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Case report-Olgu sunumu

Co-existence of primary biliary cirrhosis and ulcerative colitis

Ülseratif kolit ve primer bilier siroz birlikteliği

Sabiye Akbulut*, Firdevs Topal, Özlem Yönem, Birol Bostancı, Musa Akoğlu

Department of Gastroenterology (S. Akbulut, MD., Prof. M. Akoğlu, MD.), Department of Gastrointestinal Surgery (Prof. B. Bostancı, MD.), Kartal Koşuyolu Teaching and Research Hospital, TR-34846 İstanbul, Gastroenterology Clinic (F. Topal, MD.), Çankırı State Hospital, TR-18100 Çankırı, Department of Gastroenterology, (Assoc. Prof. Ö. Yönem, MD.), Cumhuriyet University School of Medicine, TR-58140 Sivas

Abstract

A sixty-four years old female patient who had ulcerative colitis for 7 years ago and still under treatment was admitted to the clinic for her refractory pruritus. Laboratory tests revealed elevated liver function tests. Anti-mitochondrial antibody was positive. Her liver biopsy was consistent with primary biliary cirrhosis showing infiltration of portal areas with lymphocytes and histiocytes and convenient treatment was initiated. We present this case report because of the rare co-existence of ulcerative colitis and primary biliary cirrhosis.

Keywords: Ulcerative colitis, primary biliary cirrhosis

Özet

Altmış dört yaşında, yedi yıl önce ülseratif kolit tanısı konan ve hâlen tedavisi devam etmekte olan kadın hasta geçmeyen kaşıntı semptomu ile kliniğe kabul edildi. Yapılan tetkiklerde karaciğer fonksiyon testleri yüksek bulundu. Anti-mitokondrial antikor pozitifti, karaciğer biyopsisinde portal alanların lenfosit ve histiyositlerle infiltre olduğu raporlanarak primer bilier siroz tanısı ile tedavisine başlandı. Ülseratif kolit ve primer bilier siroz hastalığının birlikte nadir görülmesi nedeniyle sunmaktayız.

Anahtar sözcükler: Ülseratif kolit, primer bilier Siroz

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***Corresponding author**

Sabiye Akbulut, MD., Gastroenteroloji Anabilim Dalı, Kartal Koşuyolu Yüksek İhtisas Eğitim ve Araştırma Hastanesi, TR-34846 İstanbul. E-mail: sabiye4@hotmail.com

Introduction

Ulcerative colitis may be seen with some hepatobiliary diseases such as primary sclerosing cholangitis (PSC), cholangiocarcinoma and autoimmune hepatitis. Primary sclerosing cholangitis is the most seen disease with ulcerative colitis by approximately 5% [1-3]. Ulcerative colitis with primary biliary cirrhosis is an extremely uncommon situation and a few numbers of patients are reported in the literature as case reports [3-15]. We would like to present the presence of primary biliary cirrhosis developed in a case who had ulcerative colitis diagnosed seven years ago.

Case report

A sixty-four years old female patient was referred with a complaint of pruritus which developed 2 months ago. On her complaints of the bloody defecation with mucus for 6-7 times a day, the diagnosis of ulcerative colitis had been established and mesalazine tablet

with 3 g/day had been introduced. The bloody diarrhea with mucus had been regressed as a result of this treatment that was still continued. A pruritus was begun about 2 months ago which become refractory and she was referred to our clinic. She did not have any connective tissue disease or ulcerative colitis in her past medical history. She also did not have a story of drug use that will affect the hepatobiliary system except mesalazine. On the physical examination, her skin and sclera's were in a mild icteric appearance, and the liver was exceeding the costa border about 2 cm. There was not any pathological finding on examination of the other systems. Her liver functional tests, especially alkaline phosphatase (ALP) was found to be high on the routine examination, and she was hospitalized for investigation. Hemoglobin was 10.2 g/dL and leukocyte and platelet counts were in the normal range. The sedimentation was 64 mm/h and CRP was defined high with a value of 2.08 mg/dL (N: 0-0.8). Her other liver tests were: AST: 64 U/L (normal: 0-41 U/l), ALT: 98 U/L (normal: 0-40 U/L), alkaline phosphatase (ALP): 826 U/L (normal: 30-91 U/L), GGT: 124 U/L (normal: 0-61 U/L), albumin: 3.7 g/dL (normal: 3.4-4.8 g/dL) and serum direct bilirubin: 1.3mg/dL, serum total bilirubin: 2.3 mg/dL (normal: 0-1.5 mg/dL). Viral hepatitis panel was negative. On the immunological tests, anti-nuclear antibody (ANA), anti-smooth muscle antibody (ASMA), anti-liver and anti-kidney microsome antibody (LKM), soluble liver antigen (SLA), anti-endomysial antibodies (EMA) and p-ANCA were negative. IgM was high with a value of 485 mg/dl (normal: 46-304) while the levels of IgG, IgA and IgE were within a normal range. Anti-mitochondrial antibodies (AMA) was positive with a value >1/160 and anti-M2 antibody (AMA-M2) was also positive. Thyroid function tests were normal. There were not any pathological findings except the hepatomegaly on her abdominal ultrasonography. Gallbladder, common bile duct and intrahepatic bile ducts were evaluated as normal on the magnetic resonance cholangiopancreatography (MRCP). She was defecating 1-2 times a day with the blood and mucus defined in the feces. Ulcerative colitis was detected in the descending colon, sigmoid colon and the mild to moderate with rectal involvement on the total colonoscopy conducted to control and localize ulcerative colitis. Biopsy was taken from the sigmoid colon and rectum and crypts. Cryptic micro abscesses and crypt distortion was observed on the histopathology of these biopsies. Upper gastrointestinal endoscopy revealed that the esophageal mucosa and lumen were normal and there were not any esophageal varices. There was diffuse inflammation and the portal areas were infiltrated by the lymphocytes and histiocytes on the liver biopsy. The case was reported as "primary biliary cirrhosis, stage 1" as a result of the pathological examination. In addition to UC diagnosis, PBC diagnosis was also established and ursodeoxycholic acid (UDCA) was introduced as 15 mg/kg/day besides the Mesalazine therapy. On the follow-up conducted a month later, her complaint of pruritus was regressed and the outcomes of liver function test were reached to the normal ranges.

Discussion

Ulcerative colitis is a disease which may have extraintestinal complications. Wide range variations of the liver diseases may be seen in the patients with ulcerative colitis. A hepatobiliary disease, primary sclerosing cholangitis is the most seen concomitant disease by 5% in the patients with ulcerative colitis [16]. Cholangiocarcinoma may develop in about 10-15% of the patients with primary sclerosing cholangitis. In inflammatory bowel disease cases with primary sclerosing cholangitis, p-ANCA positivity is reported as 80% and HLA B8/D3 positivity higher than in the cases with PSC alone. PBC is another autoimmune disease similar to PSC characterized by itching, hyperbilirubinemia and increase in the levels of lg M. AMA is found in approximately 95% of the patients with a negative value of 5%. However PBC in the patients with UC is extremely uncommon and reported as case reports in the literature [4]. PBC with UC has been first reported in 1985 [6]. Since then few numbers of case reports had been published [3-15]. PBC generally affects the middle-aged females more than males. In a study from England, F/M rate was stated as 9:1 and the mean age as 57.5 [17]. In another study conducted in Europe, the female/male ratio was 10:1, while the mean age was in a range between 47 and 59 [18].

The prevalence of PBS in the patients with UC is 30 times higher than in the general population [5, 10]. In the studies conducted, PBC rarely develops prior to UC and as in our study, it may emerge by years after the diagnosis of UC [14]. PBC had developed 7 years after the diagnosis of UC in our patient. In the literature review, UC is seen more common with a mild activity in the UC patients with PBC. Moreover, the location of UC is usually reported as proctitis in these patients. In a study, pancolitis was reported in 2 cases and proctitis in 5 cases among those with a known location [5]. UC of our case was in a mild activity with a left sided location. Differentiation of PBC and PSC is important in these cases. Liver function tests and lgM may be found high in both of them. Liver biopsy and ERCP findings may not be found accurately distinctive. The best test used in the differential diagnosis is AMA. AMA may be always positive in PBC, while it is always negative in PSC [19]. UC is in a mild activity in both of them, but unlike PBC, UC usually involves the pancolitis in PSC [14]. Although the mechanism of the relationship between the ulcerative colitis and primary biliary cirrhosis cannot completely be explained, possible common mechanisms are tried to be explained by the association among infective, genetic and immunological factors [4, 16]. Genetics plays an important role in both diseases. The association of UC and PBC with genes located on the short arm chromosome 6 (HLA class II) could suggest that these regulatory genes of inflammation have pathogenetic importance in both disease [20]. The common pathogenic mechanism of the autoimmune diseases with ulcerative colitis is tried to be explained by involvement of the immune system, lymphocyte infiltration in the portal area, antibody reacting with the bile ducts and existence of the circulating immune complexes [5].

In conclusion, primary biliary cirrhosis with ulcerative colitis is an uncommon condition. AMA-M2 positivity and elevated liver function tests are the criteria used in the differential diagnosis of primary sclerosing cholangitis and primary biliary cirrhosis. Primary biliary cirrhosis should also be considered in the differential diagnosis of the hepatobiliary diseases in the patients with ulcerative colitis.

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