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Gastroduodenal Perforation in a Child With Sickle Cell Anemia: A Case Report

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Abstract
Objectives: Gastroduodenal perforation (GDP) is a rare cause of acute abdominal pain in children.
Case Presentation: A 12-year-old boy presented with abdominal pain around the periumbilical and epigastric region on 7previous days. His familial history was significant for sickle cell anemia in the mother and minor thalassemia in the father. He had been diagnosed with sickle cell anemia at the age of 4 years and had received blood transfusion and hydroxyurea every month. At physical examination, a distended abdomen and decreased intestinal sounds were seen. Also, he had a history of cholecystectomy (for cholelithiasis). Fecal occult blood was detected, while no Helicobacter pylori antigen was present. Abdominal ultrasound examination revealed splenomegaly and evidence of free fluid. Surgical results showed bile discharge in the abdominal cavity, and perforation of duodenum and pylorus.
Conclusions: Since the symptoms of GDP in children are non-specific, especially in children with sickle cell anemia, it should be considered in these patients with abdominal pain.
Keywords: Peptic ulcer, Anemia, Sickle cell, Child

Introduction

Sickle cell anemia, known as the homozygous state for hemoglobin S, is prevalent in the Middle East, Africa, and India (1). A point mutation in the β-globin chain of hemoglobin, changing glutamic acid to valine, is the leading cause of the disease (1). Secondary gastroduodenal perforation (GDP) may rarely cause by some diseases such as sickle cell anemia. One of the common symptoms in these patients is abdominal pain, attributed to circulatory stasis and vascular occlusion (2,3). There are only a few reports regarding GDP in children with sickle cell anemia. On the other hand, GDP is one of the rare causes of acute abdominal pain in children and can lead to peritonitis and shock (4). The most frequent finding of GDP is extraluminal free air, and surgery is the necessary treatment (2).Here, we report a 12-yr-old boy who presented with abdominal pain and was diagnosed with sickle cell anemia and GDP.

Case Presentation

A 12-year-old boy who was presented with abdominal pain around the periumbilical and epigastric region on 7 previous days referred to Amirkola Children's Hospital, Babol, Iran. He had oral intolerance and biliary vomiting following eating. Loss of appetite and difficulty in defecation without fever or respiratory symptoms were reported during last week. He was the first child of his family and was born term. He had been diagnosed with sickle cell anemia at the age of 4 years by hemoglobin electrophoresis and had received blood transfusion and hydroxyurea every month. Moreover, he had a history of cholecystectomy (for cholelithiasis). His familial history was significant for sickle cell anemia in the mother and minor thalassemia in the father.

On admission, he was alert with mild dehydration, and his vital signs were as follows: pulse rate=100/min, respiratory rate =25/min, temperature=36/5c, blood pressure = 90/70 mm Hg.

Physical examination revealed that he was pale, the abdomen was distended, bowel sounds were hypoactive, and there was generalized tenderness, especially in the epigastric region. Splenomegaly was detected. The other examinations were unremarkable. Other laboratory analyses including C-reactive protein, erythrocyte sedimentation rate, blood sugar, blood urea nitrogen, amylase, lipase, creatinine, alkaline phosphatase, total protein, prothrombin time, albumin, international normalized ratio, partial thromboplastin time, calcium, phosphorus, and magnesium were in the normal range (Table 1).

Fecal occult blood was detected, while no helicobacter pylori antigen was present. Abdomen ultrasonography revealed splenomegaly and signs of free fluid in the abdomen, while the pancreas, liver, kidneys, and bladder were normal. X-ray examination of the chest and abdomen showed an air-fluid level below the diaphragm (Figure 1). Hydration as well as cefotaxime (50 mg/kg/q6h), metronidazole (10 mg/kg/q8h), and pantoprazole

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Table 1. Case's Laboratory Data on the Admission Time

CBC	
WBC	16.3×10^3 µL
RBC	3.63×10^6 µL
Hb	8.6 g/dL
HCT	26%
MCV	71.6 Fl
MCH	23.4 pg
MCHC	32.7 g/dL
PLT	242×10^3 µL
LFT	
AST	70 IU/L
ALT	68 IU/L
UA/ UC	
SG	1030
Blood	Positive
WBC	2-1
RBC	14-12
UC	Negative
VBG	
PH	7.38
PCO2	28 mm Hg
HCO3	18 mmol/L
Na	132 mEq/L
K	2.8 mEq/L

CBC: Complete blood count, WBC: White blood cell, RBC: Red blood cell, Hb: Hemoglobin, HCT: Hematocrit, MCV: Mean corpuscular volume, MCH: Mean corpuscular hemoglobin, MCHC: Mean corpuscular hemoglobin concentration, PLT: Platelet, AST: Aspartate aminotransferase, ALT: Alanine aminotransferase, Na: Natrium, K: Kalium, VBG: Venous blood gas, UA: Urine analysis, SG: Specific gravity, UC: Urine culture.

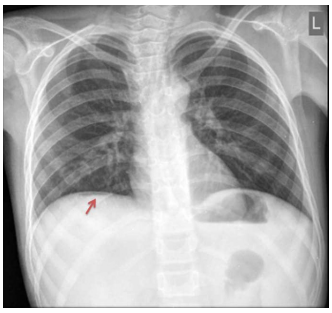

Figure 1. Free Air Under Right Diaphragm.

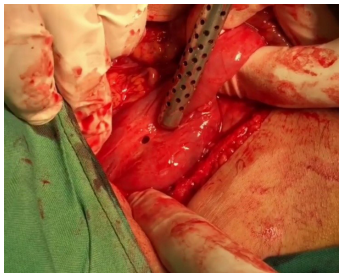

Figure 2. Perforation of Distal Stomach in a 12-Year Old Boy With the Sickle Cell Anemia.

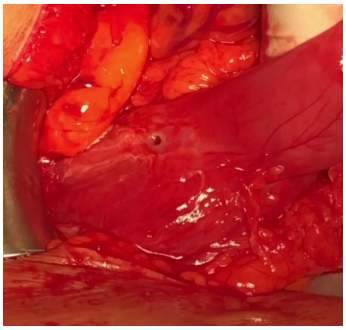

Figure 3. Perforation of Proximal Duodenum in a 12-Year Old Boy With the Sickle Cell Anemia.

(1 mg/kg/q12h) were administered via intravenous route. Because of the patient's emergency condition, no computed tomography (CT) scan of the abdomen and pelvis was performed, but a laparotomy was performed after emergency surgical consultation. The original diagnosis was gastrointestinal (GI) perforation. Surgical results indicated bile leakage into the abdominal cavity, and perforation of the duodenum and pylorus, which were repaired (Figure 2 and 3).

He was hospitalized for 12 days and discharged in good condition. One-month follow-up with upper GI endoscopy showed normal GI tract.

Discussion

This report was of a rare complication of sickle cell anemia, namely of GDP, in a 12-year-old boy. One of the most common symptoms of sickle cell anemia as a hemoglobinopathy is abdominal pain (3). Nearly 10% of these patients are hospitalized annually with abdominal pain. They are the result of vaso-occlusive or infarctive crisis (2). Abdominal vasoconstriction crisis is thought to be due to mesenteric vasoconstriction and may present with pain in other parts of the body or only as a complaint (5). It may occur singly or usually as part of a generalized vasoconstrictive crisis.

A study on 20 sickle cell anemia cases with abdominal pain suggested that the severity of pain was moderate to severe in 70% of patients. The most common symptoms associated with abdominal pain were vomiting (60%) and bone pain (45%). Abdominal pain occurred in one location in 70% of patients, with the epigastric and suprapubic regions being the most commonly affected (5). However, in these patients, abdominal pain is due to the vaso-occlusive crisis. It is clinically difficult to distinguish abdominal vaso-occlusive crises from other causes, leading to acute abdominal pain (5). Non-vaso-occlusive conditions that present with abdominal pain in sickle cell disease are cholelithiasis with or without cholecystitis, splenic infarction and abscess, acute splenic sequestration crisis, acute pancreatitis, acute appendicitis, peptic ulcer disease, hepatic infarction and abscess, hepatic crisis, ischemic colitis and mesenteric lymphadenitis (6).

Peptic ulcer disease is a cause of abdominal pain in

children with sickle cell anemia. Peptic ulcer disease is seen in 7.7%-35% of patients with sickle cell anemia (7). It is usually caused by the use of nonsteroidal anti-inflammatory drugs and helicobacter pylori infection (4), as well as can lead to GDP, which is a rare condition in children, caused mainly by ulcers (8). The most common site for gastric perforation is the pyloric area, whereas duodenal perforation involves the duodenal bulb (4). Symptoms of GDP in children are non-specific, and paraclinical evaluation is necessary. In this case, abdominal pain, biliary vomiting, abdominal distension, hypoperistaltic bowel and loss of appetite, leukocytosis, and splenomegaly occurred. Although the treatment of abdominal vaso-occlusive crisis is conservative, most of these attacks resolve spontaneously without surgical intervention. In this case, close observation, as well as laboratory and imaging evaluation, were initially performed. The abdominal pain worsened, and the abdominal X-ray revealed subdiaphragmatic free air, arguing for a diagnosis of GDP. Abdominal ultrasonography showed splenomegaly and signs of free fluid in the abdomen.

There are few studies on GDP in children with sickle cell anemia. Acıpayam and co-workers described a 14-year-old boy with homozygous sickle cell anemia and duodenal perforation who presented with abdominal pain, anorexia, and vomiting. He had no organomegaly, but abdominal tenderness was noted on palpation. Surgical intervention revealed a perforation in the second part of the duodenum (2). Johnson and colleagues described a 13-year-old boy with homozygous sickle cell anemia who presented with persistent epigastric pain, generalized abdominal distension, tachycardia, and febrile. Deep palpation of the abdomen was mildly tender. Intraoperative esophagogastroduodenoscopy confirmed the presence of perforation in the duodenum and pylorus. Of note, he had a history of taking daily naproxen along with narcotics for generalized pain. In contrast, the GDP in our case occurred with no known history of daily use of nonsteroidal anti-inflammatory drugs (9).

Sickle cell anemia is a homozygous and lifelong disease associated with hemolytic anemia and widespread organ damage (10). Patients suffer from various GI complications, including cholelithiasis, cholecystitis, acute pancreatitis, peptic ulcers, ischemic bowel disease, and pseudomembranous colitis (11). Peptic ulcers can lead to perforation, especially in more severe cases. Peptic ulcer disease in sickle cell anemia results in ischemia and mucosal hypoxemia because of episodes of abdominal vaso-occlusive crisis (11). This may lead to GDP in the absence of epigastric symptoms. However, there was no evidence of peptic ulcer based on surgical exploration and pathological findings in our case. Moreover, there was no evidence of peptic ulcer and helicobacter pylori infection on GI endoscopic follow-up. If the diagnosis was delayed, peritonitis and shock might occur. Our case was treated quickly, while surgical findings indicated bile leakage in the abdominal cavity and the duodenum and pylorus perforation. The critical point is that perforation has no specific symptoms, and acute abdominal pain can confuse the diagnosis.

Authors' Contribution

All authors contributed to patient management, collected the data, drafted the case report, and approved the final version of the manuscript.

Conflict of Interests

The authors declare that they have no conflict of interest.

Ethical Issues

Informed consent was obtained from the child's parents for publication of this report.

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