J Ped Surg Case Reports 13 (2016) 10–12

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

An uncommon case of late-onset congenital diaphragmatic hernia with bloody stool

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ARTICLE INFO

Article history:
Received 20 June 2016
Received in revised form
24 July 2016
Accepted 24 July 2016

Key words:
Acute abdomen
Bloody stool
Late-onset congenital diaphragmatic hernia

ABSTRACT

Late-onset congenital diaphragmatic hernia (CDH) is an uncommon subset of CDH and distinct from neonatal CDH with respect to presenting symptoms, diagnosis, management, and prognosis. In particular, CDH diagnosed after 30 days of age (late-onset CDH) is uncommon and has an atypical presentation and a more favorable prognosis. In the present report, an infantile late-onset CDH case that presented with bloody stool and had a severe clinical course is described. In previous reports, no late-onset CDH case developed bloody stool. After diagnosis with image inspections, emergency surgery was performed. At operation, via the hernia orifice, the jejunum was seen to have prolapsed into the thoracic cavity with focally significant intestinal and mesenteric congestion, but no intestinal necrosis. In general, other disorders such as intussusception may be considered in the differential diagnosis of acute abdomen with bloody stool in patients of this age. However, in such late-onset CDH cases, immediate differentiation from other causes of acute abdomen that present with bloody stool is life-saving.

Congenital diaphragmatic hernia (CDH) is a congenital anomaly of the diaphragm in which abdominal organs herniate through the defect into the thoracic cavity; its incidence is about one per 2500 births [1]. CDH diagnosed immediately after birth is associated with significant respiratory distress and mortality. In contrast, CDH diagnosed after 30 days of age, which we call late-onset CDH, has been considered to be rare and generally follows a variable course, with a favorable outcome [2]. However, incorrect or delayed diagnosis of late-onset CDH may lead to serious morbidity and mortality [3].

In the present report, an infantile late-onset CDH case, which presented unconventionally and had a severe clinical course, is reported. In addition, we obtained permission from his parents to write up this case.

1. Case report

A 5-month-old Japanese boy admitted to our institution with choking and bloody stool. He was born to a healthy Japanese primigravida via cesarean section following induction of labor at 37 weeks of gestation due to prolonged labor. His birth weight was 3480 g. A fetal ultrasound at 30 weeks' gestation was reported as normal. No significant abnormalities were identified in his family history but his weight gradually decreased from 9100 g over 31 days to 8600 g (+1.0 standard deviation) at admission. At 5-month-old, he suddenly presented with choking. A few hours later, massive dark red bloody stool was observed 3–4 times in a 2 h period. He was immediately transferred to our emergency room and admitted to our institution. At admission, he presented with pallor, tachypnea (58 breaths per minute), tachycardia (194 beats per minute), desaturation (SpO_2: 80% on room air), and no response to pain. Because the blood pressure was impossible to measure, shock was diagnosed, and a bone marrow needle was immediately inserted at his right distal tibia at the root to ensure rapid fluid replacement. The patient then gradually started to react to a pain stimulus with improvement of vital signs. A complete blood count showed: white blood cell count 45,900 per μL with 34% neutrophils; hemoglobin 11.4 g/dL; and platelets 501,000 per μL. Serum biochemistry results showed: C-reactive protein (CRP) 0.7 mg/dL; blood urea nitrogen (BUN) 22.9 mg/dL; creatinine 0.85 mg/dL; and potassium 7.0 mEq/L. Blood gas analysis showed a metabolic acidosis (pH 7.173, base excess −17.1 mmol/L, $PaCO_2$ 27.1 mm Hg, and HCO_3^- 9.7 mmol/L).

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http://dx.doi.org/10.1016/j.epsc.2016.07.011

Initially, an abdominal ultrasound evaluation was performed because intussusception had been suspected from his clinical history and symptoms. However, the ultrasound showed normal findings of the ileocecal valve, which was not consistent with ileocolic intussusception, significant intestinal dilation, and poor peristalsis. In addition, the superior mesenteric artery and vein were pulled toward the left upper quadrant, and an edematous wall-thickened jejunum surrounded with pleural effusion was detected in the left thoracic cavity. Jejunal blood flow was impossible to confirm on power Doppler evaluation (Fig. 1). On chest X-ray, the mediastinum was deviated to the right side, and intestinal gas could be detected in the left thoracic cavity (Fig. 2). Thus, this patient was diagnosed as having late-onset CDH based on the ultrasound and X-ray findings, and emergency surgery was performed. Intraoperative findings showed that the hernia orifice opened slightly to the left from the diaphragmatic center and was covered with the left lobe of the liver. The liver and spleen were observed in the subdiaphragmatic abdominal cavity. Although the jejunum had prolapsed into the thoracic cavity, with focally significant intestinal and mesenteric congestion, no intestinal necrosis was observed (Fig. 3). Therefore, bowel resection was not performed, but the prolapsed jejunum was replaced into the abdominal cavity with direct closure of the hernia orifice. Meckel's diverticulum was not detected. The patient was extubated and started on enteral nutrition on day 7 after surgery, and he was discharged on day 14 after surgery. On outpatient follow-up, his psychomotor development and head magnetic resonance imaging were within normal limits, and no recurrence of bloody stool has been seen for about 2 years.

2. Discussion

Late-onset CDH is an uncommon subset of CDH and distinct from neonatal CDH with respect to presenting symptoms, diagnosis, management, and prognosis. CDH diagnosed after 30 days of age is particularly uncommon and has an atypical presentation with a more favorable prognosis. Late-onset CDH patients may present with various respiratory or gastrointestinal symptoms or may be absolutely asymptomatic [4,5]. In accordance with a review summarized by a CDH study group, presenting symptoms could be identified in 46 cases of late-onset CDH. They were classified as respiratory symptoms and diseases (respiratory tract infections, respiratory distress, cough, wheezing, etc) in 20 cases, gastrointestinal symptoms and diseases (vomiting, abdominal pain,

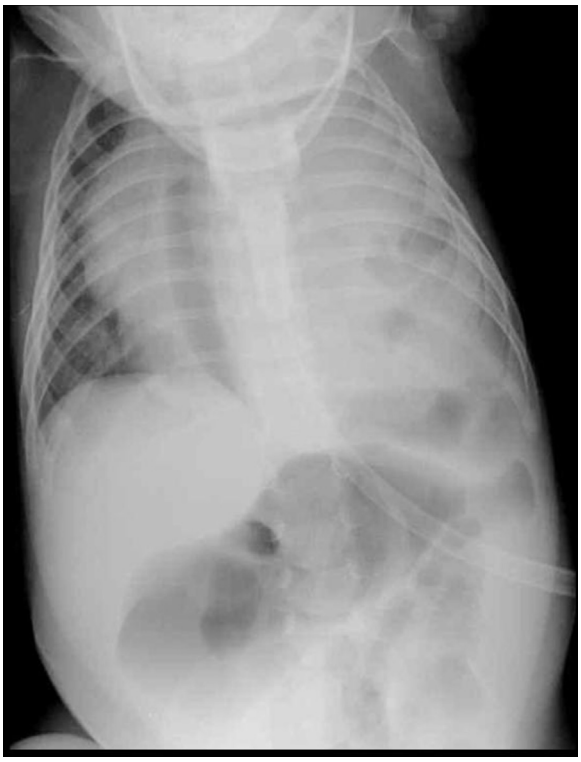

Fig. 2. The radiographic appearances immediately after arrival to the emergency department. The mediastinum is deviated to the right side, and intestinal gas is detected in the left thoracic cavity.

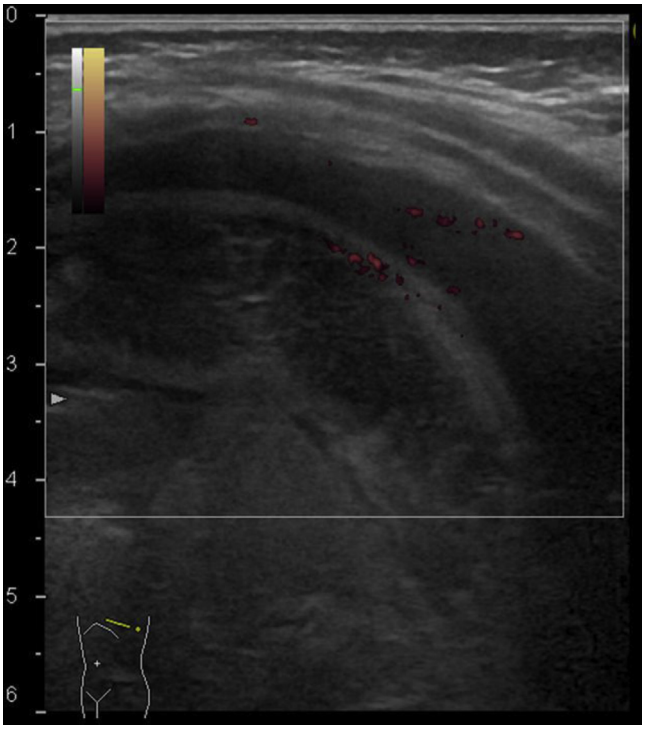

Fig. 1. The ultrasonographic appearances immediately after arrival to the emergency department. Ultrasonographic examination shows an edematous wall-thickened jejunum surrounded with pleural effusion. Jejunal blood flow was impossible to confirm on power Doppler evaluation.

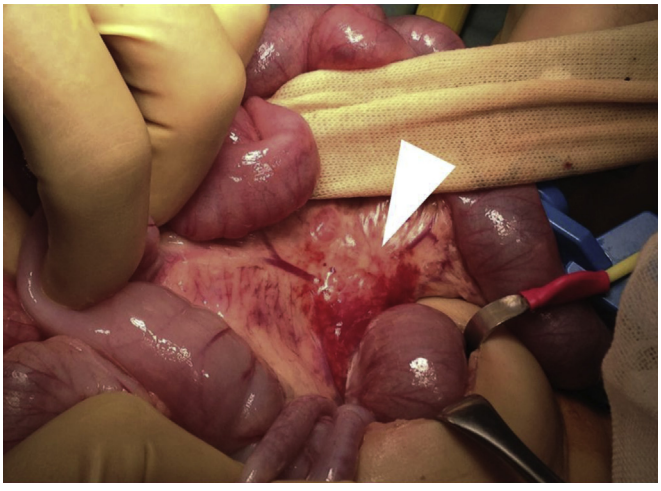

Fig. 3. The appearance at emergency surgery. The strangulated organ is the jejunum, showing no intestinal necrosis. Instead, significant intestinal and mesenteric congestion could be found (arrow head).

failure to thrive, constipation, etc) in 15, both in six, and asymptomatic in five [4]. Although the present case developed obvious bloody stool and respiratory and cardiovascular distress, no late-onset CDH case with bloody stool has been previously reported.

In general, ileo-colic intussusception may be the prime suspect in the differential diagnosis of acute abdomen with bloody stool in patients at this age. Bloody stool is also a characteristic of intussusception, but it is detectable in only 16% of cases with duration of other symptoms less than 12 h. In the diagnosis of intussusception, abdominal ultrasound has almost 100% sensitivity and specificity, and it is sometimes useful for detecting other abdominal disorders in children with suspected intussusception in whom no intussusception is detected. Thus, ultrasonography has priority for diagnosis of intussusception, but if the diagnosis is unclear, or when there is no medical staff with ultrasonic expertise, a contrast enema study is necessary [6]. In the present case, the symptoms in this patient were choking and bloody stool making late-onset CDH as one of the diagnosis that should be considered. A chest X-ray should be part of the initial diagnostic work-up and the diagnosis would have been made. However, at the time of transfer, we performed resuscitation with priority because vital signs of the patient were severe. Therefore, it was forced to perform in synchronization with resuscitation and ultrasonography by pediatricians.

As to the pathophysiology of this late-onset CDH case, the following may have been involved. Initially, the jejunum invaginated into the left thoracic cavity when intraabdominal pressure increased for some reason. Then, the invaginated jejunum, which compressed the left lung and mediastinum, caused circulatory and respiratory compromise. In addition, impairment of intestinal blood flow due to jejunal strangulation provoked ileus and ischemic enteritis. Finally, a cardiopulmonary disorder with dehydration derived from the intestinal ileus provoked shock, and the ischemic enteritis induced bloody stool. The pathophysiology of the present case is not entirely clear. Although significant intestinal congestion was confirmed at operation, hemorrhagic spots at the mucosal surface could not be directly confirmed because intestinal resection and pathological evaluation were performed. Small intestinal endoscopy was also impossible to perform because of the patient's poor general condition and small physique. However, the congested portion of the jejunum might have been the main lesion, because no episode of bloody stool has been observed in the postoperative two-year follow-up, and no obscure bleeding sources such as colonic polyps were detected on several abdominal ultrasound examinations.

3. Conclusion

A rare case of late-onset CDH that presented with bloody stool was described. Although late-onset CDH may involve gastrointestinal symptoms, no previous reports of late-onset CDH with bloody stool were identified. In late-onset CDH cases that develop poor vital signs and rare symptoms, as in the present case, immediate differentiation with other causes of acute abdomen that present with bloody stool is essential for life-saving management.

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