sure to IFN and the appearance of symptoms may be variable. Symptoms generally resolve following the cessation of the drug (3). IFN-α therapy may induce choreiform movements due to affecting the dopaminergic pathways. It may act as a dopamine antagonist in the long term and may cause choreic movements by dysfunction of basal ganglia-thalamocortical loops (6).

In conclusion, movement disorders can be observed as rare complications of PEG-IFN-α. Physicians should be aware of these rare neurologic side effects during therapy.

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Clinical remission after strict gluten-free diet in a patient with celiac disease, advanced cryptogenic cirrhosis and splenic atrophy

Çölyak hastalığı, ilerlemiş kriptojenik siroz ve splenik atrofisi olan olguda glutensiz diyet sonrası klinik remisyon sağlanması

To the Editor,

Splenic atrophy and liver cirrhosis are known complications of celiac disease (CD). We report a case with CD associated with decompensated cryptogenic cirrhosis and splenic atrophy who was withdrawn from the liver transplantation schedule after adhering to a strict gluten-free diet.

A 58-year-old man with a history of intermittent diarrhea for 10 years and of abdominal swelling and edema in the legs for six months was admitted to our clinic in September 2009. The patient reported having 10 to 12 defecations without blood per day for the last two months. There was no relevant family, alcohol, or drug history. On the physical examination, there were no pathological findings except muscle weakness, tense ascites and bilateral pretibial edema. Laboratory investigation revealed the following: hematocrit: 26.8%, hemoglobin: 8.5 g/dl, platelets: 384000/mm^3, white blood cells: 5060/m^3, aspartate aminotransferase (AST): 106 U/dL, alanine aminotransferase (ALT): 69 U/dl, alkaline phosphatase: 403 U/dl, gamma glutamyl transferase: 93 U/dl, total bilirubin: 0.6 mg/dl, total protein: 6.1 g/dl, albumin: 2 g/dl, prothrombin time: 18.6 seconds, international norma-

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lized ratio: 1.4, and calcium: 7.8 mg/dl. Peripheral blood smear showed Howell-Jolly bodies, target cells and acanthocytes. Serum markers for hepatitis B virus (HBV) and HCV as well as serum antinuclear antibodies, smooth muscle antibodies, antimitochondrial antibodies, and anti LKM-1 antibodies were negative. Anti gliadin IgA, anti gliadin IgG, anti endomysium IgA, and anti endomysium IgG class antibodies and anti-tissue transglutaminase were strongly positive.

Abdominal ultrasound and abdominal computerized tomography (CT) showed atrophy of the right lobe, hypertrophy of the left lobe and caudate lobe and irregularity on the liver surface, enlargement of the portal vein, large volume of ascites in the abdomen, and splenic atrophy (6.8 cm in length). Esophagogastroduodenoscopy showed esophageal varices (F2) and portal gastropathy associated with duodenal edema. All these findings were compatible with decompensated cirrhosis associated with splenic atrophy. A small bowel mucosal biopsy specimen showed near-total villous atrophy and mixed inflammatory cell infiltration in the mucosa, which was consistent with CD (Figure 1). The patient was referred to the liver transplantation center and scheduled for orthotopic liver transplantation. He was also placed on a gluten-free diet, and furosemide 40 mg per day (oral) was prescribed. Within four weeks, the symptoms of diarrhea and ascites had disappeared, and the patient was followed without diuretic administration. One year after a strict gluten-free diet, the patient was completely symptom-free, and laboratory investigation revealed a hematocrit of 40% and hemoglobin of 13.5 g/dl. Platelets were 235000/mm^3, white blood cells 5890/m^3, AST 93 U/dl, ALT 99 U/dl, alkaline phosphatase 244 U/dl, gamma glutamyl transferase 157 U/dl, total bilirubin 1.1 mg/dl, total protein 7.5 g/dl, albumin 3.6 g/dl, prothrombin time 11.5 seconds, international normalized ratio 0.96, and calcium 8.6 mg/dl. A follow-up endoscopic small bowel biopsy was refused by the patient. Follow-up abdominal CT showed cirrhotic liver and splenic atrophy without ascites (Figure 2). One year later, there was no change in the diameter of the spleen compared to initial CT findings.

Modest elevation in serum aminotransferase levels is common in untreated CD, occurring in 15%-55% of patients (1,2). It may also be associated with severe forms of liver disease (3). The mechanisms underlying liver injury in CD are poorly understood. The increased intestinal mucosal permeability may facilitate the entry of toxins, antigens and inflammatory substances to the portal circulation, and these mediators may have a role in the liver involvement seen in patients with CD.

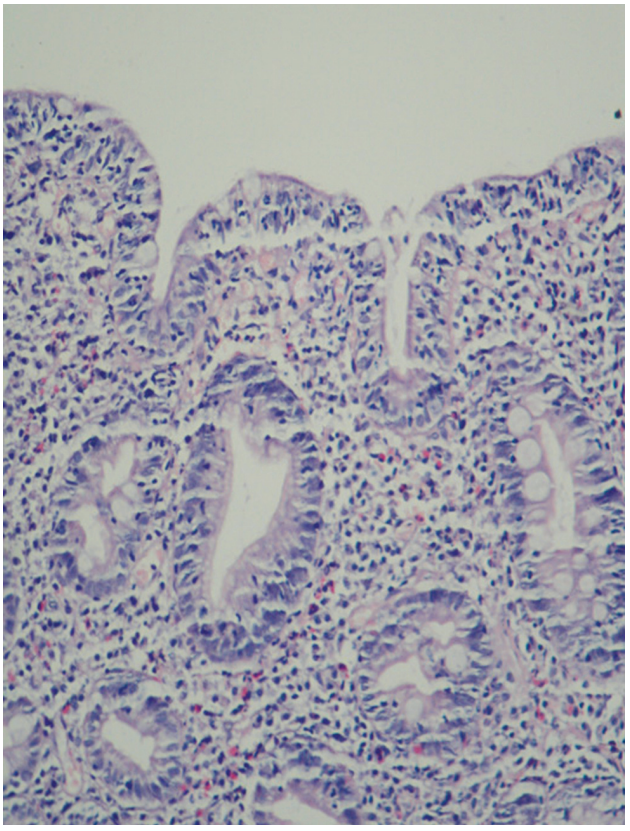

Figure 1. A small bowel mucosal biopsy specimen showed near-total villous atrophy, crypt hyperplasia and increased intraepithelial mixed inflammatory cell infiltration (H&E, x200).

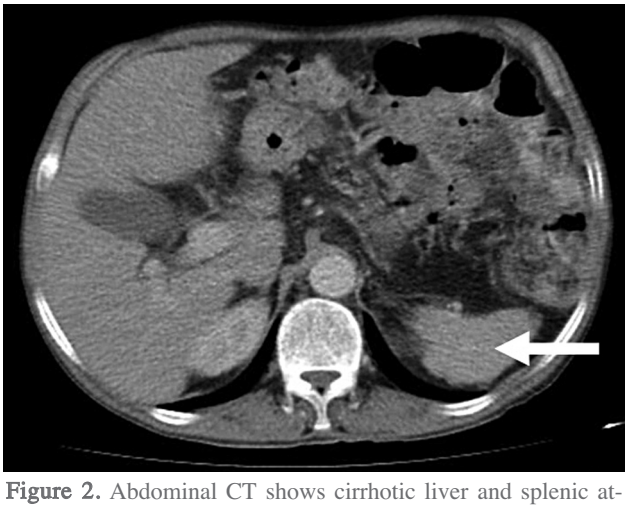

Figure 2. Abdominal CT shows cirrhotic liver and splenic atrophy (arrow) after adherence to a strict gluten-free diet for one year.

Histologically, only minimal hepatic steatosis or reactive non-specific hepatitis is seen in cases of CD in which liver enzyme levels are slightly increased (2). It can be hypothesized that untreated CD with subclinical hepatic involvement can lead to a more serious liver disease in some cases in the long term.

A gluten-free diet leads to normalization of serum transaminases in 75%-95% of patients with CD, usually within a year of good adherence to the diet (1,2). A response to dietary treatment and an improvement in clinical manifestations and laboratory abnormalities after a gluten-free diet was described in three patients with liver cirrhosis who had been referred for consideration of liver transplantation (4).

The association between CD and hypofunction of the spleen is well recognized. The pathogenesis of hyposplenism in CD remains unknown. The degree of hyposplenism is usually related to the severity of jejunal mucosal atrophy. Two types of hyposplenism were described with CD: functional hyposplenism and splenic atrophy (5). Functional hyposplenism may be reversible with gluten withdrawal and subsequent decrease in circulating immune complexes reflecting as a decrease in anti-transglutaminase, anti-endomysial and anti-gliadin antibody titers, while splenic atrophy is usually irreversible. Howell-Jolly bodies, target cells and acanthocytes are considered indicators of splenic dysfunction. The spleen is 10±1.5 cm in length in adults (6).

In case of cryptogenic liver cirrhosis associated with CD and splenic atrophy, classical hematological manifestations of hypersplenism like thrombocytopenia and leukopenia may not be seen. A strict gluten-free diet should be the first option of treatment.

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An unusual presentation of Seckel syndrome: Fatty liver

Seckel sendromunun nadir bir prezentasyonu: Yağlı karaciğer

To the Editor,

Seckel syndrome (SCKL) [OMIM 210600], belonging to the group of osteodysplastic primordial "dwarfism" first defined by Seckel in 1960, is characterized by microcephaly, dwarfism of prenatal onset, proportionate pre- and post-natal growth retardation, and a typical "bird-headed" profile (beaked nose, receding forehead, prominent eyes, and micrognathia) (1). The exact pathogenesis of

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