

Case Report

Primary Biliary Cholangitis in a Patient with Ulcerative Colitis: About an Unusual Association

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Received: 12.03.2022

Accepted: 18.04.2022

Published: 12.05.2022

Journal homepage:<https://www.easpublisher.com>**Quick Response Code**

Abstract: Primary biliary cholangitis may be another hepatobiliary association of ulcerative colitis, and although less common than primary sclerosing cholangitis should be considered as a cause of cholestasis in these patients. Here, we report a case of A 26-year-old female patient.

Keywords: Ulcerative colitis, inflammatory bowel disease, Primary Biliary Cholangitis.

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INTRODCUTION

Inflammatory bowel diseases (IBD) can be associated with a variety of hepatobiliary disorders, reported both in Crohn's disease (CD) and ulcerative colitis (UC). The most common association described is that of the Primary Sclerosing Cholangitis (PSC) with IBD [1]. Primary Biliary Cholangitis (PBC) on the other hand, rarely occurs in patients with IBD, although it shares some clinical features with PSC such as suspected autoimmune response to bile duct epithelial cells. Only 30 cases have been reported to date. The aim of this article is to report a case of this rare association, in a 26-year-old female patient.

CASE REPORT

A 26-year-old female patient, with no known history of chronic inflammatory bowel disease or autoimmune pathology, was admitted to our hospital in 2007, with bloody diarrhoea. The patient reported a pattern of bloody diarrhoea episodes alternating with periods of remission, over the past 2 years. On admission, she presented with 10 stools per day, both during the day and at night, associated with a marked rectal syndrome consisting of tenesmus and cramps, as well as extra-intestinal manifestations such as arthralgia of the major and medium-sized joints with an inflammatory like. On physical examination, the patient was asthenic with obvious muco-cutaneous pallor. Her

biological check-up revealed a microcytic hypochromic anaemia with a haemoglobin level of 10, her white blood cells count was around 12,000 with a neutrophilic predominance. Her C-reactive protein was 45 and her sedimentation rate 50. The rest of her check-up, i.e. in terms of liver and kidney function, was normal. Endoscopic examination of the rectum revealed signs of severity such as deepening ulcerations exposing the submucosa with bridge-like detachment. Based on these findings, the patient was diagnosed with UC and started on intravenous corticosteroid therapy, withdrawn due to lack of therapeutic improvement, classified as cortico-resistant and managed with ciclosporin. Her abdominal symptoms were drastically improved, and her diarrhea was resolved. The subsequent maintenance therapy was azathioprine 2.5mg/kg/d. The patient's symptoms have since been well controlled and she was followed as an outpatient with a good clinical, biological and endoscopic response.

Three years later, biological cholestasis was discovered incidentally during azathioprine monitoring and the patient is admitted to our hospital for further exploring. She had no history of drug abuse or significant alcohol consumption prior to her admission. She complained of skin itching, and general fatigue with no other symptoms such as jaundice, abdominal pain or diarrhoea. Abdominal examination revealed hepatomegaly with no splenomegaly. She did not exhibit any signs indicating the co-existence of other

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autoimmune diseases, such as Sjögren syndrome, chronic thyroiditis, and rheumatoid arthritis. Her work-up on admission revealed cholestasis with elevated alkaline phosphatase (PAL), and gammaglutamyl transpeptidase (GGT) levels (GGT: 462 and PAL: 168). On the other hand, serum aspartate aminotransferase, alanine aminotransferase and bilirubin levels were correct. Serum immunoglobulin IgM levels were elevated. Serologic makers for hepatitis B and hepatitis C viruses were negative. Serum anti-M2 antibody was positive as well as serum anti SP-100 antibodies. Ultrasonography and Hepatic MRI did not reveal any intra or extrahepatic biliary abnormalities, but showed a hepatic angioma straddling segment II and III. The patient underwent a liver biopsy, which showed non-suppurative destructive lesions of the small bile ducts at the anatomopathological examination. The patient began therapy with ursodeoxycholic acid (900 mg daily), and she is being followed as an outpatient for the past year showing a good response to the therapy.

DISCUSSION

Ulcerative colitis (UC) is an inflammatory disease of the colonic mucosa. It is believed to result from a complex series of interactions between predisposing genes, the environmental factors and the immune system. While the exact aetiology and pathogenesis of UC is not known to date, the involvement of the immune system appears to be essential [1]. UC and CD are usually associated with a range of hepatobiliary disorders, which are considered to be extra-intestinal manifestations of inflammatory bowel disease, and may occur at any time throughout the natural history of the disease [2]. Although, PSC and Non Alcoholic Fatty Liver Disease (NAFLD) are the two most common hepatobiliary disorders found in patients with inflammatory bowel disease (IBD). Less frequently, other IBD-related hepatobiliary manifestations can be seen, including cholelithiasis, granulomatous hepatitis, hepatic amyloidosis and primary biliary cirrhosis (PBC) [3].

PBC is an autoimmune disease of the liver characterised by chronic inflammatory damage and obliteration of the intrahepatic bile ducts, as well as infiltration of lymphocytes and plasma cells into the portal tract. It is a chronic disease, that can progress to fibrosis that may lead to cirrhosis and end-stage liver disease [2]. It is thought to be the result of a combination of multiple genetic factors and overlapping environmental factors [4]. PBC usually affects middle-aged women with a sex ratio of 10:1 (females to males), and the mean age at diagnosis being approximately 57.5 years. The patients who have been reported in literature with the association of both diseases have shown different features. The sex ratio for PBC in these cases was lower (2:1), thus affecting more males, and the mean age was younger, similar to that of UC [5, 6]. The symptoms of PBC usually occur during the active phase of UC, suggesting that PBC activity evolves in tandem

with UC. Therefore, leading most authors to consider PBC as an extra-intestinal (hepatobiliary) manifestation of UC [5].

The exact nature of the relationship between PBC and UC remains uncertain as only a few cases have been reported to this date. Nevertheless, some common characteristics have been displayed.

- **Both CBP and UC are known to be autoimmune diseases:** The autoimmune character of PBC is suggested by increased levels of immunoglobulin M, abnormal T cells infiltrating the liver, immune complex formation, high circulating autoantibody titers as well as its association with other autoimmune diseases [4]. Ulcerative colitis also meets some of the requirements for being considered an autoimmune disease. including the presence of autoantibodies to antigens expressed by enteric epithelial cells and leukocytes, the identification of HLA haplotypes, the response to corticosteroid therapy and the association with autoimmune diseases [7].
- **Genetics also have their say:** The involvement of genes located on the short arm of chromosome 6 (HLA class II) with both UC and PBC may imply that these inflammation regulating genes have pathological implications in both diseases [7].

The largest cohort in literature by Liberal et al, described 6 patients with the association. The UC in these patients, was also found to be mild with limited bowel lesions, usually a proctitis or left colitis, compared to patients with PSC, and were treated with Sulfasalazine as a first therapy choice [6, 8].

The identification of these patients is of utmost importance as UDCA treatment improves biochemical and histological markers of disease activity and lengthens the transplant-free survival time for a significant proportion of patients with diagnosed PBC [1].

CONCLUSION

In conclusion, the association of PBC with IBD, especially UC, while rare with only 25-30 sporadic cases reported so far, is real. Therefore, not only PSC, NAFLD or drug-induced hepatobiliary disorders but also PBC should be taken into account in the differential diagnosis of hepatobiliary diseases among UC patients with unexplained cholestasis.

REFERENCES

1. Liberal, R., Gaspar, R., Lopes, S., & Macedo, G. (2020). Primary biliary cholangitis in patients with inflammatory bowel disease. *Clinics and Research in Hepatology and Gastroenterology*, 44(1), e5-e9.
2. Polychronopoulou, E., Lygoura, V., Gatselis, N. K., & Dalekos, G. N. (2017). Increased cholestatic enzymes in two patients with long-term history of

- ulcerative colitis: consider primary biliary cholangitis not always primary sclerosing cholangitis. *Case Reports*, 2017, bcr-2017.
3. Fousekis, F. S., Theopistos, V. I., Katsanos, K. H., Tsianos, E. V., & Christodoulou, D. K. (2018). Hepatobiliary manifestations and complications in inflammatory bowel disease: A review. *Gastroenterology Research*, 11(2), 83-94.
 4. Al-Harthy, N., & Kumagi, T. (2012). Natural history and management of primary biliary cirrhosis. *Hepatic medicine: evidence and research*, 4, 61-71.
 5. Xiao, W. B., & Liu, Y. L. (2003). Primary biliary cirrhosis and ulcerative colitis: a case report and review of literature. *World journal of gastroenterology: WJG*, 9(4), 878-880.
 6. Tada, F., Abe, M., Nunoi, H., Azemoto, N., Mashiba, T., Furukawa, S., ... & Onji, M. (2011). Ulcerative colitis complicated with primary biliary cirrhosis. *Internal medicine*, 50(20), 2323-2327.
 7. Koulentaki, M. (1999). [No title found]. *Dig Dis Sci*, 44(10):, 1953-1956.
 8. Losurdo, G., Brescia, I. V., Lillo, C., Mezzapesa, M., Barone, M., Principi, M., ... & Rendina, M. (2021). Liver involvement in inflammatory bowel disease: What should the clinician know?. *World journal of hepatology*, 13(11), 1534-1551.

Cite This Article: S. Boulajaad, M. El Bouatmani, A. Ait Errami, S. Oubaha, Z. Samlani, K. Krati (2022). Primary Biliary Cholangitis in a Patient with Ulcerative Colitis: About an Unusual Association. *East African Scholars J Med Surg*, 4(5), 107-109