

Case Report

Acute pancreatitis in a child with sickle cell anemia

Manoj Kumar D.*, Komalatha Choppari, Suresh R. J. Thomas

Department of Pediatrics, Kamineni Institute of Medical Sciences, Narketpally, Nalgonda, Telangana, India

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*Correspondence:

Dr. Manoj Kumar D.,

E-mail: dr.manojkumar420@gmail.com

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ABSTRACT

Sickle cell disease (SCD) is a term used for a group of genetic disorders characterized by production of Hb "S". Sickle cell hemoglobinopathy occurs due to mutation of beta-globin gene situated on short arm of chromosome 11, where adenine is replaced by thymine in base of DNA coding for the amino acid in the sixth position in beta-globin chain. This leads to an amino acid change in beta chain of Hb molecule, from glutamic acid to valine. The result is profound change in the molecular stability and solubility of Hb "S". Authors are reporting a 8-year-old girl who is a known case of sickle cell disease presented with complaints of intermittent pain abdomen and vomiting since 30 days. Investigations revealed elevated pancreatic enzymes with radiological evidence of pancreatitis. Packed red blood cell transfusion and appropriate supportive therapy given and child recovered well.

Keywords: Acute pancreatitis, Sickle cell disease

INTRODUCTION

Acute pancreatitis, the most common pancreatic disorder in children, is increasing in incidence.¹ In children, blunt abdominal injuries, multisystem disease such as the hemolytic uremic syndrome and inflammatory bowel disease, biliary stones or microlithiasis, and drug toxicity are the most common etiologies. In sickle cell anemia, a vaso-occlusive or infarctive crisis often causes acute abdominal pain associated with ischemia and infarction.²

CASE REPORT

Among 8-year-old girl with sickle cell disease was admitted to the hospital with a 1 month history of intermittent abdominal pain in epigastric and left hypochondrium regions with radiation to left subscapular region, 5 episodes of non-projectile non bilious vomiting. The pain was of sufficient severity to have kept her awake the night prior to admission. There was no history of recent illness, fever, burning micturition, or trauma. Past history revealed that she was diagnosed at 9 months with sickle cell disease by hemoglobin

electrophoresis. She was repeatedly hospitalized since then for anemia and respiratory tract infections, and received about 3 packed RBC transfusions. She remained healthy subsequent to the last episode. Family history indicated that her parents were sickle cell traits. The child was well immunized and developmentally normal.

On physical examination, the patient's temperature was 99°F, heart rate was 94 beats/minute, respiratory rate was 20/minute, and blood pressure was 100/60 mm Hg. On abdominal examination, there was tenderness in epigastrium umbilical and left hypochondriac regions. The liver was palpable at 1 cm below right costal margin and spleen was not palpable. Bowel sounds were normal. The remainder of the physical examination was unremarkable. Preliminary investigations included a hemoglobin of 8.6gm/dL, a white blood cell count of 18,100/cu.mm and a platelet count of 4.5L. Serum electrolytes, glucose, and creatinine levels were normal. Laboratory values were elevated for total bilirubin, aspartate aminotransferase. Urine for bile salts and bile pigments were negative. Abdominal radiographs were normal, and an abdominal ultrasound demonstrated a tiny

gall bladder calculi and bulky pancreatic head. CECT abdomen was suggestive of acute edematous pancreatitis with hyperdense sludge in gall bladder.

Initial management consisted of intravenous fluids, inj. pantoprazole, inj. zofer, inj. piptaz, tab. combiflam, tab hydroxyurea, tab. fiolvite and oral fluids as tolerated. Abdominal pain continued with associated anorexia. Serum amylase and lipase levels measured after admission were elevated: 459 U/L (normal range: 40 to 80U/L) and 144 U/L (normal range: 40 to 240 U/L), respectively. At that time, oral intake was discontinued, and total inj. octreotide was started. Twenty four hours later, the patient's she improved clinically. Repeat abdominal ultrasound again showed biliary sludge with a normal pancreas, liver. As admission hemoglobin was 8.6gm/dL 10ml/kg of packed red blood cell transfusion was given, repeat haemoglobin 24 hours after transfusion was 10.8gm/dL. She progressed slowly to a full diet without any recurrence of her symptoms. Her pancreatic enzymes, total serum bilirubin and liver enzymes gradually decreased to normal levels over a week. She was continued with the same treatment and was discharged after 7 days. She was advised to continue tab Hydroxyurea 50mg and tab Folvate 5mg. She remained healthy at a follow-up visit 2 weeks later.



Figure 1: Image taken from the ultrasound scan showing the presence of gallstones within the gallbladder.

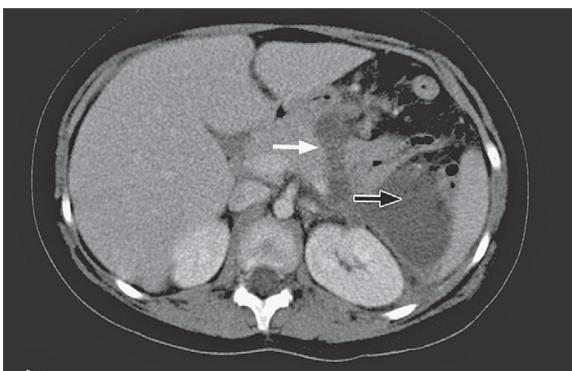


Figure 2: CECT abdomen of the same patient showing enlargement of pancreas suggestive of acute pancreatitis.

DISCUSSION

Abdominal pain in children with sickle cell anemia usually occurs during a vaso-occlusive or infarctive crisis and may be severe enough to mimic an acute surgical abdomen.³ Multiple factors causing this pain include: vertebral infarction with compression of nerve roots, mesenteric and retroperitoneal lymphadenitis with infarction, and occlusion of the blood supply to abdominal organs with ischemia and infarction.³ With the latter injury, target organs include the spleen, liver, and less commonly the small intestine and colon.⁴ Other causes of abdominal pain in patients with sickle cell anemia include viral hepatitis, acute cholecystitis, cholelithiasis, cholelithiasis, and peptic ulcer disease.

Surprisingly, pancreatitis is rarely included as a cause of abdominal pain in sickle cell crisis. In Serjeant's extensive review of the clinical manifestations of sickle cell disease, he states that the frequency of pancreatitis and its role in the abdominal painful crisis are entirely unknown.⁵ An extensive literature review disclosed no additional cases of pancreatitis in children with sickle cell disease.

Recently, the importance of acute pancreatitis presenting as a complication of multisystem diseases in children was recognized.¹ Diseases including Reye's syndrome, hemolytic-uremic syndrome, collagen vascular syndromes, sepsis, and shock accounted for 35% of all cases of pancreatitis. This was the most common cause of pancreatitis in children followed by idiopathic, traumatic, structural, drug-related, and metabolic diseases.²

A recent review of the gastrointestinal manifestations of hemolytic-uremic syndrome demonstrated a high incidence of pancreatitis with 66% of children displaying both clinical and laboratory evidence of pancreatitis during acute presentation.⁶ The authors hypothesized that the pancreatitis was due to endothelial damage that caused microthrombi formation and ischemia. Similarly, in the vaso-occlusive crisis of sickle cell anemia, the characteristic pathophysiological mechanism of organ injury is that of circulatory stasis with vascular occlusion and resultant ischemia.⁴ This likely initiates the activation of enzymes known to trigger the auto-digestion of the pancreas, which results in acute pancreatitis.⁷

In the case reported here, the diagnosis of pancreatitis was based on the clinical picture, the persistent elevation of pancreatic enzymes, and the subsequent finding of a bulky pancreas on abdominal ultrasound. The patient's abdominal pain gradually subsided with conservative treatment for acute pancreatitis. Other causes of pancreatitis including trauma, infection, and metabolic disease were not found in this patient. The anemia, reticulocytosis, elevated serum lactate dehydrogenase level noted at admission were all consistent with a vaso-occlusive crisis. In the absence of other etiological

factors, it must be assumed that the pancreatitis in this 8-year-old girl with sickle cell anemia was due to circulatory stasis and ischemia of the pancreas caused by an infarctive crisis. This case provides further evidence to support the fact that pancreatitis can occur in children who develop multi system diseases, especially those that are associated with ischemic injury'.

CONCLUSION

Efforts in looking for pancreatitis in patients with sickle cell anemia with abdominal pain will provide a more accurate assessment of its incidence in this multisystem disease and lead to improved patient care.

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