RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 2 | FEBRUARY 2016 | ISSN - 2249-555X

Association Between Celiac Disease and Primary Biliary Cirrhosis- a Case Report

KEYWORDS: Celiac Disease, Primary Biliary Cirrhosis, Association, Case report, Autoimmune disorders.

Dr. Hardeep SinghDeep
Professor, Department of Medicine, MD Medicine, Sri Guru Ram Das Institute of Medical Sciences and Research, Sri Amritsar.

Dr. Barjinder Pal Singh
Junior Resident, Department of Medicine, MBBS, Sri Guru Ram Das Institute of Medical Sciences and Research, Sri Amritsar.

Dr. Roopa Chopra
Senior Resident, Department of Medicine, MD Medicine, Sri Guru Ram Das Institute of Medical Sciences and Research, Sri Amritsar.

Dr. Mridu Manjari
Professor, Department of Pathology, MD Pathology, Sri Guru Ram Das Institute of Medical Sciences and Research, Sri Amritsar.

ABSTRACT Celiac Disease is an autoimmune disorder which mainly affects gastrointestinal tract causing malabsorption and chronic diarrhea. It is thought to result from the activation of both a cell-mediated (T-cell) and humoral (B-cell) immune response on exposure to the glutens of wheat, barley, rye, and rarely oats, in a genetically susceptible person. Primary Biliary Cirrhosis is a chronic cholestatic autoimmune disease with a progressive course of many decades leading to cirrhosis of liver. Both celiac disease and primary biliary cirrhosis share several features, including a higher prevalence in females, autoimmune comorbidities and specific autoantibodies. There is neither causative association between the two diseases nor the activity of one influences the course of the other, but presumably a shared susceptibility of biliary and small bowel epithelium to attack by autoimmune mechanisms can lead to association of both these diseases in the same patient. Here is a case report of a patient with coexistant celiac disease and primary biliary cirrhosis.

INTRODUCTION:

Celiac disease (also known as celiac sprue, gluten-sensitive enteropathy, and nontropical sprue) is common throughout the world and affects around 1:100 to 1:300 of the population.[1] The female-to-male ratio is 2 : 1. It is an autoimmune disorder which mainly affects gastrointestinal tract causing malabsorption and chronic diarrhea. It is thought to result from the activation of both a cell-mediated (T-cell) and humoral (B-cell) immune response on exposure to the glutens of wheat, barley, rye, and rarely oats, in a genetically susceptible person.

Primary Biliary Cirrhosis is a chronic cholestatic disease with a progressive course of many decades leading to cirrhosis of liver. It has a female predominance with 8:1 (F:M) ratio.[2] Primary Biliary Cirrhosis is considered a model autoimmune disease because of its hallmark serologic findings, the antimitochondrial antibodies (AMA) and specific bile duct pathology.

Both celiac disease and primary biliary cirrhosis share several features, including a higher prevalence in females, autoimmune comorbidities and specific autoantibodies.

Association between Celiac disease and Primary Biliary Cirrhosis is rare and has been very rarely reported in India.

History and Examination:

We recently diagnosed a patient with primary biliary cirrhosis and co-existant celiac disease. The patient(41yrs, female) was admitted to the hospital with repeated complaints of abdominal distension, pruritis and loose stools since 4 years which increased since last 2 months. Patient also gave history of severe weight loss during this period. The patient was treated by many physicians without any relief.

On examination, patient was conscious, oriented to time, place and person. Pulse rate was 90/min, Blood pressure was 120/70mm of Hg, Respiratory rate was14/min, and oral temperature was 98.4^{0}F. On general physical examination, she had anemia. Cardio respiratory examination was normal. On abdominal examination, she had ascites and splenomegaly.

Laboratory investigations:

Hemoglobin-10.9gm%(11.5-15), RBCs-3.78million(3.8-5.2), WBC-4400/mm^3(4000-11000), Platelets-1.5lakh(1.5-4.5). RFTs were within normal limits. Liver enzymes were raised-ALP-373(50-136), SGOT-180(15-37), SGPT-87(30-65), GGT-274(9-52). Serum albumin was 2.3 gm/dl(3.4-5.0). Anti mitochondrial antibodies(AMA) were positive. Tissue transglutaminase(TTG) was raised-76.7(0-7). Serology for HIV, HCV & HBsAg was negative. HBA1C, thyroid profile were within normal range. RA factor and ANA were negative. 2D Echo was also normal. USG abdomen showed liver cirrhosis with splenomegaly and mild to moderate ascites. Upper GI endoscopy showed early esophageal varices, portal hypertensive gastropathy and duodenitis. Biopsy from duodenum was suggestive of flattening of villi with marked infiltration of lamnia propria by lymphocytes.

Volume : 6 | Issue : 2 | FEBRUARY 2016 | ISSN - 2249-555X

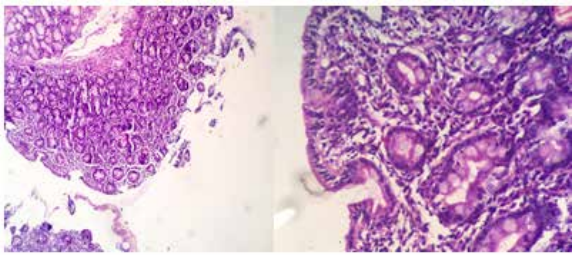

Figure no.1 Duodenal biopsy at low power field. Figure no.2 Duodenal biopsy at high power field.

Treatment:
The patient was being managed with antibiotics and prebiotics earlier, but there was no relief in symptoms. After establishing diagnosis of CD and PBC, patient was managed with gluten free diet, Ursodeoxycholic acid and steroids. Frequency and consistency of stools improved drastically after start of definitive treatment. Patient had increase in weight during hospital stay and during follow up.

Discussion:
The breakdown of the gut–liver axis balance plays a crucial role in the development of intestinal and liver disorders. An aberrant intestinal T lymphocyte and immunologically active molecules reaching the liver through the portal circulation owing to the increased intestinal permeability are thought to be critical for the development of the T cell mediated hepatic disorders associated with gut inflammation, whose best example is the association between celiac disease (CD) and primary biliary cirrhosis (PBC).[3] CD and PBC share several features, including a large predominance in female patients, a high frequency of the same autoimmune comorbidities (i.e., Hashimoto thyroiditis and Sjogren syndrome), a specific immune response against a well established autoantigen (tissue transglutaminase [TG2] for CD and E2 component of pyruvate dehydrogenase multienzyme complex for PBC). Environmental infectious factors are also thought to contribute to the onset of both the disorders with the demonstration of a molecular mimicry between TG2 and a rotavirus protein (VP7) for CD and between pyruvate deydrogenase multienzyme complex and a Gram–negative bacterium (*Novosphingobium aromaticivorans*) for PBC. A genetic susceptibility seems to be present in both the disorders. There is neither causative association between the two diseases nor the activity of one influences the course of the other, but presumably a shared susceptibility of biliary and small bowel epithelium to attack by autoimmune mechanisms can lead to association of both these diseases in the same patient.[4] So, Reciprocal screening for both diseases(CD in patient presenting with PBC and PBC in patient presenting with CD) is recommended, so that an early diagnosis, with the appropriate treatment will improve the outcome of these patients.

REFERENCE 1. World Gastroenterology Organisation Practice Guidelines: Celiac Disease,2007. 2. Kumagi T, Heathcote EJ. Primary biliary cirrhosis. Orphanet J Rare Dis 2008;3:1. 3. Volta U, Caio G, Tovoli F, De Giorgio R. Gut-liver axis: An immune link between celiac disease and primary biliary cirrhosis. Expert Rev Gastroenterol Hepatol 2013;7:253-61. 4. Kingham JG, Parker DR. The association between primary biliary cirrhosis and coeliac disease: A study of relative prevalences. Gut 1998;42:120-2.