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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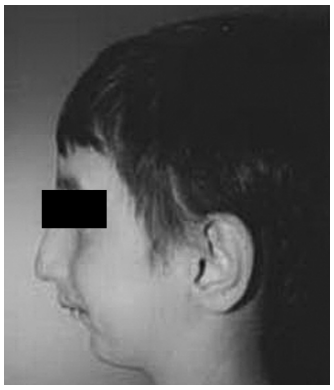

Figure 1. Image of the patient showing micrognathia, malocclusion of the teeth, receding forehead, large prominent "bird-like" nose deformities, and small, lobeless ears.

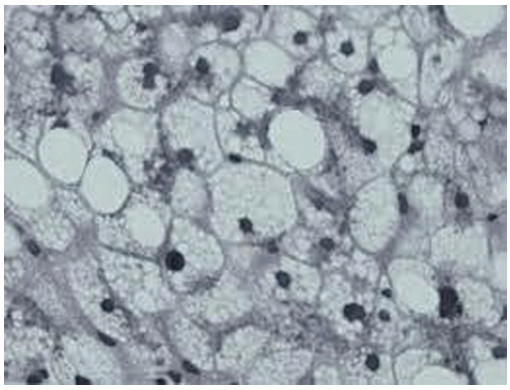

Figure 2. Fatty changes in liver biopsy (x100).

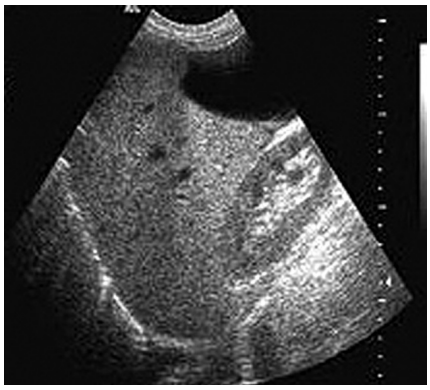

Figure 3. Ultrasonography of liver showing steatosis.

the disease currently remains unknown, but the most probable cause is an autosomal recessive inheritance (2). Since SCKL was first described, 170 cases have been reported, but there is considerable heterogeneity in their clinical characteristics. Seckel syndrome is a disorder of markedly reduced brain and body size, and is associated with defective ATR-dependent DNA damage signalling (3). SCKL is a genetically heterogeneous disorder, with variable chromosomal instability, fractures and hematological disorders (4).

Our case was the first child of a non-consanguineous healthy 32-year-old mother and 34-year-old father and was born at 28 weeks gestation by cesarean section, weighing 750 g. All family members were healthy. The medical history revealed low birth weight and mental and motor developmental delay. His parents were non-consanguineous. He was severely microcephalic with a receding forehead, hypertelorism, beaked nose, low-set ears, micrognathia, and downward-slanting palpebral fissures. There was no hepatosplenomegaly or lymphadenopathy. The patient had mental retardation and a distinct high-pitched voice. On physical examination, the weight, height and head circumference were below the 3rd percentile. His height was 95 cm (-5-1 SD), weight 13 kg, and head circumference 43 cm (-6.6 SD). The short stature was proportionate, that is, the leg-to-trunk length ratio was normal. The average Z score (standard deviation score) in SCKL syndrome is -7 SD (between -4 and -14) (5). Our case's Z score was –7.97. Micrognathia, malocclusion of the teeth, receding forehead, large prominent "bird-like" nose deformity, and small, lobeless ears were present in our patient (Figure 1). Scoliosis was determined in vertebral radiographs. His liver biopsy showed fatty changes (Figure 2). Brain computerized tomography was normal. His chromosome analysis was 46, XY. His bone age was 5 years. His intelligence quotient (IQ) score was consistent with his chronological age. There was no known history of exposure to radiation or toxins, and the family history was negative for fatty liver or Seckel syndrome. Complete blood count showed white blood cells (WBC) 3 x 10^9/L, hemoglobin 16 g/L, platelet count 150 x 10^9/L, blasts 4%, myelocytes 5%, metamyelocytes 3%, bands 28%, neutrophils 33%, lymphocytes 24%, and monocytes 3%. Laboratory findings showed hemoglobin level 11 g/dl, urea nitrogen 60 mg/dl, creatinine 1.1 mg/dl, sodium 134 mEq/L, potassium 5.8 mEq/L, calcium 9 mg/dl, albumin 3.5 g/dl, uric acid 6.3 mg/dl, and phosphate 5.6 mg/dl. The ultrasonography of the liver showed steatosis (Figure 3). Growth hormone level was 4.10 mg/ml (normal range: 0.5-6 mg/ml). Viral marker results were as follows: anti-hepatitis A virus (HAV) IgM and IgG negative, HbsAg negative, anti-Hbs positive, and anti-HCV negative. Total cholesterol was 125 mg/dl (normal), triglyceride 75 mg/dl (normal), low density lipoprotein (LDL)-cholesterol 75 mg/dl (normal), very low density lipoprotein (VLDL)-cholesterol 20 mg/dl (normal), and high density lipoprotein (HDL)-cholesterol 30 mg/dl (normal). Liver function tests revealed aspartate aminotransferase (SGOT) 86 U/L, alanine aminotransferase (SGPT) 70 U/L, gamma-glutamyl transferase (GGT) 28 U/L, direct biliru-

bin 0.2 mg/dl, and indirect bilirubin 0.7 mg/dl. The metabolic diseases screening test results were: blood ammonium level 50 mmol/L (normal), reducing substance in the urine (negative), and tandem mass spectrometry (normal). Echocardiography was normal.

To our knowledge, fatty liver degeneration in Seckel syndrome has not been reported previously. There is considerable heterogeneity in the clinical characteristics in Seckel syndrome (5-7). As a result of this case report, we suggest also investigating patients for hepatosteatosis.

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An unusual case of polycythemia vera with a complication of pancreatic pseudocyst

Polistemia vera komplikasyonu olarak gelişen bir nadir pankreatik psödokist vakası

To the Editor,

Polycythemia vera (PV) is a myeloproliferative disorder that can be complicated with thrombosis. The rate of thrombosis is as high as 50% (1-7). Budd-Chiari syndrome and portal, splenic, or mesenteric vein thrombosis are some examples of the major thrombotic events that can be seen in PV patients. Portal hypertension and hypersplenism can complicate as a result (8,9).

Pancreatic pseudocyst is defined as a fluid collection >4 weeks old and surrounded by a defined wall (10). Most pseudocysts include sterile material. There is no need for treatment for an asymptomatic pseudocyst (11,12).

In this article, we report a case of PV with a rare secondary complication of pancreatitis.

A 46-year-old male had symptoms of abdominal pain, pruritus and dyspnea. During the previous four months, he had noted these complaints with increasing severity. Physical examination revealed hepatomegaly and splenomegaly. He had also rales in the left hemithorax.

On laboratory examination, hematocrit level was 57.02 x10^3/µl (37.0–50.0), leukocyte count 17.6 x10^3/µl (4.0–11.0), granulocyte count 17.0 x10^3/µl (9.0–17.0), and platelet count 734 x10^3/µl (150–400), and JAK2 mutation was present. These findings were consistent with PV. There were no signs of another clotting disorder. Phlebotomy and hydroxyurea therapy was started.

Computed tomography (CT) revealed portal vein

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