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Case Report

Celiac disease associated non cirrhotic portal hypertension with acute variceal bleeding: a rare case report

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ABSTRACT

The interplay between the intestine and liver, commonly referred to as the gut–liver axis, plays an important role in the development of various immune-mediated and metabolic conditions. Among these, celiac disease has emerged as a potential cause of hepatic abnormalities that may clinically resemble autoimmune hepatitis, complicating the diagnostic process. We describe the case of a 45-year-old woman with a previous diagnosis of chronic liver disease and presumed autoimmune hepatitis who presented with recurrent hematemesis and hemodynamic instability. Emergency upper gastrointestinal endoscopy identified actively bleeding Grade III esophageal varices, which were managed with urgent endoscopic band ligation. Imaging studies demonstrated portal hypertension, splenomegaly, and a non-occlusive thrombus within the splenic vein. Further investigation revealed negative autoimmune serological markers alongside markedly increased tissue transglutaminase and deamidated gliadin peptide antibody levels, raising strong suspicion of celiac disease-associated hepatic involvement. This case underscores the need to investigate celiac disease in patients with otherwise unexplained chronic liver dysfunction, cryptogenic cirrhosis, or clinical features suggestive of seronegative autoimmune hepatitis. Establishing the correct diagnosis is essential to avoid potentially unnecessary immunosuppressive treatment and to guide appropriate long-term care, particularly through adherence to a strict gluten-free diet.

Keywords: Celiac disease, Autoimmune hepatitis, Celiac hepatitis, Portal hypertension, Esophageal varices, Gluten-free diet.

INTRODUCTION

Portal hypertension and its complications remain major causes of morbidity and mortality among patients with chronic liver disease. Variceal hemorrhage, in particular, represents a medical emergency that requires prompt diagnosis and management. While viral, metabolic, alcohol-related, and autoimmune causes account for most cases of cirrhosis, a subset of patients remain classified as having cryptogenic liver disease despite extensive evaluation.

Celiac disease is a chronic immune-mediated disorder triggered by gluten exposure in genetically susceptible individuals. Although traditionally associated with gastrointestinal symptoms such as diarrhea, malabsorption, and weight loss, many adults present with extraintestinal manifestations. Liver abnormalities are among the most frequently reported non-intestinal features and may range from mild transaminase elevation to advanced fibrosis and cirrhosis. Clinical overlap between autoimmune hepatitis and celiac disease-associated liver injury can present a significant diagnostic challenge.[1] Both conditions may manifest with elevated liver

enzymes, hypergammaglobulinemia, and nonspecific constitutional symptoms. In the absence of classical gastrointestinal complaints, celiac disease may remain undiagnosed for years, resulting in delayed treatment and progressive hepatic injury.

This study describes a patient with severe portal hypertension and recurrent variceal bleeding who had been treated for presumed autoimmune hepatitis for several years before a comprehensive evaluation revealed underlying celiac disease.

CASE REPORT

A 45-year-old woman presented to the emergency department with multiple episodes of hematemesis accompanied by hemoptysis. On arrival, she was pale, tachycardic, and hypotensive, requiring immediate resuscitative measures.

Her medical history was notable for chronic liver disease, portal hypertension, and esophageal varices. Several years earlier, she had been diagnosed elsewhere with autoimmune hepatitis and had received prolonged oral corticosteroid therapy. However, no previous diagnostic records, serological reports, or liver biopsy findings were available for review. She had undergone endoscopic variceal ligation in 2017 and required intermittent blood transfusions because of chronic anemia.

Following stabilization, the patient was admitted to the intensive care unit. Initial laboratory investigations demonstrated severe anemia, evidence of hypersplenism, and elevated blood urea levels consistent with upper gastrointestinal bleeding. Serological testing for hepatitis B, hepatitis C, and human immunodeficiency virus was negative.

Table 1: Haematology and routine biochemistry profiles.

Parameters	P(February 3)	P(February 4)	Reference range	Clinical significance
Hemoglobin	9.8	-	12-15 g/dl	Chronic iron deficiency anemia
Platelet count	136000	-	150000-450000	Thrombocytopenia secondary to hypersplenism
MCV	74	-	80-100 Fl	Microcytic anemia consistent with malabsorption
Blood urea	104	54	15-45mg/dl	Severe pre-renal azotemia from GI bleeding
BUN	48.88	25.38	7-20 mg/dl	Elevated due to upper GI bleed protein breakdown
SGOT	37	-	5-40 U/L	Mild baseline hepatocellular activity
SGPT	57	-	7-56 U/L	Mildly elevated transaminase levels

Urgent upper gastrointestinal endoscopy revealed large Grade III esophageal varices with active bleeding. Endoscopic band ligation was performed successfully, achieving complete hemostasis. In addition, severe portal hypertensive gastropathy was noted. Blunting of the duodenal folds was also observed, raising suspicion for underlying celiac disease.[2]

Abdominal ultrasonography demonstrated portal vein dilatation and significant splenomegaly. Subsequent portal Doppler examination revealed an irregular hepatic surface suggestive of cirrhosis. Portal venous flow remained patent; however, a small non-occlusive thrombus measuring approximately 12×9 mm was identified within the splenic vein near the splenic hilum.

Diagnostic paracentesis yielded a low-protein transudative ascitic fluid without evidence of spontaneous bacterial peritonitis.

Given the uncertainty regarding the etiology of her liver disease, an expanded autoimmune and celiac serological workup was undertaken. Autoimmune hepatitis markers, including ANA, AMA, and anti-LKM antibodies, were negative. In contrast, celiac serology demonstrated markedly elevated tissue transglutaminase IgA/IgG and deamidated gliadin peptide IgG antibody levels, confirming active celiac disease.

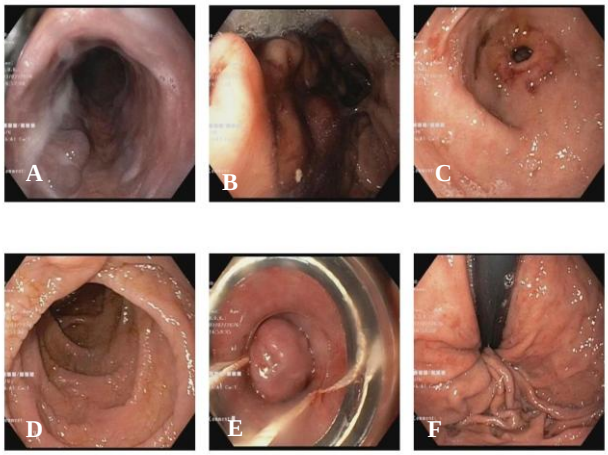


Figure 1 (A-F): Duodenal biopsy.

Table 2: Hepatic and enteric autoimmune panel.

Diagnostic biomarker	Patient value	Reference Range	Interpretation
ANA (Hep-2)	Negative (1:100)	Negative (<1:100)	Excludes classic Type 1 Autoimmune Hepatitis
Antimitochondrial antibody (AMA)	Negative (1:40)	Negative (<1:40)	Excludes Primary Biliary Cholangitis
Liver kidney microsomal (LKM)	Negative (1:40)	Negative (<1:40)	Excludes Type 2 Autoimmune Hepatitis
DGP AntibodyIgA	20.05 Units	<20.00 Units	Weak positive Celiac Enteropathy marker
DGP Antibody IgG	144.37 Units	<20.00 Units	Highly positive Celiac Enteropathy marker
tTG Antibody IgA	31.09 U/mL	<6.00 U/mL	Elevated; confirms gluten driven autoimmunity

During hospitalization, the patient received standard therapy for acute variceal bleeding and portal hypertension, including intravenous fluids, antibiotics, proton pump inhibitors, terlipressin, vitamin K supplementation, and supportive care. Following stabilization, she was transitioned to oral medications for long-term management of portal hypertension and ascites. Detailed counseling regarding lifelong adherence to a gluten-free diet was provided prior to discharge.

The patient was discharged in stable condition with plans for close gastroenterology follow-up.

DISCUSSION

This case illustrates an uncommon but clinically important presentation of celiac disease. Although hepatic involvement is well recognized in celiac disease, advanced portal hypertension with life-threatening variceal hemorrhage remains an unusual manifestation.[3] It also signifies how repetitive antigenic stimulation along the portal vein in untreated celiac disease can precipitate severe portal hypertension and variceal bleeding.[4]

The gut–liver axis provides a plausible explanation for the association between celiac disease and chronic liver injury. Persistent intestinal inflammation and increased intestinal permeability facilitate the translocation of microbial products and inflammatory mediators into the portal circulation. Continuous exposure of the liver to these factors may contribute to chronic hepatic inflammation and progressive fibrosis.[5]

A major challenge in this case was the clinical resemblance between autoimmune hepatitis and celiac disease-associated liver injury. The association between celiac disease and autoimmune hepatitis has been reported previously, including cases in which both conditions occurred in the same patient.[6] Both disorders share several laboratory and clinical features, including elevated transaminases and evidence of systemic immune activation.[7] Consequently, patients with undiagnosed celiac disease may be incorrectly classified as having seronegative autoimmune hepatitis.

The patient had received long-term corticosteroid therapy based on a presumptive diagnosis of autoimmune hepatitis. Despite years of treatment, progressive portal hypertension and cirrhosis developed, ultimately culminating in severe variceal bleeding. The absence of autoimmune markers and the presence of strongly positive celiac serology prompted reconsideration of the original diagnosis.[8]

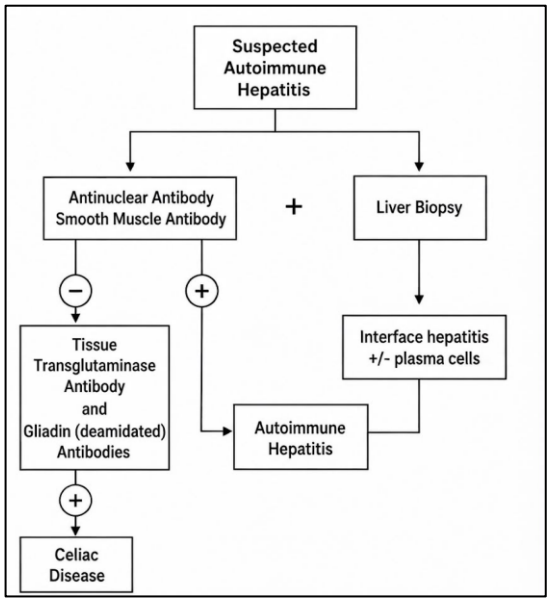


Figure 2: Diagnostic algorithm for evaluation of association between autoimmune hepatitis and celiac disease.[9]

Another noteworthy finding was the presence of splenic vein thrombosis. Several mechanisms may contribute to thrombotic risk in untreated celiac disease, including chronic systemic inflammation, endothelial dysfunction, nutritional deficiencies, and alterations in natural anticoagulant pathways. Although a direct causal

relationship cannot be definitively established in this case, active celiac disease may have contributed to the development of the thrombotic event.

Management of such patients requires a multidisciplinary approach. Acute bleeding episodes should be treated according to established protocols, including endoscopic therapy and pharmacological reduction of portal pressure. Equally important is identification and treatment of the underlying disorder. In patients with celiac disease, strict adherence to a gluten-free diet remains the cornerstone of therapy and may lead to improvement in hepatic abnormalities over time.

This case emphasizes the importance of maintaining a high index of suspicion for celiac disease in individuals with unexplained liver disease, particularly when autoimmune markers are absent or when clinical response to immunosuppressive therapy is inadequate.[10]

CONCLUSION

Celiac disease may rarely present with advanced portal hypertension and life-threatening variceal hemorrhage, masking its intestinal origin and mimicking autoimmune liver disease. This case demonstrates how celiac disease-associated hepatic injury can be mistaken for autoimmune hepatitis, resulting in prolonged and potentially unnecessary corticosteroid therapy.

Patients with cryptogenic cirrhosis, unexplained liver dysfunction, atypical autoimmune profiles, or poor response to immunosuppressive treatment should undergo evaluation for celiac disease. Early diagnosis and institution of a strict gluten-free diet may improve hepatic outcomes, reduce complications related to portal hypertension, and prevent progression of liver injury.

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