

Idiopathic Portal Hypertension and Celiac Disease: Two Case Reports

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Abstract

The association of celiac disease and idiopathic portal hypertension is a rare entity whose etiopathogenic mechanism has not yet been elucidated. We report two cases of celiac disease associated with idiopathic portal hypertension. In both cases celiac disease diagnosis was made based on the positivity of anti-tissue transglutaminase IgA and villous atrophy on the pathological study of duodenal biopsies. Idiopathic portal hypertension was retained after complete biological, radiological and endoscopic assessment excluding all possible causes that can manifest as portal hypertension. Both patients improved after gluten-free diet.

Keywords: Celiac disease, idiopathic portal hypertension, anti-tissue transglutaminase, villous atrophy, gluten-free diet.

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INTRODUCTION

Idiopathic portal hypertension (IPH) is a disorder generally considered to be of non-cirrhotic origin and of unknown etiology. It is clinically characterized by portal hypertension, splenomegaly and pancytopenia [1].

Celiac disease (CD) is a systemic condition resulting from an inappropriate immune response to gluten in genetically predisposed individuals [2].

The association of CD with IPH has been rarely reported in the literature [3]. We report two cases of association between celiac disease and idiopathic portal hypertension observed in our center.

CASE REPORTS

Case 1

A 24 year-old male patient, with no particular medical history, admitted in our center for portal hypertension syndrome revealed by gastrointestinal bleeding due to variceal esophageal rupture.

The clinical examination found an afebrile, pale patient with scleral icterus, conjunctiva and splenomegaly on abdominal examination.

The biological assessment revealed hypochromic microcytic anemia with hemoglobin at 8 g/dl, thrombocytopenia at 90000 platelets/mm³,

ferritinemia at 3 ng/ml, a serum albumin at 35 g/l and a complete hepatic assessment without abnormalities.

Abdominal ultrasonography coupled with Doppler showed dilatation of the portal vein at 16 mm with splenomegaly and a liver of normal morphology, without obstruction of the hepatic veins. The abdominal CT scan did not show any obstruction of the portal venous system.

Upper gastrointestinal endoscopy revealed grade II esophageal varices with red marks with reduced appearance of duodenal folds. The pathological examination of these biopsies showed sub-total villous atrophy. The diagnosis of celiac disease was confirmed by the positivity of anti-tissue transglutaminase IgA. The viral serologies B and C were negative. The search for anti-mitochondria, anti-smooth muscle, anti-nuclear and anti LKM-1 antibodies was also negative.

This was an idiopathic portal hypertension syndrome associated with celiac disease. A gluten-free diet, ligation and beta-blockers as secondary prophylaxis for rupture of esophageal varices were introduced.

With the gluten-free diet, the patient gained weight with significant improvement in hemoglobin and platelet count. Esophageal varices were eradicated after four sessions of ligations and there was no recurrence of upper gastrointestinal bleeding. The

patient continued to be asymptomatic on a gluten-free diet at last follow up, 18 months after presentation.

Case 2

A 40 year-old male patient, with no particular medical history, admitted in our center for an assessment of chronic diarrhea. The clinical examination found an afebrile patient, pale with discolored conjunctiva and splenomegaly on abdominal examination.

The biological assessment revealed pancytopenia with hypochromic microcytic anemia with hemoglobin at 9 g / dl, thrombocytopenia at 120000 / mm³, white blood cell at 3000 cells/mm³. Ferritinemia at 5 ng/ml, albuminemia at 33 g/l and a complete liver test without abnormalities.

Abdominal ultrasonography coupled with Doppler showed dilatation of the portal vein with splenomegaly and a liver of normal morphology, without obstruction of the hepatic veins.

Upper gastrointestinal endoscopy revealed grade II esophageal varices without red marks and a reduced appearance of duodenal folds. Pathological examination of the biopsies revealed sub-total villous atrophy. The diagnosis of celiac disease was confirmed by the positivity of anti-tissue transglutaminase IgA. The viral serologies B and C were negative. Auto-immune markers were also negative.

The diagnosis of idiopathic portal hypertension associated with celiac disease was thus retained. The patient was put under gluten-free diet and beta-blockers as primary prophylaxis for esophageal varices rupture.

On treatment with a gluten-free diet, the diarrhea subsided and the patient gained weight. The patient continued to be asymptomatic on a gluten-free diet at last follow up, one year after presentation.

DISCUSSION

Idiopathic portal hypertension (IPH) is a rare condition. It represents about 1% of portal hypertension [4]. It is a heterogeneous and multifactorial disorder with a potential genetic contribution [3]. It is defined by the presence of unexplained portal hypertension with a macroscopically and microscopically normal liver [5]. It is most commonly found in the Indian subcontinent and East Asia [3].

Hepatic involvement is common during celiac disease (CD) and is dominated by increased regressive transaminases under the gluten-free diet and by association with true autoimmune hepatopathies or fibrosis or hepatic cirrhosis [6]. The CD and IPH association is much more rare and little reported in the literature.

Improvement of portal hypertension with a gluten-free diet, a rare entity reported in some case, would imply a causal relationship between portal hypertension and increased inflammatory responses during celiac disease [7].

Autoimmune reactivity to other auto- antigens, apart from transglutaminase, has been reported in celiac disease such as actin, ganglioside, collagen, calreticulin and laponin. These abnormalities could be associated with specific clinical presentations or extra-intestinal manifestations of CD. There are several associations between CD and other pathologies, most of which are autoimmune. Cellular immunity is important with increased CD8 T cells and inflammatory cytokines found in the tissues involved [8,9]. IPH may be an aberrant immune response initiated by gluten exposure, suggesting that there is an immunological link between these two conditions and would require screening for CD in patients with IPH [3]. Repetitive antigenic stimulation along the portal system has been recognized as a possible cause of IPH [10].

CONCLUSION

Celiac disease and idiopathic portal hypertension is a rare association whose pathophysiological mechanism is unclear. CD could trigger the onset of IPH. Screening for CD during IPH would be necessary and the publication of other clinical cases would help to understand the etiopathogenesis of this association.

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