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CASE REPORT

Association of ulcerative colitis, celiac disease, primary sclerosing cholangitis and liver cirrhosis in a young male

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Abstract

Inflammatory bowel diseases (IBD), including ulcerative colitis, are chronic autoimmune conditions characterized by inflammation of the digestive system. The exact cause of IBD is unknown, but they often start during adolescence or early adulthood with symptoms such as urgency, rectal bleeding, diarrhea, abdominal pain and tenesmus. Primary sclerosing cholangitis and autoimmune hepatitis are recognized as co-occurring conditions associated with ulcerative colitis. However, the combination of ulcerative colitis, primary sclerosing cholangitis, liver cirrhosis, and celiac disease occurring concurrently has only been reported once before in a female patient. Here, we present the exceptional case of a Syrian adult male with all four of these conditions. This highlights the importance of screening for both celiac disease and cirrhosis in patients with ulcerative colitis and primary sclerosing cholangitis together, despite this combination of comorbidities is rare.

Introduction

Inflammatory bowel diseases (IBDs) are chronic conditions characterized by recurrent immune activation and inflammation within the gastrointestinal tract.[1,2] Although the precise etiology of IBD is not fully understood, it is believed to involve dysregulated intestinal immune responses to microbiota, which are influenced by environmental exposures in genetically susceptible individuals.[3] IBD has been reported to be associated with certain autoimmune hepatobiliary conditions, including autoimmune hepatitis, primary sclerosing cholangitis (PSC), and celiac disease.[4–7] We report here the rare case of a young male who was previously diagnosed with ulcerative colitis and later developed liver cirrhosis. Subsequently he was also diagnosed with PSC and celiac disease. This case emphasized the importance of screening for concurrent autoimmune conditions that can occur in patients with IBDs.

Case presentation

A 17-year-old male presented to our hospital with a history of abdominal distension lasting for 10 days. His past medical history was significant for ulcerative colitis, which was diagnosed 7 years ago and has been effectively managed with sulfasalazine, resulting in a good clinical response.

On admission, the physical examination revealed conjunctival pallor, with no other notable findings. Laboratory studies demonstrated pancytopenia, hypoalbuminemia, elevated levels of aspartate aminotransferase and alkaline phosphatase, normal bilirubin levels, and prolonged prothrombin time. Further laboratory investigations excluded hepatitis B, hepatitis C, autoimmune hepatitis, and Wilson's disease.

Abdominal ultrasonography showed a shrunken liver with irregular nodular contours and coarse parenchymal echo texture. The intrahepatic biliary system and common bile duct appeared normal. However, the portal vein was observed to be dilated at 17 mm, along with findings of splenomegaly and mild ascites. A diagnostic paracentesis was conducted, with the ascitic fluid analysis revealed a high serum-ascites albumin gradient and low protein content, which is consistent with cirrhosis.

Magnetic resonance cholangiopancreatography (MRCP) showed the presence of multifocal biliary strictures, which are typical of PSC. The common bile duct and gallbladder, however, appeared normal (Fig. 1). The upper endoscopy showed four esophageal varices, with two of them measuring over 5 mm in diameter and lacking cherry red spots, along with portal hypertensive gastropathy. The duodenum exhibited a scalloped appearance, and the biopsy showed Marsh 3b changes (Figure S1, Supporting information). The serum tissue transglutaminase IgA came back positive. During the colonoscopy, conducted to assess for ulcerative colitis activity, severe pancolitis with cecal ulcerations was observed.

In summary, this 17-year-old male with a known history of ulcerative colitis was found to have new diagnoses of PSC, liver cirrhosis, and celiac disease. He was stabilized during hospitalization and subsequently discharged with instructions for outpatient gastroenterology follow-up.

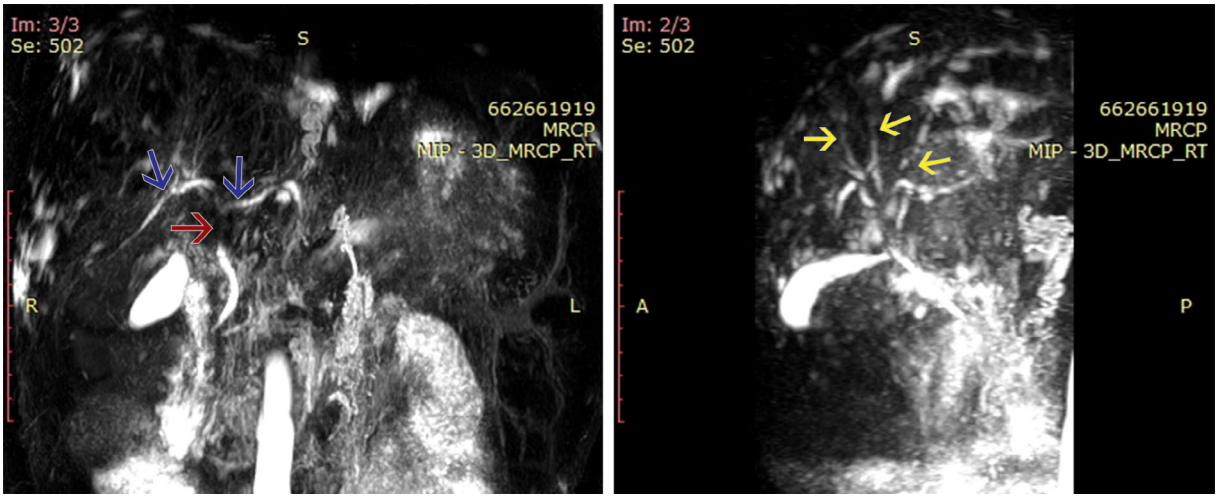

Figure 1 MRCP showing stenosis in the proximal part of the common biliary duct (red arrow), in the right and left hepatic ducts (blue arrows) and in the intrahepatic ducts (yellow arrows).

Discussion

This case highlights an association between ulcerative colitis and other autoimmune conditions, which was previously thought to be rare. However, with heightened clinical vigilance and screening, this association is being increasingly recognized.

Earlier reports documented cases of PSC and celiac disease co-occurring with IBD.[8] More recently, Habior et al. reported the case of two sisters with ulcerative colitis, PSC, and celiac disease. One of the sisters developed cirrhosis and cholangiocarcinoma.[9] Additionally, in a separate study conducted by Wurm *et al*., two cases were outlined involving individuals with concurrent ulcerative colitis, PSC, and celiac disease.[10]

A comprehensive review of 47 325 individuals with IBD revealed higher rates of comorbid autoimmune diseases compared with matched controls, with a greater association in ulcerative colitis than Crohn's disease.[11] The incidence of autoimmunity in IBD is estimated to be 11.11% *versus* 3.9% in the general population.[12] The most prevalent autoimmune conditions in patients with IBD are PSC, celiac disease, and psoriasis, although the exact relationship between these conditions remains debated.[13]

The occurrence of ulcerative colitis, PSC, celiac disease, and cirrhosis in a single patient is exceptionally rare, with only one prior documented case. This infrequent clustering of conditions warrants attention. Based on this potential association, we recommend screening for celiac disease and cirrhosis in patients with inflammatory bowel disease and concurrent PSC given this potential association. This proactive approach can aid in early detection and management of these comorbidities.

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Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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Supporting information

Additional supporting information may be found in the online version of this article at the publisher's website:

Figure S1. Duodenal biopsies of our patient showing MARCH 3b classification