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Cirrhosis and intestinal B-cell lymphoma: two entities that are rarely associated with celiac disease

Siroz ve intestinal lenfoma; çölyak hastalığı ile ilişkili iki nadir durum

To the Editor,

Celiac disease (CD) is an immune-mediated permanent small bowel disorder triggered by the ingestion of gluten-containing food. Although this disease primarily affects the gut, many other tissues and organs may be affected in at least 20-30% of patients, as shown in recent studies on the association of CD and cryptogenic cirrhosis (1,2). Malignant intestinal lymphomas are mainly enteropathy-type T-cell lymphomas (ETCLs), defined as an independent category in the World Health Organization's classification, which may complicate CD in patients, whereas intestinal B-cell lymphomas associated with CD are seen less commonly (3,4).

A 62-year-old female was admitted to our hospital with malaise. Ten months before, she was diagno-

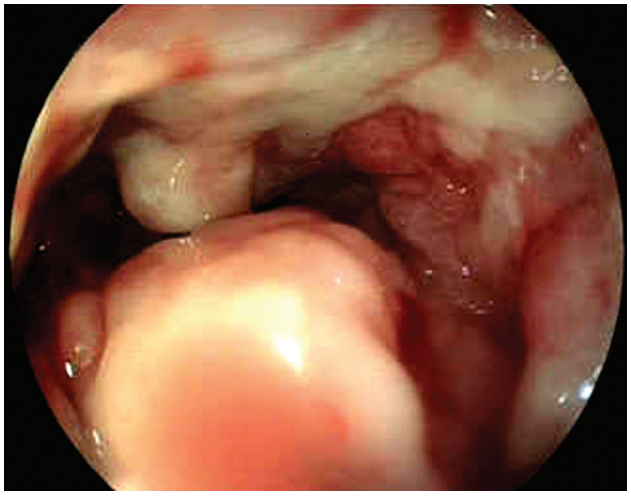

Figure 1. Endoscopic view of postbulbar area showing ulcerations.

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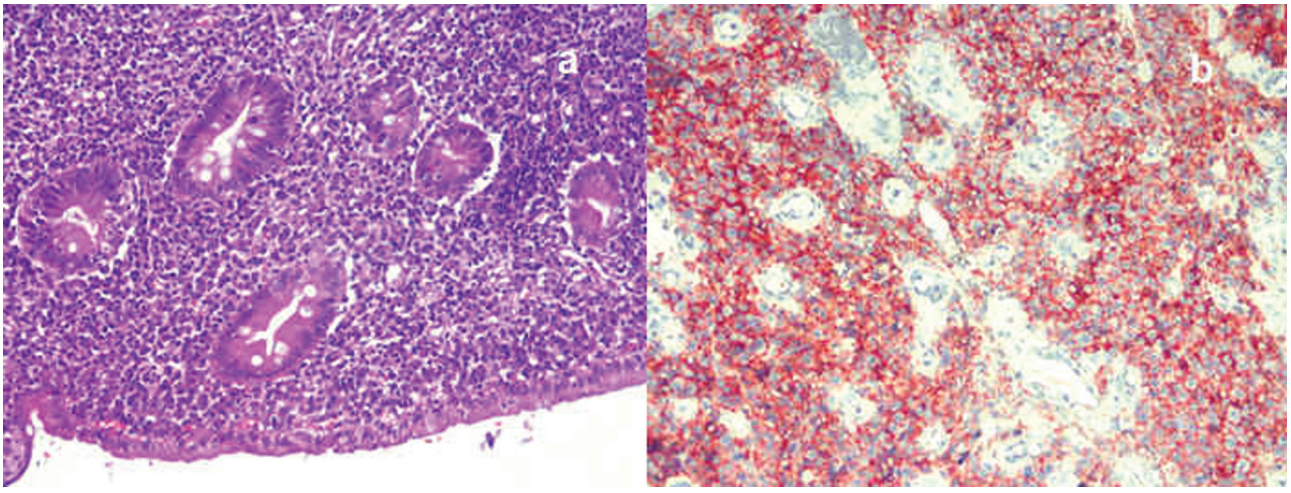

Figure 2. CD 38 and CD 79 immunostaining of biopsy specimen.

sed with cirrhosis and CD. An investigation including autoimmune, metabolic and virological marker tests for the etiology of cirrhosis was carried out, but findings did not suggest a specific condition. CD was thus thought to be responsible for the development of cirrhosis.

On her second admission, the patient appeared pale. An upper gastrointestinal endoscopy was performed following her complaints of intractable nausea and vomiting, and granular spots were observed in the post-bulbar area. The mucosa appeared fragile, and ulcerations surrounding the whole lumen were observed (Figure 1). Multiple biopsies were taken, and histological examination revealed CD 38 and CD 79 with positive high-grade diffuse large B-cell lymphoma (Figure 2). Computerized tomography (CT) of the thorax and abdomen showed hepatosplenomegaly, lymphadenopathies around the celiac truncus and mild atelectasis of the right lung on the diaphragm border. The patient was prescribed the R-CHOP protocol (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone) and improvement was seen after the first intake. Endoscopic findings showed remarkable improvement after the patient was treated with four rounds of chemotherapy (Figure 3).

The patient reported in this study was diagnosed with CD and associated cirrhosis. Although it is evident that cirrhosis increases the risk of developing hepatic malignancies (5), studies still lack information about the risk of developing extrahepatic malignancies in cirrhotic patients. Sorensen et al. (6) found an increased risk for hepatic and extrahepatic cancer in patients with cirrhosis, but part of this risk was attributable to alcohol and tobacco use. In this case, CD itself is a strong risk factor for the development of intestinal lymphoma, but the presence of cirrhosis in conjunction with CD might increase the development likelihood to a relatively rare form (diffuse large B-cell lymphoma).

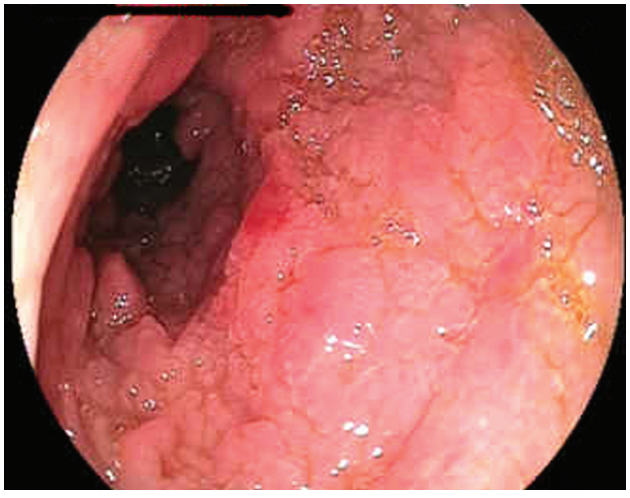

Figure 3. Endoscopic view of postbulbar area after chemotherapy.

In conclusion, recurrence of a patient's gastrointestinal complaints despite strict adherence to a gluten-free diet should raise suspicion of the development of a CD-related comorbidity such as an intestinal malignancy. Although ETCLs are seen more commonly in patients with CD, it should not be too surprising to diagnose B-cell lymphomas in the context of the increasing number of reports in the literature.

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Lymphoma-like presentation of hepatocellular carcinoma

Lemfoma benzeri hepatoselüler karsinoma vakası

To the Editor,

Hepatocellular carcinoma (HCC) is the fifth most common cancer in the world, with 500.000-1.000.000 new cases per year, causing 600.000 deaths globally per year (1,2). Late HCC usually metastasizes to regional lymph nodes and lungs (3).

In this letter, we describe a patient with no known prior liver disease who was diagnosed as HCC and lymphangitis carcinomatosis of the lung secondary to HCC and metastasis to the lungs, mediastinal, cervical and paraaortic lymph nodes, and bilateral adrenal glands.

A 39-year-old male patient applied to our clinic with complaints of weight loss and epigastric and substernal pain. His mother and daughter had died due to hepatitis B cirrhosis.

On admission, body temperature was 38.2°C and pulse was 120/minute. His physical examination revealed bilateral posterior cervical painless lymphadenopathies (LAPs); the liver was palpated at the costal border and dullness at the space of Traube. Hepatitis B virus (HBV) DNA level was 1.67 x 1.000.000 IU/L, and alpha-fetoprotein (AFP) was 3.53 ng/ml.

Hepatomegaly and multiple hypoechoic and hyperechoic masses with indeterminate borders were detected by abdominal ultrasonography. Cervical ultrasonography showed multiple LAPs at the upper cervical chain and supraclavicular region.

Upper abdomen magnetic resonance imaging (MRI) findings were T1 hypointense and T2 hyperintense multiple liver masses in different segments of the liver. Diameter of the largest mass was 8 cm. Ascites was present around the caudate lobe. A lobulated contoured LAP package, which was pressing the pancreas head, was detected in the paraaortic region. Bilateral, hypointense, nodular masses measuring 2 cm were also detected in the surrenals. Radiologic diagnosis was metastatic liver disease.

Thorax tomography revealed bilateral hilar LAP

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