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Case Report

Late-presenting congenital diaphragmatic hernia in a child with gastric perforation and acute pancreatitis

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ABSTRACT

Background. Late-presenting congenital diaphragmatic hernia occurs beyond the neonatal period, and is relatively rare, presenting with nonspecific respiratory and gastrointestinal symptoms.

Case. We report a rare case of late-presenting congenital diaphragmatic hernia in a 7-year-old girl, who presented with abdominal pain, shortness of breath and fever on admission. Work-up revealed intrathoracic gastric perforation, acute pancreatitis and septic shock with a diaphragmatic defect. Due to the high content of amylase in pleural effusion, we suspected the presence of a pancreaticopleural fistula, and we were also puzzled whether the gastric perforation was caused by a pleural indwelling catheterization, but this was ruled out. We about performed a laparotomy to reposition the herniated organs, repair the hernia and the gastric perforation, and undergo the gastrostomy. The girl had an uneventful post-operative recovery.

Conclusions. Late-presenting congenital diaphragmatic hernias are often misdiagnosed. Clinicians should combine multiple imaging modalities to make a definite diagnosis and perform surgery as soon as possible to avoid severe complications.

Key words: late-presenting congenital diaphragmatic hernia, hydropneumothorax, gastric perforation, acute pancreatitis, children.

Congenital diaphragmatic hernia (CDH) is characterized by an embryonic defect of the diaphragm, resulting in the abdominal organs herniating into the thoracic cavity, with a reported incidence of 1 in 2500-5000 live births.[1,2] The majority of CDHs suffer from respiratory distress in the neonatal period due to pulmonary dysplasia and pulmonary hypertension. About 5-20% of the cases occur beyond the neonatal period, which is called late-presenting CDH. They present with nonspecific respiratory and gastrointestinal symptoms and are always misdiagnosed, sometimes even fatal.[3] Here we report a pediatric case of late-presenting CDH with gastric perforation, acute pancreatitis and septic shock.

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Case Report

A 7-year-old girl presented to our pediatric emergency department with progressive abdominal pain accompanied by shortness of breath for 1 day and fever for 10 hours. There was no history of trauma or surgery. Her vital signs showed tachycardia (196 beats/minute) with a fever of 39.5°C, hypotension (56/32 mmHg), and tachypnea (60 breaths/minute) with a transcutaneous oxygen saturation of 83%. On physical examination, she was lethargic, cyanotic and her capillary refill time was delayed. She had slight epigastric tenderness, muffled cardiac sounds and her breath sounds on the left hemithorax were diminished on auscultation. Blood gas analysis showed a PaO_2/FiO_2 ratio less than 300 mmHg and metabolic acidosis with lactate of 7.21 mmol/L. Emergency fluid resuscitation and endotracheal intubation were performed, and she was admitted to the pediatric intensive care unit.

Initial chest X-ray in supine position revealed the left lung field hyperlucent with the mediastinum shifting to right, which suggested pneumothorax (Fig. 1). Instant thoracentesis released a large amount of gas and 400 ml of brown turbid pleural effusion, then a thoracic drainage tube was placed. The girl's vital signs improved apparently. The laboratory test showed the leukocyte count was within the normal limit, but c-reactive protein (130.73 mg/L) and procalcitonin (41.22 ng/ml) were significantly elevated. In addition, serum amylase was 498 U/L, serum lipase was 600 U/L, and amylase in pleural effusion was 28108.1 U/L. They were all above normal. We suspected the presence of a pancreaticopleural fistula, but the abdomen ultrasound was consistent with acute pancreatitis and didn't detect a fistula. We gave her conservative treatment, including antibiotics, and somatostatin analogues. One day later, the level of serum amylase (382 U/L) and serum lipase (69 U/L) decreased, but the patient's condition did not improve significantly. The subsequent X-ray showed an air-filled oval-shaped outline of the left lower hemithorax in continuity with the abdomen, and the rightward mediastinal shift remained. The bedside thorax ultrasound detected some solid tissue echo in the left thoracic cavity, but the sonographer on duty didn't identify it due to a lack of experience. So we performed an upper gastrointestinal series at bedside with iodic contrast medium injected through a nasogastric (NG) tube, demonstrating the intrathoracic stomach filled with contrast, and contrast leaking into the thoracic cavity (Fig. 2). At that time, the diagnosis of a left late-presenting CDH with gastric perforation, acute pancreatitis and septic shock was basically clear.

An exploratory laparotomy was performed urgently, and a defect of about 5cm×3cm in the left posterolateral diaphragm (Bochdalek hernia) was found. Surgeons enlarged the tight hernia ring and repositioned the herniated abdominal organs, including the stomach, spleen, a small portion of the omentum, transverse colon and proximal small intestine. Exploring the left thoracic cavity revealed mild pulmonary dysplasia, then they closed the

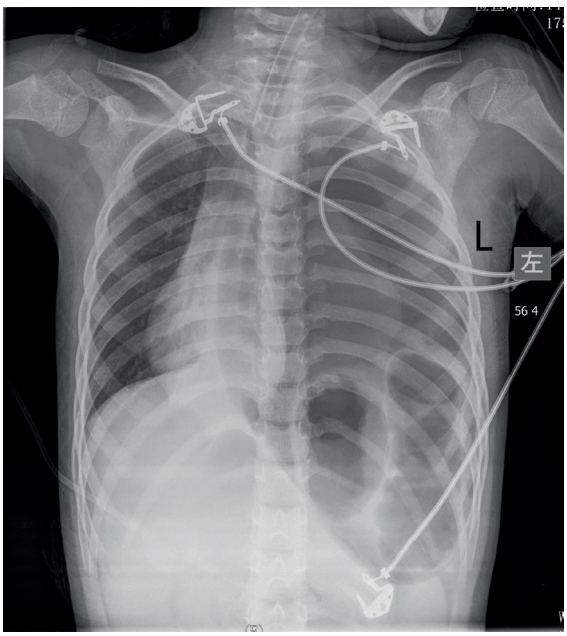

Fig. 1. Initial chest X-ray in supine position showing the left lung field is hyperlucent with a rightward mediastinal shift.

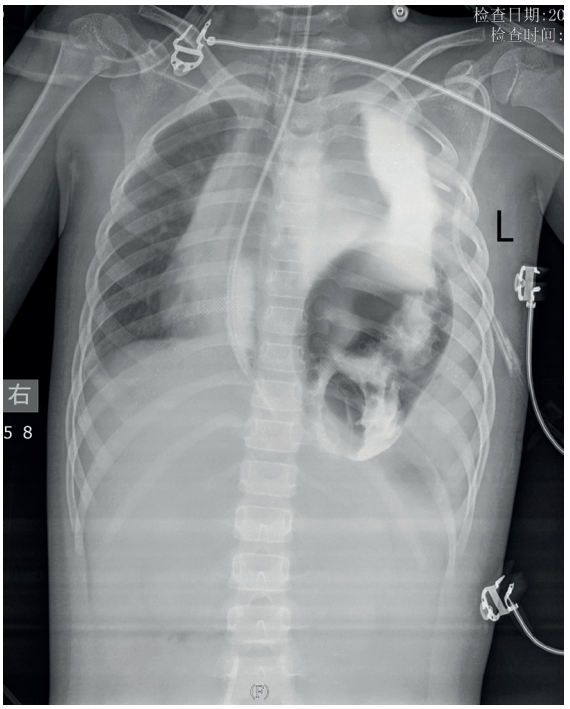

Fig. 2. The upper gastrointestinal series demonstrating the intrathoracic stomach filled with contrast, and contrast leaking into the thoracic cavity.

diaphragmatic defect with 2-0 non-absorbable interrupted sutures and found high tension at the suture site, so they reinforced it with a bovine pericardial patch. The herniated organs appeared congested, edematous, and closely adhered to each other. They released the adhesions and detected two gastric perforations with surrounding necrosis on the lesser curvature (3cm×2cm) and fundus (4cm×3cm) respectively, and multiple focal ischemia could be seen on the gastric wall (Fig. 3 and Fig. 4). There was no evidence that the thoracic drainage tube had damaged the stomach, surgeons repaired the gastric perforation and performed a gastrostomy and implanting a mushroom head tube into the stomach through the skin, and found no evidence of a pancreaticopleural fistula intraoperatively. The girl had an uneventful post-operative recovery, she was discharged on the 10th post-operative day and followed up for 6 months asymptomatically. Informed consent was obtained from the girl's family for this case report.

Discussion

Late-presenting CDH is less common than neonatal CDH despite the fact that the diaphragmatic defects are both congenital in anatomy, but the clinical characteristics are quite different from each other.[3] Late-presenting CDH has no obvious symptoms in the early stage that may be attributed to the small amount of herniated organs in the thoracic cavity or without any herniation.[4] Intrathoracic pressure increases while the abdominal organs suddenly herniate into the chest cavity, accompanied by pulmonary compression, contralateral mediastinal displacement and incarcerated intestinal obstruction, resulting in diverse clinical manifestations.[3]

According to the radiological findings on admission, pneumothorax was the most important emergency diagnosis for this patient. Considering that trauma can also lead to pneumothorax, we repeatedly confirmed the trauma history of this case and ruled it out. We also suspected the presence of a pancreaticopleural fistula due to the high amylase content in the pleural effusion. It has been documented that a pleural effusion amylase level higher than 50,000 IU/L could be directly diagnosed as a pancreaticopleural fistula.[5] But our case had only an intermediately increased amylase level of 28108.1U/L in the pleural effusion, and we eventually disproved that suspicion because there was no further indication of the fistula. It is also inappropriate to analyze the pneumothorax and pleural effusion separately in our case. High levels of amylase in pleural effusion can be found in other pathologies, including acute pancreatitis.[5]

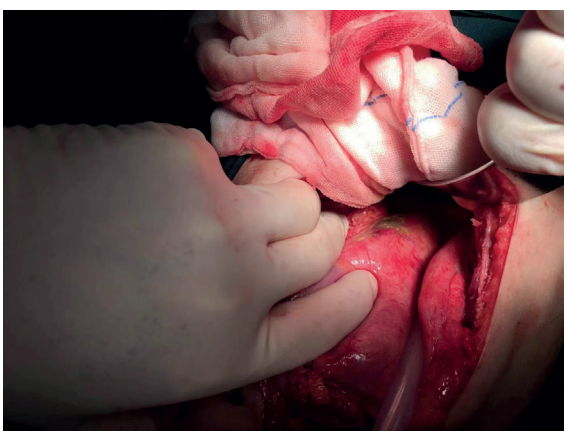

Fig. 3. Intraoperative photograph showing a gastric perforation of about 3cm×2cm on the lesser curvature of the stomach with surrounding necrosis.

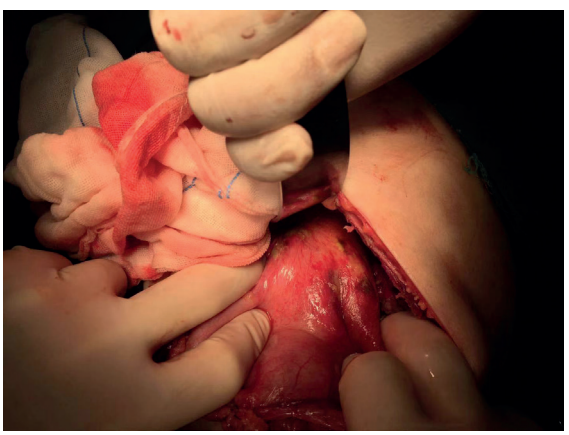

Fig. 4. Intraoperative photograph showing multiple focal ischemia on the gastric wall.

The leakage of salivary amylase into the thorax was probably due to the gastric perforation. Acute pancreatitis secondary to diaphragmatic hernia is relatively rare. Harrington et al.[6] reported an adult Bochdalek hernia case with acute pancreatitis, but the pancreas was not the hernia content, and the acute pancreatitis was considered to be due to traction. Our case was similar in that pancreatitis was triggered by mechanical traction of other hernia content.

Chest and abdomen X-rays are usually the initial imaging studies performed in late-presenting CDH, but they are not enough to make a definitive diagnosis. CDH may even be misdiagnosed as another disease due to the resemblance of radiological features like pneumothorax, pneumonia, pleural effusion, or congenital pulmonary airway malformation.[3] As a gradually popularized diagnostic method, the point-of-care ultrasound can be used independently for the diagnosis of pneumothorax or diaphragmatic hernia, which provides real-time dynamic images and is easily repeatable, but requires appropriate training and quality assurance.[7,8] Although, in this case, bedside ultrasound did not provide valid diagnostic information due to the inexperience of the young sonographer at the time, more attention should be paid to the role of ultrasound. The upper gastrointestinal series is also a helpful diagnostic modality, which played a crucial role in our case. The computed tomography (CT) can clearly detect the diaphragmatic defect and intrathoracic viscera, which is the more important examination.[3] This case was not transferred for CT because of her critical condition. So, apart from perfect medical history collection, appropriate imaging examination is the key to a correct diagnosis.

However, the misdiagnosis rate for late-presenting CDH is as high as 38.2%.[3] Interventions following misdiagnosis may lead to iatrogenic complications, such as gastric perforation resulting from thoracentesis and pleural indwelling catheterization.[9,10] Clinicians should be more cautious about these interventions while they seem reasonable. Fortunately, in our case, the surgical exploration verified that the two large gastric perforations had no relation to the thoracic drainage tube. We speculate that the gastric perforations may have existed before admission and resulted from the high tension of the intrathoracic stomach, which was the severest complication in our case.

Once the late-presenting CDH is clearly diagnosed, surgical treatment should be performed promptly, laparotomy is superior to dealing with cases that have visceral complications.[11] In this case, we performed a gastrostomy in addition to the repair to provide effective decompression and adequate drainage of the dilated stomach in order to prevent secondary perforation. In addition, considering there was a risk of recurrence of a diaphragmatic hernia, we used a bovine pericardial patch to reinforce the primary repair because of the high tension at the suture site. It has been documented that a biologic mesh has a better application prospect in terms of low reherniation, low calcification and infection rate.[12]

In conclusion, late-presenting CDH in children presenting as gastric perforation, acute pancreatitis and septic shock is rare. Clinical and imaging misdiagnosis is much more common. Clinicians should combine multiple imaging modalities to make a definite diagnosis and perform surgery as soon as possible to avoid severe complications.

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Ethical approval

Informed consent to publish the case report has been obtained. And this report does not contain any personal information that could lead to the identification of the patient.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: QL, CL; data collection: QL; analysis and interpretation of results: QL, CL ; draft manuscript preparation: QL. Both authors reviewed the results and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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