



Abdominal pain in a patient with sickle cell disease with multiple complications

Çoklu komplikasyonlarla birlikte orak hücre hastalığı olan bir hastada karın ağrısı

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Abstract

Sickle cell disease is an inherited autosomal recessive hemoglobinopathy. Acute abdominal pain is the cause of hospitalization in 10% of patients with sickle cell disease and usually occurs during vaso-occlusion or distal tissue ischemia. Determining the etiology of abdominal pain is very difficult in these patients because it is associated with several rare diagnoses, such as pancreatitis and splenic abscess in some patients. We represent a 14-year-old boy with sickle cell disease who was hospitalized due to acute abdominal pain and indicated multiple and scarce disturbances in the spleen and hepatobiliary system.

Keywords: Abdominal pain; cholelithiasis; pancreatitis; sickle cell disease; splenic abscess

Öz

Orak hücre hastalığı, otozomal resesif kalıtım geçişli bir hemoglobi-nopatidir. Orak hücre hastalığı olan hastaların %10'unda hastaneye yatış nedeni akut karın ağrısıdır ve bu durum genellikle vazo-oklüzif tıkanma ya da enfarktif kriz, distal doku iskemisi esnasında ortaya çıkar. Bu hastalarda, karın ağrısının etiyolojisinin tanısı çok zordur, Çünkü bazı hastalarda pankreatit ve dalak absesi gibi bazı nadir durumlarla ilişkili olabilir. Burada, akut karın ağrısı nedeni ile yatırılan, orak hücre hastalığı olan ve dalak ve hepatobiliyer sistemde nadir görülen çoklu bozukluklar gösteren 14 yaşında bir erkek çocuğunu sunuyoruz.

Anahtar sözcükler: Dalak absesi; karın ağrısı; kolelitiazis; orak hücre hastalığı; pankreatitis

Introduction

Sickle cell disease (SCD) is an autosomal recessive abnormality of the β -globin chain of hemoglobin; its most common clinical manifestation is anemia due to chronic hemolysis (1).

Sickle cell disease can affect any part of the body. Nearly 10% of patients with SCD are hospitalized due to acute abdominal pain. It usually occurs during vaso-occlusion or distal tissue ischemia (2, 3).

The common causes of abdominal pain include bone marrow hyperplasia, mesenteric and retroperitoneal lymphadenitis, ischemia and infarction of the abdominal or-

gans such as the spleen, liver, small intestine, colon, and pancreas. Other causes include viral hepatitis, acute cholecystitis, cholelithiasis, and peptic ulcers (4). Available treatments include transfusions, hydroxycarbamide, stem cell transplantation, and several new therapeutic options such as gene therapy and gene editing in development (5, 6).

This article describes a case of acute abdominal pain in a 14-year-old boy with SCD who presented with multiple and scarce disturbances in the spleen and hepatopancreatobiliary system.

Case

A 14-year-old boy, known to have sickle cell anemia, was

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admitted with a history of abdominal pain and four vomiting episodes, which were non-bilious and non-bloody. He had no recent illness and no history of fever or chills and trauma. His medical history indicated that he was diagnosed as having sickle cell anemia and was put on hemoglobin electrophoresis at the age of 6 months, and he received his last transfusion 2 years prior to the study. His family history indicated that his brother also had SCD.

On physical examination, constant and non-positional abdominal pain, not aggravated by food, was located in the left upper quadrant (LUQ) and epigastric area. The abdomen was not distended, and no mass could be felt. The other physical examination findings were unremarkable.

His vital signs were as follows: temperature 37.6°C, pulse rate 100 beats/minute, blood pressure 80/50 mm Hg, and respiratory rate 20 breaths/minute. At the time of admission, the patient's complete blood count findings were as follows: white blood cell (WBC), 18 000/mm³ with 85% polymorphonuclear cells and 13% lymphocytes; hemoglobin (Hgb), 7.9 g/dL; platelets, 140 000/mm³; and red blood cell, 1 900 000/mm³.

He was then admitted to the hospital, and an abdominal ultrasound was performed in which cholelithiasis was revealed. During the next few days, his abdominal pain persisted, and the patient gradually became febrile and dyspneic; thus, imaging studies were conducted.

Chest X-ray showed elevation of the left side of the diaphragm (Fig. 1).

Based on these findings and the patient's symptoms, emergency abdominal ultrasound revealed pancreatitis, cholelithiasis with several stones, and enlarged spleen in which a mass was observed on one-third of its lower part, which was suggestive of splenic infarction, inflammatory mass, and/or tumoral lesions. Free fluid was observed in the pelvic and peritoneal cavities. Doppler ultrasound revealed splenomegaly and non-visible vessels in the spleen parenchyma.

A computed tomography (CT) scan with oral and intravenous (IV) contrasts of the abdominopelvic region showed splenic abscess and splenomegaly associated with infarction. Further, free fluid was observed in the pleural cavity (Fig. 2).

The laboratory values were as follows: WBC, 21,200/mm³; neutrophils, 82%; lymphocytes, 16%; monocytes, 2%; Hgb, 7.2 mg/dL; C-reactive protein (CRP), 123 mg/dL; lipase, 395 U/L; gamma-glutamyl transferase (GGT),

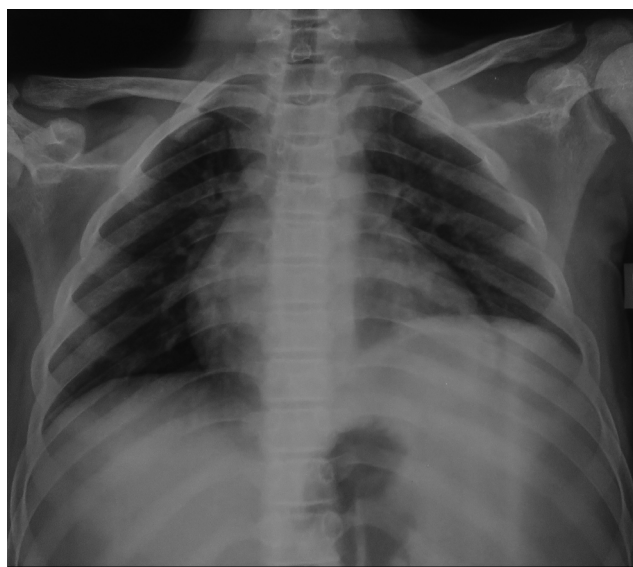


Figure 1. Chest X-ray showed left diaphragm elevation



Figure 2. Abdominal CT scan showed infarcts and abscess regions in spleen with splenomegaly

48 U/L; albumin, 3.42 g/dL; erythrocyte sedimentation rate (ESR), 21 mm/h; total bilirubin, 3.1 mg/dL; indirect bilirubin, 0.5 mg/dL; sodium (Na), 125 meq/L; and BS, 147 mg/dL. The urine and blood culture findings were negative, and vein blood gas (VBG) test findings were as follows: pH, 7.34; PCO₂, 53 mm Hg; PO₂, 44 mmHg; and HCO₃, 19 mmol/L. The other laboratory data, such as the Serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), phosphorus (P), potassium (K), cholesterol, triglycerols (TG), amylase, blood urea nitrogen (BUN), and creatinine (Cr) levels, were within the normal limits.

The patient was initially managed conservatively with analgesic administration, IV fluid infusion, and oral nutrition, as tolerated. With the presentation of symptoms such as fever, abdominal tenderness, and guarding be-

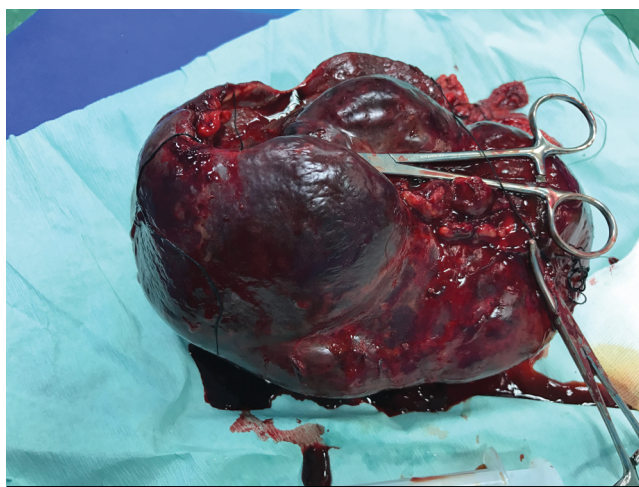


Figure 3. Splenectomy image that showed multiple infarct and abscess

havior, and laboratory and imaging assay findings, he was treated with IV broad spectrum antibiotherapy, blood transfusion, and total parenteral nutrition; splenectomy and cholecystectomy were considered (Fig. 3). Nine days after admission and the patient becoming stabilized, he was transferred to the surgical unit.

During the surgery, a midline skin incision from the subxiphoid to the subumbilical region was performed. In the peritoneal cavity, multiple severe adhesions were noted between the enlarged spleen and the omentum, transverse colon, and stomach. After adhesion excision, splenectomy was performed. The gallbladder serosal surface was also incised to retract it in the cephalic direction to explore Calot's triangle. Cholecystectomy was completed. Broad spectrum antibiotic therapy was provided, and packed red cells and fresh frozen plasma (FFP) were transfused. Oral nourishment was established. Eight days after surgery, the patient was discharged without complications and followed up in the outpatient clinic. The pathology was reported as splenic necrosis and abscess.

Informed consent was obtained from the patient and his parents who participated in this study.

Discussion

Sickle cell disease is one of the common inherited hemoglobinopathies, which results from a single change in one amino acid, valine, instead of glutamic acid (7). Many patients with SCD sometimes experience acute abdominal pain with fever, leukocytosis, and mild jaundice (4); these findings serve as difficult diagnostic and management problems, because the clinical presentation may mimic an acute surgical condition (8).

When patients with known SCD present with symptoms of abdominal pain, the characteristics of the symptoms, precipitating events, WBC count, bilirubin level, and fever should be compared with those in previous crises. A deviation from previous patterns suggests an illness caused by problems other than sickle cell crisis (3).

Pancreatitis in children with SCD is a complication of cholelithiasis (9). Cholelithiasis is one of the most common manifestations of SCD in the hepatobiliary system (2). The prevalence is high in patients with SCD and increases with age (4). It is observed in 26–58% of these patients (1) as a result of elevated serum bilirubin levels due to erythrocyte hemolysis, and bilirubin is in the form of its calcium salt (1, 10). Patients with cholelithiasis can be symptomatic or asymptomatic (2, 6, 10). Abdominal pain is the most common finding in symptomatic patients with cholelithiasis, which might localize in the epigastric area or RUQ (10). Cholelithiasis can cause cholecystitis, choledocholithiasis, cholangitis, and pancreatitis, yielding high morbidity rates in patients with SCD (2).

Although pancreatitis during childhood is not common, the high incidence of cholelithiasis in patients with SCD should be considered in the differential diagnosis of SCD with abdominal pain (4, 9). Cholecystectomy is an appropriate treatment in symptomatic patients (2). However, the general condition of our patient worsened. Thus, it was necessary to evaluate more and pancreatitis was determined. Pancreatitis is treated conservatively with analgesic administration, antibiotherapy, intravenous fluid infusion, and sometimes blood transfusion (9). We also treated our patient in this manner.

Splenic abscess and infarction were also observed in this case. One of the earliest affected organs in SCD is the spleen (7). It is commonly enlarged during the first decade of life, but undergoes progressive atrophy thereafter as a result of multiple splenic infarctions leading to autosplenectomy (11). However, this does not always occur, and splenomegaly sometimes persists in an older age group (7, 11), making this group vulnerable to developing splenic complications necessitating splenectomy, such as splenic abscess and massive splenic infarction (7, 11). Splenic abscess occurs in 0.14–0.7% of necropsy specimens. It is very rare in patients with SCD (12). The early development of functional asplenia makes these patients prone to systemic infections and predisposes patients with SCD to the development of splenic abscess in the presence of splenic infarction (12). In a review of 173 patients with splenic abscess, 12% of the causes were related to hemoglobinopathies (7, 12).

The diagnosis of splenic abscess is very difficult in children who frequently present with fever because of other infective reasons and abdominal pain, which is commonly associated with vaso-occlusive crisis in SCD (12). In a review of 10 patients with splenic abscess and SCD, all presented with fever and abdominal pain (12); however, in our patient, who initially presented with abdominal pain and vomiting without fever, infective causes were less possible. Based on the first ultrasound, only cholelithiasis and splenomegaly were reported; however, the patient gradually became febrile, and the other diagnosis was made with the presence of persistent leukocytosis. The increased CRP level, high lipase level, and the findings of the second ultrasound indicated pancreatitis; thus, appropriate treatment was provided. However, as the patient's general condition worsened in addition to the presence of respiratory distress, the results of the chest X-ray, and tenderness in the LUQ, inflammatory lesions based on the second ultrasound became more possible.

Ahmed et al. (12) reported that splenic abscess should be considered when fever, abdominal pain, and tenderness and enlarged spleen were observed in patients with SCD. Thus, it was necessary to perform other imaging evaluations to confirm the diagnosis; a CT scan was performed because inflammation and infarction in the spleen were probable on the second ultrasound. Patients should be evaluated using CT scans if there are doubts with ultrasound findings because CT is more reliable (7, 12). In our patient, abdominal CT revealed splenic abscess. Some authors recommended aspiration under ultrasound guidance to distinguish between splenic infarction and abscess; however, we performed Doppler ultrasound in which splenic infarction was found (12).

There are two treatment options for splenic abscess: splenectomy and abscess drainage. It is suggested that the treatment of choice for splenic abscess in patients with SCD is splenectomy and antibiotherapy (7, 12) because the mortality rate is higher with percutaneous abscess drainage than with splenectomy (12). Our patient was treated with splenectomy. Laparoscopic surgery is mostly performed for cholelithiasis (1) but our patient underwent laparotomy because of the pancreatitis and splenic abscess.

The repeat abdominal ultrasound one week after surgery showed normal pancreas, liver, and biliary tree. Hydroxyurea was administered daily for the patient. In the follow-up, there was no iron overload, there was no recurrence of abdominal pain attacks, and monitoring was performed as follows; CBC monthly, ferritin every 3 months, cardiac and liver MRI annually (5, 6).

In patients with SCD and acute abdominal pain, vaso-occlusive crisis consequences should be considered in multiple gastrointestinal and hepatobiliary organs, especially in the spleen. Furthermore, spleen abscess may occur.

Informed Consent: Verbal informed consent was obtained from patients' parents who participated in this case.

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