis surrounding pancreas. Deep cannulation was not possible due to calculi packed in the head region and the cystic lesion could not be punctured due to fibrosis, therefore only papillotomy was performed. Patient improved symptomatically after the papillotomy.

On the day 15 after admission, patient complained of weakness and giddiness. On examination, vitals were stable and severe pallor was noted along with a hemic murmur. An urgent CT angiography was done to rule out an active bleed from papillotomy site, pseudoaneurysm and bleeding into the cyst. Blood investigations showed a Hemoglobin of 3.4g/dl, Total Leukocyte Count of 15,300, Platelet Count of 449000, ESR of 140, Presence of Nucleated RBCs, polychromasia, anisopoikilocytosis and left shift of neutrophils were noted in the Peripheral Smear. The serum LDH was 677, urine for Hemoglobin was negative and Serum Iron Profile was normal. Direct and Indirect Coombs Test was positive and anti-nuclear antibody and anti-double-stranded DNA antibody tests were negative. In view of the following, a diagnosis of autoimmune hemolytic anemia was made and patient was transfused with packed cells and started on Injection Hydrocortisone. Blood Transfusion was discontinued after 3 days and repeat Hemoglobin was 7 g/dl. After a week’s course of Hydrocortisone, patient was put on oral Prednisolone 60mg OD.

Discussion

Chronic Pancreatitis is an end stage, irreversible disease with progressive functional loss as a result of pancreatic fibrosis due to various etiologies. The incidence of chronic pancreatitis is estimated to be between 4-5% in the general population with 60 to 70% cases related to alcohol abuse followed by tropical pancreatitis. Tropical Pancreatitis is chronic calcific, non-alcoholic pancreatitis, prevalent in
developing countries like India which is seen in malnourished patients and in diets rich in tapioca. 
Hypertriglyceridemia, Hypercalcemia, genetic mutations such as CFTR-gene 9 and the SPINK-1 N34S mutation 10 implicated in tropical pancreatitis, obstruction of the main pancreatic duct by stenosis, stones, or cancer 7 are some of the less common causes. Recently a newer entity known as Autoimmune pancreatitis was described as an idiopathic inflammatory and sclerotic process associated with hypergammaglobulinemia which responds with corticosteroid therapy11. Patient with chronic pancreatitis typically present with either intermittent or continuous pain, diabetes and malabsorption as a result of loss of organ function, and complications of chronic pancreatitis. 1 Diagnosis of Chronic Pancreatitis is generally made through clinical history, imaging modalities such as CT, endosonography, ERCP or MRCP . Typical radiological features are atrophic gland with calcifications, irregularity of main pancreatic duct, duct dilatation with beaded side branches and enlargement of gland. 1 The natural history of chronic pancreatitis is variable in individual patients and depends on the etiology.

End stage complications of chronic pancreatitis such as exocrine pancreatic insufficiency and calcification generally appears faster and lasted 2-5 fold longer in hereditary and idiopathic juvenile pancreatitis as compared to alcoholic chronic pancreatitis. 13 Diabetes develops in 58% of patients and tends to be brittle 13. Common complications include pseudocyst and fistula formations, pseudoaneurysms, stenosis of the common bile duct, splenic/portal venous obstruction..13 Other rare complications include mediastinal pancreatic pseudocyst 14, pancreaticoportal fistula 15, splenic rupture and splenic pseudocyst. 16

Hemolytic anemia is defined as premature destruction of RBCS and may be classified as cellular and extracellular defects. Cellular defects include membrane defects and enzyme deficiencies. Extracellular defects include autoimmune hemolytic anemia, transfusion reactions, burns, infections, prosthetic valves and liver disease. 17Autoimmune hemolytic anemias are characterized by increased destruction of RBCS as a result of production of autoantibodies. There are 1-3 cases per 100,000 of 18Autoimmune hemolytic anemia (AIHA) per year. AIHA is slightly more common in the middle aged and females18. Warm autoimmune hemolytic anemia (WAIHA) results from production of an IgG isotype autoantibody that reacts against unclassified red cell antigens or red cell membrane protein at body temperature. They may arise spontaneously or in association with diseases like SLE, lymphoma, chronic lymphocytic leukemia and drugs like α-methyldopa, levodopa. 19 Cold agglutinin disease (cold antibody disease) is caused by autoantibodies that react at temperatures <37° C. It sometimes occurs with infections especially mycoplasma, infectious mononucleosis and lymphoproliferative states20. Autoimmune hemolytic anemia is also associated with other autoimmune illnesses such as systemic lupus erythematosis, rheumatoid arthritis, scleroderma, and ulcerative colitis. Other less common associations include AIHA with hyperthyroidism, reactive arthritis and HIV. 17 The hallmark finding in AIHA is a positive Coombs’ test which is used to confirm the diagnosis. 20 In this case, the patient presented with typical clinical history and radiological features suggestive of chronic calcific pancreatitis. During this episode, the patient developed concurrent autoimmune hemolytic anemia. The workup for secondary causes of autoimmune hemolytic anemia such as SLE was negative. SLE is an autoimmune disease with multorgan involvement which may involve one or several organ systems and over time additional manifestations may occur. The hematologic abnormalities are common manifestations in patients with SLE. It has been found that hemolytic anemias are cause of anemia in 7-15% of SLE patients. (21-25) and a study done by Nossent and Swaak showed that hemolytic anemia is the presenting symptom of SLE in 68.8% of hemolytic anemia patients.23 Likewise, patients with SLE may present with chronic pancreatitis but the association between the two is extremely rare and only six cases were reported in English literature. 25 In our patient it is not clear whether the autoimmune hemolytic anemia was coincidental or related to the underlying condition that predisposed the patient to develop the condition. To our knowledge, this is the first reported case of chronic calcific pancreatitis in combination with autoimmune hemolytic anemia.

References


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Illustrations

Illustration 1

Calcification in the head of Pancreas

Illustration 2

Pseudocyst in the head of Pancreas
Illustration 3

Peripheral Smear showing spherocytes and Nucleated RBCs
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