Research

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# Celiac Disease Associated with Idiopathic Portal Hypertension: A Case Report and Literature Review

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## 1. Abstarct

Idiopathic portal hypertension (IPH) is a rare condition characterized by features of portal hypertension without evidence of cirrhosis or portal vein obstruction. The etiology remains unclear, and diagnosis is usually established after exclusion of other causes. We report a case of a 39-year-old female who presented with severe pancytopenia, massive splenomegaly, and tense ascites. Comprehensive evaluation excluded common etiologies of portal hypertension. Upper gastrointestinal endoscopy revealed esophageal varices and mucosal abnormalities in the duodenum suggestive of malabsorption. Histopathological examination of the duodenal biopsy demonstrated significant villous atrophy with increased intraepithelial lymphocytes, and serological tests showed high titers of anti-tissue transglutaminase IgA and anti-endomysial antibodies, confirming celiac disease. The patient was started on a strict gluten-free diet, which led to significant clinical and laboratory improvement, including reduction in splenomegaly and ascites. This case emphasizes the rare association between celiac disease and idiopathic portal hypertension and highlights the importance of considering celiac disease in patients with unexplained portal hypertension.

## 2. Introduction

Idiopathic portal hypertension (IPH), also referred to as non-cirrhotic portal hypertension, is an uncommon disorder characterized by increased portal venous pressure in the absence of liver cirrhosis or extrahepatic portal vein obstruction. The clinical presentation typically includes splenomegaly, hypersplenism with pancytopenia, ascites, and the development of esophageal varices. Although several mechanisms have been proposed, the exact cause of IPH remains uncertain.

Celiac disease (CD) is a chronic immune-mediated disorder triggered by ingestion of gluten-containing foods in genetically predisposed individuals. Gluten proteins present in wheat, rye, and barley initiate an inflammatory immune response in the small intestine, leading to villous atrophy and malabsorption. The prevalence of celiac disease is estimated to be approximately 1% worldwide.

Although celiac disease mainly affects the gastrointestinal tract, numerous extraintestinal manifestations have been described. Hepatic involvement is among the recognized extraintestinal features and may present as isolated elevation of liver enzymes, autoimmune liver disorders, or, rarely, portal hypertension.

In this report, we present a patient with severe manifestations of portal hypertension who was subsequently diagnosed with celiac disease. The patient showed remarkable clinical improvement after initiation of a gluten-free diet.

## 3. Case Presentation

A 39-year-old woman was referred to the gastroenterology clinic from the hematology department for further evaluation of pancytopenia, splenomegaly, and ascites. The patient reported persistent fatigue, generalized weakness, and early satiety over several months.

On physical examination, the patient appeared pale. Abdominal examination revealed significant distension with shifting dullness suggestive of ascites. A markedly enlarged spleen was palpable extending well below the left costal margin.

Laboratory investigations revealed pancytopenia. Liver function tests demonstrated normal serum albumin, normal international normalized ratio (INR), and normal total bilirubin levels. However, mild elevations were noted in alanine aminotransferase (ALT), aspartate aminotransferase (AST), and alkaline phosphatase.

Abdominal ultrasonography revealed a normal-sized liver with homogeneous echotexture and no focal lesions. The spleen was significantly enlarged, measuring approximately 21 cm. Doppler ultrasonography showed dilation of the portal vein measuring up to 18 mm without evidence of thrombosis. The hepatic veins appeared normal, and the splenic vein was also dilated.

A contrast-enhanced magnetic resonance imaging (MRI) scan of the abdomen confirmed a normal liver size and morphology with marked splenomegaly measuring 22.3 cm. The splenic vein measured 1.8 cm and the portal vein 1.7 cm in diameter. Multiple collateral vessels were observed around the splenic hilum, indicating portal hypertension.

Further laboratory investigations were performed to determine the underlying cause of portal hypertension. Tests for viral hepatitis including hepatitis B surface antigen and hepatitis C antibodies

were negative. Autoimmune markers such as antinuclear antibodies (ANA), anti-smooth muscle antibodies (ASMA), and anti-mitochondrial antibodies (AMA) were also negative. There was no laboratory evidence suggesting hemochromatosis.

Diagnostic paracentesis revealed ascitic fluid with a high serum-ascites albumin gradient (SAAG), which is consistent with portal hypertension. A liver biopsy was initially planned after correction of coagulation parameters.

During evaluation, upper gastrointestinal endoscopy revealed Grade I–II esophageal varices. In addition, mucosal changes including atrophic folds and scalloping were noted in the second part of the duodenum, raising suspicion for celiac disease.

A biopsy from the second part of the duodenum showed marked villous atrophy with increased intraepithelial lymphocytes. Serological analysis revealed strongly positive anti-tissue transglutaminase IgA antibodies and anti-endomysial antibodies, confirming the diagnosis of celiac disease.

The patient was started on a strict gluten-free diet. During follow-up after three months, there was significant improvement in clinical symptoms and laboratory parameters. Hemoglobin levels, platelet counts, and white blood cell counts improved considerably. Liver enzyme levels also normalized. Imaging studies demonstrated a noticeable reduction in splenomegaly and ascites.

## 4. Discussion

Celiac disease is a chronic immune-mediated disorder of the small intestine triggered by dietary gluten. It occurs in genetically susceptible individuals and results in inflammatory damage to the intestinal mucosa. Although the disease primarily affects the small intestine, a variety of extraintestinal manifestations have been described.

The diagnosis of celiac disease is usually established based on clinical presentation, serologic testing for specific antibodies, histopathological examination of duodenal biopsy samples, and improvement following a gluten-free diet.

Patients with celiac disease may present with classical gastrointestinal symptoms such as chronic diarrhea, weight loss, abdominal discomfort, and malabsorption. However, many patients exhibit atypical or extraintestinal manifestations including anemia, metabolic bone disease, neurological symptoms, and autoimmune disorders.

Hepatic involvement in celiac disease has been widely reported. Several studies have described a spectrum of liver abnormalities ranging from mild elevations in liver enzymes to autoimmune liver diseases. The most common hepatic manifestation is mild hypertransaminasemia, often referred to as “celiac hepatitis,” which typically resolves after adherence to a gluten-free diet.

Previous research has also suggested a relationship between celiac disease and autoimmune liver disorders. Studies by various authors have indicated that screening for celiac disease may be beneficial in patients with unexplained liver disease, particularly when autoimmune conditions or cryptogenic cirrhosis are suspected.

The occurrence of idiopathic portal hypertension in association with celiac disease is extremely uncommon. Only a few cases have been described in the literature. The exact pathophysiological mechanism underlying this association remains uncertain, although immune-mediated vascular injury or chronic inflammatory processes have been proposed as possible explanations.

In the present case, the patient exhibited severe portal hypertension with hypersplenism. Remarkably, the patient’s clinical condition improved substantially after initiation of a gluten-free diet. The improvement in hematological parameters, reduction in splenomegaly, and resolution of ascites suggest that untreated celiac disease may have contributed to the development of portal hypertension.

This case highlights the importance of considering celiac disease as a potential underlying cause in patients presenting with unexplained portal hypertension. Early diagnosis and treatment may significantly improve clinical outcomes and reduce the need for invasive procedures such as liver biopsy.

The association between celiac disease and idiopathic portal hypertension is rare but clinically important. Physicians should maintain a high level of suspicion for celiac disease in patients presenting with unexplained portal hypertension, especially when other common causes have been excluded. Early recognition and adherence to a gluten-free diet can result in significant clinical improvement and may prevent unnecessary diagnostic interventions.

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