Case Report

Sickle cell anemia with perforated duodenal ulcer as a complication: when to suspect this rare entity?

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Abstract

Perforated duodenal peptic ulcers are often not considered when making a differential diagnosis of abdominal pain, especially in the context of sickle cell disease, and cases have not been frequently described in the literature. This study reports the case of a 14 year-old girl with sickle cell anemia complicated with duodenal ulcer perforation, focusing mainly on the imaging aspects. Abdominal CT should be considered as a method for this diagnosis and it requires the knowledge of this entity and its characteristic imaging findings.

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Introduction

Peptic ulcer disease (PUD) has a multifactorial origin, and it is considerably more common in adults than in children. The diagnosis of PUD has increased in the last few years due to improvements in endoscopic techniques used with this age group; however, studies that provide objective data about its incidence are still scarce [1,2]. It is estimated that the prevalence of the disease in large pediatric centers is 4 to 7 new cases per year [3]. Data from our pediatric service at the Escola Paulista de Medicina of the Universidade Federal of São Paulo reported by Kawakami et al. [4] showed that there were 7.2 diagnoses and/or year.

Duodenal perforation in children is an even rarer condition and has high mortality rates if not treated promptly. It is most commonly caused in this age group by Helicobacter pylori infection – responsible for 56.8%-83.1% of cases in children living in
the poorest regions of Brazil, and about 10% of cases in children under 10 years old [2] living in developed countries – or by systemic diseases such as Crohn’s disease, cystic fibrosis or sickle cell disease [5].

In the context of sickle cell disease, characterized by genetic alterations in the hemoglobin metabolism, there can be multiple causes of abdominal pain, with this symptom being most commonly attributed to vaso-occlusive crises (potentially associated with ischemic intestinal damage), paralytic ileus, cholecystitis or episodes of pulmonary sequestration [6,7]. Duodenal perforation due to peptic-ulcer disease is rarely considered as a differential diagnosis and is not often described in literature [8].

The present study reports the case of a pediatric patient with sickle cell anemia complicated by duodenal ulcer perforation, and aims to focus mainly on the imaging aspects of this diagnosis.

Case Summary

A thirteen-year-old female patient attended the emergency department with emesis not associated with her recent food consumption, intense abdominal pain that had started the day before, without factors that improved or aggravated the pain, and without fever. The patient was tachycardic, tachypneic, dyspneic and pale; the abdomen was semi-globose, flaccid, diffusely painful, and with no signs of peritoneal irritation. The patient had a history of sickle cell anemia with monthly blood transfusions, was submitted to a cholecystectomy when she was five years old and auto-splenectomy related to the underlying disease. Laboratory exams showed: hemoglobin, 10.5 g/dL (reference range: 12.2 to 14.8 g/dL); leukocytes, 56,950/μL (reference range: 4,000 to 9,000/μL) with a predominance of bands and segmented neutrophils; and elevated transaminases (AST 61 U/L, ALT 71 U/L - reference range: AST up to 28 U/L, ALT up to 33 U/L), amylase (183 U/L – reference range: up to 100 U/L), lipase (571 U/L – reference range: up to 60 U/L), and lactate (41 mg/dL – reference range: 4.5 to 14.4 mg/dL). Arterial blood gas demonstrated metabolic acidosis. Clinically, there were no criteria for septic shock or pancreatitis.

Imaging Findings and Diagnosis

The imaging exams were performed all on the same day. Initially, an abdominal and chest radiography were performed for acute abdomen investigation and showed no signs that would lead to a diagnosis of this condition (Figs 1, 2, and 3). The abdominal ultrasonography identified a moderate amount of free fluid in the abdominal cavity, especially in the pelvis (Fig. 4).

The contrast-enhanced computed tomography (CT) performed showed free fluid in the cavity and pneumoperitoneum, mainly in the upper abdomen. There was also a focal duodenal thickening with mucosal discontinuity in its first portion, with low-attenuation material going through the submucosa towards the serous layer, findings that led to a high suspicion of ulcer perforation (Figs. 5 and 6).
Fig. 3 – Chest radiography in orthostasis.

Exploratory laparotomy and an ulcerorrhaphy were performed, and no Helicobacter pylori infection was found. In the immediate postoperative period, the patient evolved with ischemic stroke, recovering the level of consciousness and reversing the motor deficit. The patient is now in follow-up, with no sequela.

Discussion

Abdominal pain in children with sickle cell disease, in most cases, is attributed to subacute causes. The most frequent cause of abdominal pain in childhood, in emergency services or primary care, is constipation [9-11]. The list of differential diagnoses is wide and includes splenic and hepatic sequestration, cholelithiasis, renal infarcts, vaso-occlusive crisis or bezoars. All these conditions are facilitated by the pathophysiology of the underlying disease [8]. However, there are few reported cases of a perforated duodenal ulcer as a complication of sickle cell anemia in the pediatric population, and there is an absence of prevalence studies that correlate both entities.

The mucosal ischemia and hypoxia resulting from the vaso-occlusive crises (relatively common in patients with sickle cell disease) must be considered as factors that can contribute to the occurrence of perforated duodenal ulcers, as well as the use of non-steroidal anti-inflammatory drugs (NSAIDs) to control pain crises (medication that is known to be an important risk factor for the genesis of peptic ulcers), and the use of iron chelators (a drug that has a peptic ulcer as an infrequent adverse event) for patients who have received multiple blood transfusions [8].

Although upper gastrointestinal endoscopy is the method of choice for the diagnosis of peptic ulcers, even in young children [2], a study by Ilgar et al. [12] demonstrated that CT was able to correctly identify the perforation site in 85.7% of patients with gastroduodenal perforation. The most common CT

Fig. 4 – Abdominal ultrasonography. Free fluid in the pelvic cavity (arrow).

Figs. 5 – (A, B). High amount of free fluid in the abdominal cavity (arrows) with focus of free air (arrowhead) more evident in the upper abdomen.
findings in gastrointestinal tract perforation were free fluid in the abdominal cavity (89.4%), free air (76.6%), segmental wall thickening (48.9%), and wall discontinuity (25.5%). The use of oral contrast could help in the tomographic identification of the perforation site, as it showed extravasation of the contrast agent into the abdominal cavity (23.3%) [12].

**Conclusion**

Although perforated duodenal ulcers are a rare complication of sickle cell disease, this entity should be considered in patients with the disease who report abdominal pain and have significant symptoms such as shock or sepsis. Abdominal CT should be considered as a method for their diagnosis in the emergency department, but this requires an active search for duodenal alterations, and knowledge of this entity and its characteristic imaging findings described here.

**Patient consent**

We would like to inform that the written consent term has been given by both patient and parents.

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