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CASE REPORT

Delayed presentation of strangulated congenital diaphragmatic hernia: learning from our experience

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Abstract
Background Strangulation is very rare in congenital diaphragmatic hernia (CDH) of the Bochdaleck variety. Here, we share our experience with six cases of delayed presentation of strangulated CDH. The aim of this article is to provide information on how to diagnose and manage this situation using a systematic approach.
Materials and methods A retrospective review identified six cases of strangulated/obstructed CDH from 1998 to 2011. Demographic data, clinico-radiological findings, management and complications, along with final outcome were recorded.
Results Small bowel gangrene was found in one patient, gastric perforation in three, transverse colon perforation in one and colonic obstruction in one patient. Video-assisted thoracoscopic surgery (VATS) was used in all but one patients for definitive diagnosis, diaphragmatic repair, pleural lavage and management of empyema. Laparotomy was needed for management of strangulated or perforated bowel. Three patients in this study survived.
Conclusion Clinicians should always consider a diagnosis of obstructed Bochdaleck hernia in children, because X-ray findings are not always typical or even normal in complicated CDH. VATS may be considered as both diagnostic and therapeutic. Preventive measures for empyema or early intervention in the evolving stage can significantly reduce morbidity.

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Introduction

Variability in the clinical presentation of congenital diaphragmatic hernia (CDH) depends on topography, dimensions of the diaphragmatic defect, and the age of the affected child [1, 2]. The incidence of late-presenting CDH in the pediatric population is 5–25% [1, 2]. Late-presenting CDH has been generally regarded as a benign condition with a very good prognosis [1, 2]. However, this does not always holds true, as rare complications like bowel strangulation, pneumothorax and fecothorax can have a lethal course if not treated promptly [3–5]. Because of the rarity of this condition, we reviewed our records retrospectively to develop a management strategy for this entity.

Materials and methods

This retrospective study was performed at the Department of Pediatric Surgery of the Medical University between 1998 and 2011. The review found six complicated cases of delayed presentation CDH. Diagnosis was made by content of intercostal drainage (ICD), chest X-ray, or laparotomy. We analyzed demographic data (age, sex), together with clinical presentation, management, per-operative/post-operative complications and their management, and final outcome. Chest X-ray was the initial diagnostic procedure supplemented with video-assisted thoracoscopy (VAT) or contrast enhanced CT (CECT) of the thorax, if needed. In the postoperative period, for confirmation of empyema

thoracis, CECT of the chest was performed. None of our patients had any history of past trauma.

Case 1

A 10-year-old boy presented with a left-sided ICD placed elsewhere for respiratory distress. The pre-ICD chest X-ray suggested hydro-pneumothorax, along with contralateral mediastinal shift. At the time of admission, the child had tachypnea, and ICD was draining feculent content. After resuscitation, VATS was performed, which revealed a left-sided 5 × 3 cm diaphragmatic defect, along with perforated stomach due to the ICD tube. After reduction of the contents, thoracoscopic repair of the diaphragmatic defect was performed. Thereafter, a left subcostal incision was made and a 2 × 2 cm gastric perforation was primarily repaired (Fig. 1). The patient was kept on ventilatory and vassopressor support. Unfortunately, the child succumbed to death in the immediate postoperative period because of malnutrition, ongoing sepsis, respiratory distress, and multi-organ failure.

Case 2

An 8-year-old boy presented with acute respiratory distress and features of large bowel obstruction. The auscultation revealed decreased air entry on the left side of the chest. An abdominal X-ray revealed an elevated left hemi-diaphragm with dilated bowel in the left hemithorax, and multiple air-fluid levels. After resuscitation, VATS was performed and the gangrenous colon and small bowel were reposited back into the abdomen, along with the repair of a 4 × 2 cm diaphragmatic defect. The gangrenous colon (Fig. 2) was resected and a colostomy was performed. At postoperative day 6, the patient had minimal, purulent ICD drainage with decreased ipsilareral lung expansion. CECT of the thorax revealed loculated empyema thoracis, for which pleural adhesiolysis and lavage were carried out. The patient was discharged on day 12. The colostomy closure was performed after 3 months. At 1 year of follow up, the child had no fresh complaints.

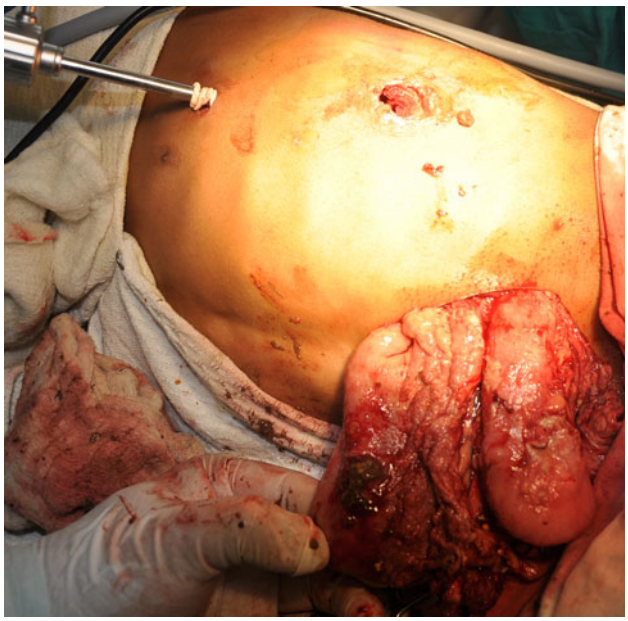

Fig. 1 Video-assisted thoracoscopic surgery (VATS) followed by laparotomy showing iatrogenic gastric perforation due to intercostal drainage (ICD)

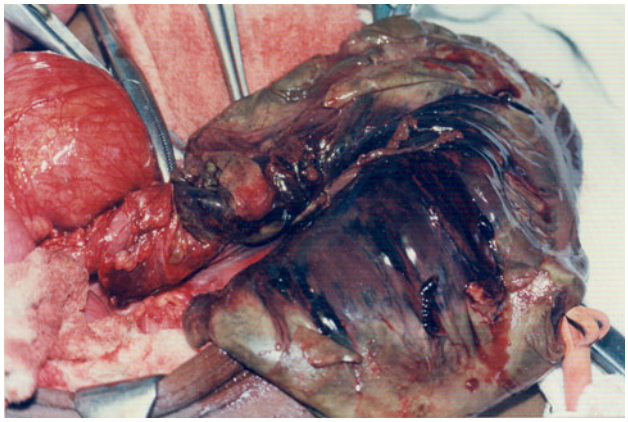

Fig. 2 Gangrenous colon due to strangulation in congenital diaphragmatic hernia (CDH)

Case 3

A 6-year-old boy presented with a right-sided ICD placed elsewhere, which was draining feculent content (Fig. 4). The pre-ICD X-ray chest suggested an opacified left lower lung field resembling pleural effusion. VATS revealed a 3 × 3 cm CDH. The liver, along with part of stomach, small bowel, and transverse colon, were herniated into the thoracic cavity. Through exploratory laparotomy, resection of the strangulated jejunum was performed. Postoperatively, the child was kept on a ventilator but succumbed to death on postoperative day 3 due to ongoing sepsis.

Case 4

An 8-year-old girl presented with acute respiratory distress, circulatory collapse, and a single episode of hemetemesis. Chest X-ray revealed a left-sided diaphragmatic hernia with a single large air fluid level and mediastinal shift to the right side. Nasogastic intubation was unsuccessful. A clinical diagnosis of strangulated CDH with gastric volvulus was made. The VATS revealed the questionable viability of the gastric fundus and body, and the body had a 1 × 1 cm gastric perforation. There was 5 × 4 cm

diaphragmatic defect. After reduction of the contents, the diaphragm was repaired. A gastrostomy was placed through the perforation site. Postoperatively, the child was kept in the pediatric intensive care unit but expired the following day due to circulatory collapse.

Case 5

A 5-year-old boy presented with acute respiratory distress and circulatory collapse. Chest X-ray revealed a left-sided pneumothorax with a shift of the mediastinum to the right side. An emergency tube thoracostomy was carried out, which improved respiration. On the 2nd day, bile mixed with serosanguinous content was draining through the ICD. VATS revealed a left-sided diaphragmatic defect of 4 × 3 cm, with the strangulated stomach coming through it. The defect was repaired and pleural cavity lavage was performed. Diversion gastrostomy and feeding jejunostomy were also performed. Despite all these conservative measures, the patient developed partial collapse of the left lung. Twenty days later, the child underwent VATS adhesiolysis. Thereafter, the child improved and was discharged with proper follow up advice. The ICD was removed after 15 days. The gastrostomy and jejunostomy tubes were removed after 2 months. At present, the child is healthy.

Case 6

A 1.5-year-old female child presented with acute large bowel obstruction. X-ray of the chest was absolutely normal, while that of abdomen had features of intestinal obstruction. The laparotomy revealed a 2 × 1 cm diaphragmatic defect, through which spleen, omentum, and splenic flexure of the colon were herniated (Fig. 3). The

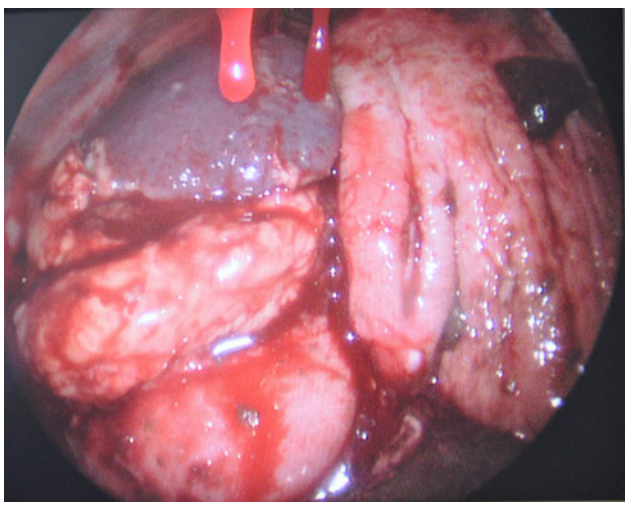

Fig. 3 Thoracoscopic view showing herniated spleen, stomach, omentum and colon into the thorax

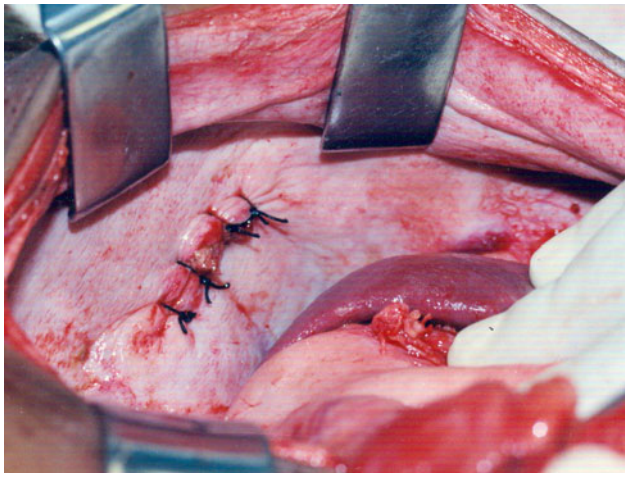

Fig. 4 Repaired diaphragmatic defect

contents were reduced and the defect was repaired (Fig. 4). The patient was discharged on the 4th day.

Results

Over a period of 13 years, six patients with CDH presented to the department, of which four were male and two female. All had left-sided CDH. Chest X-ray showed features of CDH in only two cases, pneumothorax in two, moderate pleural effusion in one, while in one patient the X-ray was absolutely normal. VATS was used for definitive diagnosis in all patients. In all patients, the diaphragm was repaired by simple interrupted suturing, using non-absorbable sutures (polypropelene/silk suture). Mesh was not required in any of them, as we were able to repair the defect primarily. Laparotomy was needed for repair of strangulated or perforated bowel. In the postoperative period, three patients developed empyema thoracis. Pleural adhesiolysis and lavage was performed via VATS in these patients. Of the six patients, only three survived. Of these, two expired in the immediate postoperative period, while the third died later due to ongoing sepsis, malnutrition and lung collapse.

Discussion

Contrary to neonatal posterolateral diaphragmatic hernia, which manifests uniformly with respiratory distress, patients with CDH diagnosed beyond infancy may be completely asymptomatic or may present with chronic, acute, or even sudden symptoms [1, 2, 4, 6]. As far as pathogenesis is concerned, late-presenting CDH seems to be an isolated congenital defect of the diaphragm only, whereas neonatal CDH is a complex anomaly, which is associated with many abnormal physiological components [4, 6].

As compared to right-sided CDH, the frequency of left-sided CDH is higher, due to the protecting effect of the liver on the right hemi-diaphragm [4]. Obstruction of the bowel may occur in CDH, but fecothorax and pneumothorax are uncommon complications of delayed CDH [3–5]. Carter and Giuseffi [5] analyzed all reported cases of strangulated diaphragmatic hernia and concluded that, in 90% of cases, strangulation occurs most commonly in traumatic diaphragmatic hernia, while it is rare in bochdaleck hernia, possibly due to the size of the diaphragmatic defect.

Radiology in strangulated CDH is not very suggestive of the diagnosis, especially if strangulation leads to perforation of the bowel, as the pneumothorax itself will not then reveal the diagnosis [7–10]. Contrast studies, described classically, cannot be performed if strangulation of the bowel is suspected [7]. Only CECT thorax of the abdomen can inform the diagnosis but this requires a high suspicion of diagnosis [7, 10]. In this series, patients 1 and 3 presented with ICD draining feculent fluid; therefore, CT was not needed in these patients. In patients 2 and 4, we were able to diagnose CDH on chest X-ray. Inadvertent ICD placement revealed CDH in patient 5. Patient 6 presented with abdominal symptoms, for which laparotomy was performed, and CECT thorax was not used to make the diagnosis. However, we do agree that CECT is the best modality to diagnose this condition. On the other hand, there may be CDH with strangulation but chest X-ray may be absolutely normal (case 6). Hence, the treating surgeon should always keep in mind the possibility of this entity in the differential diagnosis of acute abdomen or respiratory distress in children.

After analyzing the clinical data from these patients, it seems that intrathoracic displacement of the stomach is associated with the highest risk of a complicated course of late-presenting CDH [3, 4].

Which approach is best remains controversial. The proponents of the abdominal approach consider it to be superior to thoracotomy because of the better opportunity to assess and repair all intraabdominal viscera [4–9], while opponents believe thoracoscopy to be superior, as strangulated or incarcerated hernia has adhesions with thoracic organs [5, 6, 11, 12, 13, 15]. Some prefer thoracoabdominal incision for this complex entity [12]. We used a method intermediate between these latter approaches, i.e., VATS for adhesiolysis of viscera from pleural cavity, repair of diaphragm, and thoracic cavity lavage; and laparotomy for repair of perforation and diversion/enteral feeding purposes. VATS has the added advantage of avoidance of thoracic incision, which may not heal properly in the postoperative period due to the catabolic phase (as occurred in our first case). Furthermore, thoracic incision may restrict respiratory efforts in the postoperative period, in which the child requires vigorous prophylactic chest physiotherapy, as empyema is almost inevitable due to gross contamination of the pleural cavity [14].

Although it seems unorthodox, in an emergency situation, life saving emergency gastric decompression can sometimes be done through the chest wall to relive acute respiratory distress [5]. Once the child has been resuscitated, the definitive procedure can be performed. For diversion of food from the stomach and to enhance healing, a gastrostomy should be performed in large gastric perforations. To administer early enteral feeding, a feeding jejunostomy may be performed safely as we did in our fifth patient, who survived. Early feeding allows fast wound healing, early mobilization, decreased hospital stay, and improve immunity to resist chest infection [14]. Further, if empyema develops, a good nutritional state helps the child to withstand the required surgery for this condition.

Any extent of strangulation of abdominal viscera, especially if perforation has occurred, increases the risk of empyema. Thus, attention to preventive measures for empyema or early intervention in its evolving stage can significantly reduce morbidity and mortality [13, 15]. VATS is also a good modality for managing empyema, especially in the early stage before irreversible changes develop in pleura, and can decrease the morbidity and long-term complications due to thoracotomy [13, 15].

To conclude, clinicians should always consider a diagnosis of obstructed Bochdaleck hernia in children, because X-ray findings are not always suggestive, especially if strangulation of the bowel occurs. In these situations, radiology may show a diverse picture of the hemothorax and pneumothorax with or without mediastinal shift. VATS may be considered as both diagnostic and therapeutic. It should be used before laparotomy to release adhesions and reduce viscera with pleural lavage. Preventive measures for empyema or early intervention in the evolving stage can significantly reduce morbidity.

References

1. Weber T, Tarcy T, Bailey P, Lewis JE, Westfall S (1991) Congenital diaphragmatic hernia beyond infancy. Am J Surg 162:643–646
2. Berman L, Stringer D, Ein S, Shandling B (1988) The late-presenting pediatric Bochdaleck hernia: a 20-year review. J Pediatr Surg 23:735–738
3. Radin DR, Ray MJ, Halls JM (1986) Strangulated diaphragmatic hernia with pneumothorax due to colopleural fistula. AJR Am J Roentgenol 146:321–322
4. Bagłaj M, Dorobisz U (2005) Late-presenting congenital diaphragmatic hernia in children: a literature review. Pediatr Radiol 35:478–488
5. Hoffman E (1968) Strangulated diaphragmatic hernia. Thorax 23:541–549

6. Buxi TBS, Pruthi PK, Dutta T (1991) The diagnosis of strangulated diaphragmatic hernia. Pediatr Radiol 21:527–528
7. Minagi H, Brody WR, Laing FC (1977) The variable roentgen appearance of traumatic diaphragmatic hernia. J Can Assoc Radiol 28:124–128
8. Leahy P, Liu K, Guiney E (1989) Pneumothorax—an unusual delayed presentation of congenital posterolateral diaphragmatic hernia. Pediatr Surg Int 4:415–416
9. Cruz CJ, Minagi H (1994) Large-bowel obstruction resulting from traumatic diaphragmatic hernia: imaging findings in four cases. AJR Am J Roentgenol 162:843–845
10. Rahman AA, Panda M (2009) Locked lung by looped hernia: a case report. Cases J 2:29
11. Manning PB, Murphy JP, Raynor SC, Ashcraft KW (1992) Congenital diaphragmatic hernia presenting due to gastrointestinal complications. J Pediatr Surg 27:1225–1228
12. Borrie J (1960) Strangulated diaphragmatic hernia: five cases treated surgically. Aust NZ J Surg 29:311–320
13. Wani ML, Ahangar AG, Lone GN, Singh S, Dar AM, Bhat MA, Lone RA, Irshad I (2010) Feeding jejunostomy: does the benefit overweight the risk (a retrospective study from a single centre). Int J Surg 8:387–390
14. Lardinois D, Gock M, Pezzetta E, Buchli C, Rousson V, Furrer M, Ris HB (2005) Delayed referral and gram-negative organisms increase the conversion thoracotomy rate in patients undergoing video-assisted thoracoscopic surgery for empyema. Ann Thorac Surg 79:1851–1856
15. Cassina PC, Hauser M, Hillejan L, Greschuchna D, Stamatis G (1999) Video-assisted thoracoscopy in the treatment of pleural empyema: staged-based management and outcome. J Thorac Cardiovasc Surg 117:234–238