A Death Due to Complications Following Late-Onset Congenital Diaphragmatic Hernia in a Young Child

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Abstract
Congenital diaphragmatic hernia (CDH) is a rare anatomical birth defect, causing the diaphragm fails to fuse completely, leaving a defect and causing herniation of the abdominal viscera towards the thoracic cavity. Though most cases of CDH are symptomatic and diagnosed in the early stages of life some asymptomatic cases may go undetected and later detect as incidental or may present with respiratory or gastrointestinal symptoms. We report a case of late-onset CDH which leads to a rapidly fatal medical emergency causing challenges in clinical diagnosis. An eight-year-old previously healthy schoolgirl was admitted to a primary care hospital following recurrent upper abdominal pain, nausea, and vomiting for a duration of 3 consecutive days. According to the parents she has taken treatment from a general practitioner. She was afebrile, hypotensive, tachycardic, and drowsy and was found unresponsive during transfer to a tertiary care hospital and was pronounced dead. On opening the thoracic cavity, it was noted that some of the abdominal contents like the entire stomach, duodenum, and proximal part of the small intestine, splenic flexure of the transverse colon were present within the left hemi thorax causing left lung hypoplasia, thoracic midline shift, intestinal compression, and elevated luminal pressure causing gastric erosions and chemical peritonitis. This case highlights the importance of higher clinical vigilance and the value of early diagnosis and timely medical attention to relatively uncommon but surgically correctable conditions like CDH.

Keywords: Congenital diaphragmatic hernia, Bochdalek, Morgagni, lung hypoplasia, chemical peritonitis

Received: 03 June 2022, **Revised version accepted:** 27 June 2022, **Published**: 30 June 2022. ***Corresponding author:** Warushahennadi J, Email: janaki@med.ruh.ac.lk https://orcid.org/0000-0002-6880-5513

Cite this article as: UGB Jayarathna, J Warushahennadi, LKCD Ariyarathna. A Death Due to Complications Following Late-Onset Congenital Diaphragmatic Hernia in a Young Child. Medico-Legal Journal of Sri Lanka, 2022;10(1):50-53. DOI: http://doi.org/10.4038/mljsl.v10i1.7459

Introduction

Congenital diaphragmatic hernia (CDH) is a rare anatomical birth defect, causing the embryological elements of the diaphragm fail to fuse completely, leaving a defect.[1] The incidence of CDH is 1 in 2500 births, with the left side (85%) predominance.[2,3]. Most cases of CDH are symptomatic and diagnosed in the early stages of life, prenatally or during the immediate postnatal period. Asymptomatic cases may go undetected and later detect as incidental or may present with respiratory or gastrointestinal symptoms as a result of sudden or exacerbated herniation of abdominal contents into the thoracic cavity. It also has a common association with pulmonary hypoplasia of the affected side.[4,5] These late-onset cases lead to rapidly fatal medical emergencies causing challenges in clinical diagnosis and prompt medical care.

Case report

An eight-year-old previously healthy schoolgirl was admitted to a primary care hospital following recurrent upper abdominal pain, nausea, and vomiting for a duration of 3 consecutive days. According to the parents she has taken treatment from a general practitioner. She was afebrile, hypotensive, tachycardic, and drowsy and was found unresponsive during transfer to a tertiary care hospital and was pronounced dead. Her past medical, surgical and immunization histories were unremarkable. A medico-legal autopsy was performed following an inquest.

On opening the thoracic cavity, it was noted that abdominal contents (entire stomach, duodenum and proximal part of the small intestine, splenic flexure of the transverse colon and part of the omentum) were present within the left hemi thorax (Fig.01). A 7cm x 4cm size large defect was observed on the left hemi diaphragm along the left posterolateral chest

wall resembling a Bochdalek hernia (Fig.02). The mediastinum had been pushed to the opposite or right side and the left lung was hypoplastic and atelectatic with a weight of 70g (Fig.03). The right lung was 240g in weight and normal in size. All the other organs were free of visible pathologies. The stomach was filled with brown coloured partially digested food material. There were no congenital malrotation of the gut, volvulus, or intussusception and no necrosis of the affected segment of the bowel (Fig.04). The peritoneum was inflamed, and saponification was noted on the omentum. There were multiple gastric erosions over the inferior surface of the stomach (Fig.05)

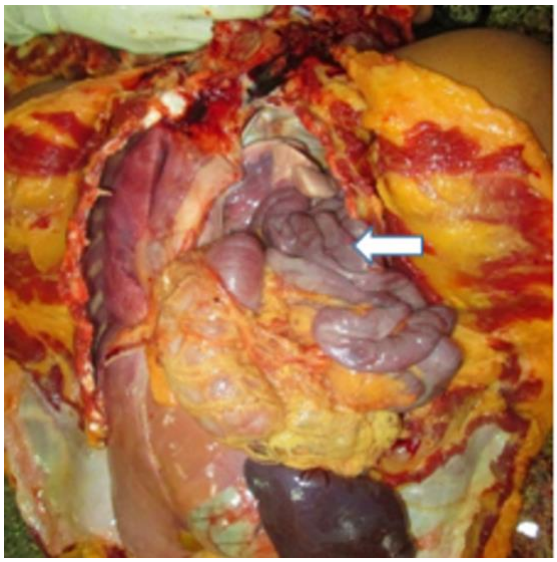


Figure 01. The stomach, duodenum and proximal part of the small intestine, splenic flexure of the transverse colon, and part of the omentum within the left hemithorax

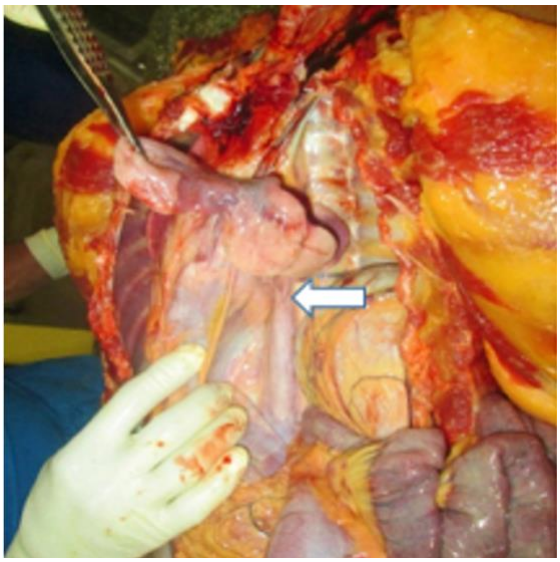


Figure 02. The defect in the left hemi-diaphragm (White arrow)

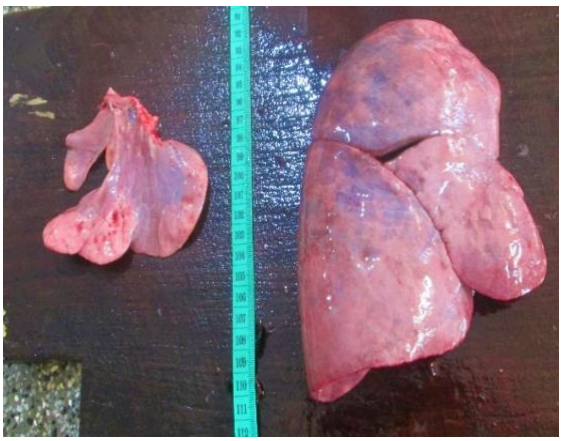


Figure 03. Hypoplastic and atelectatic left lung with relatively normally expanded right lung

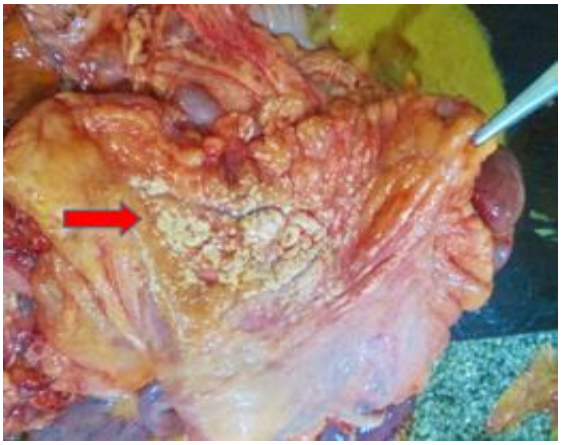


Figure 04. Inflamed omentum with Saponification. (Red arrow)

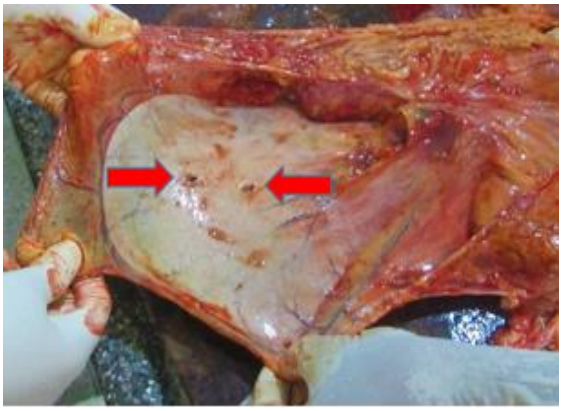
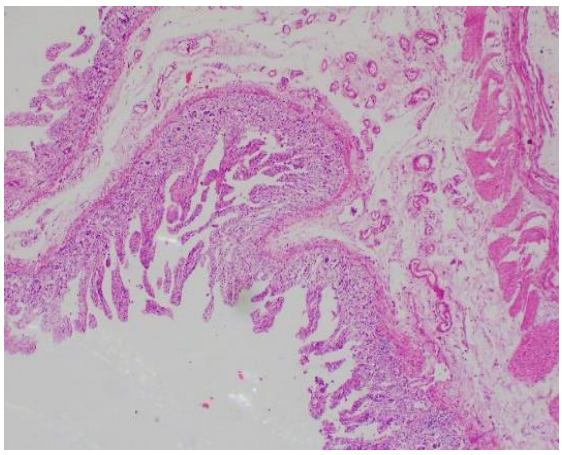


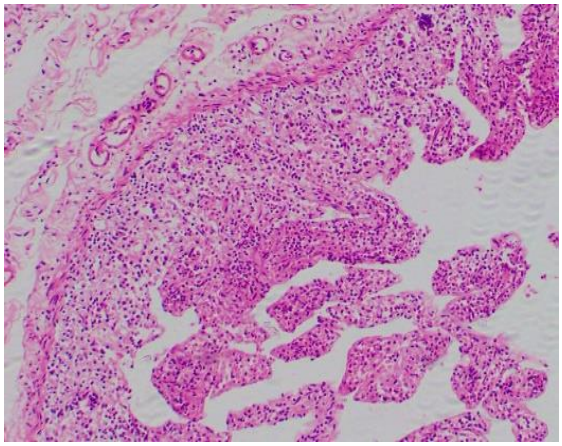
Figure 05. Multiple gastric perforations over the posterior wall of the stomach. (Red arrows)

Histology Findings

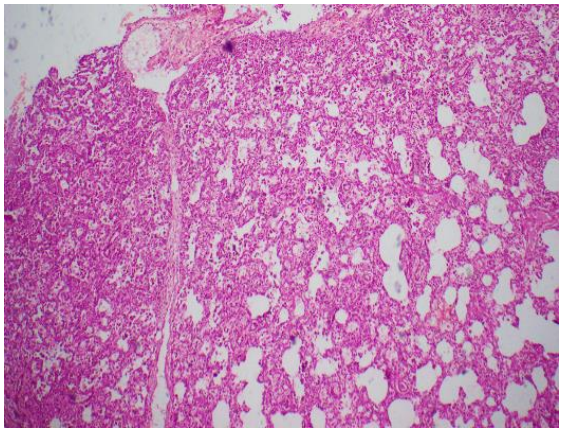
Microscopic examination of the lung tissue revealed pulmonary hypoplasia in the left side lung while the right lung was free of pathologies. There were features of ulceration and necrosis of the stomach wall in the sections taken from the areas of gastric perforations. The Cause of death was given as a death due to complications of late-onset diaphragmatic hernia.



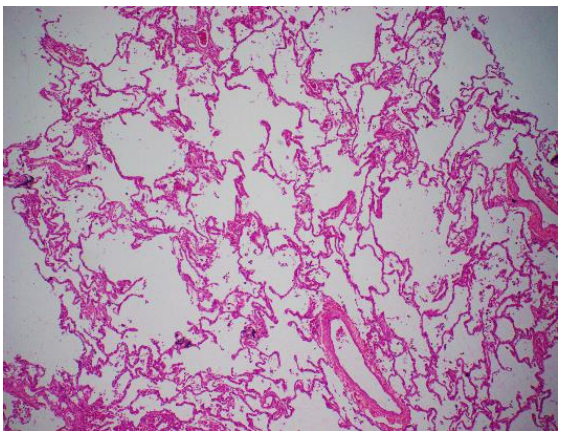
Photomicrograph 8-A. Erosions of the stomach wall (H & E x 4)



Photomicrograph 8-B. Erosions of the stomach wall (H & E x 10)



Photomicrograph 9-A. Hypoplastic atelectatic lung in histology (H & E x 4) with collapsed alveoli and limited air spaces



Photomicrograph 9-B. Right lung (H & E x 10) with normally expanded alveolar spaces

Discussion

There are two types of congenital diaphragmatic hernias (CDHs) named Bochdalek and Morgagni. Among them, Bochdalek hernias are more common in infants and mostly located on the postero-lateral part of the left hemi-diaphragm.[6] Though most diaphragmatic hernias are congenital in nature, they can be acquired following blunt force trauma, penetrating injuries, pregnancy, and complications of surgery. In this case, the defect was found on the posterolateral part of the left dome of the diaphragm and there was no history to suggest the development of an acquired diaphragmatic hernia which indicates this is a Bochdalek type CDH.

The late presenting CDH is difficult to differentiate with an initial chest X-ray and misdiagnosis is not uncommon.[7] Around 5-10% of CDH children may appear healthy during the newborn period, but abnormality can manifest in later life which was in this child. Chest X-ray is the commonest method of diagnosing a CDH but ultrasound, CT, MRI and upper and lowers GI contrast media are also useful.[8,9] In this case the child was asymptomatic and no radiological investigations had been performed prior to the death to diagnose the pathology.

In contrast to the high mortality and morbidity rates for neonatal CDH, the prognosis for late presenting CDH with early diagnosis and surgical correction is usually favorable.[10,11] Rarely they can be present with lung hypoplasia and malrotation also.[10] In this case, autopsy and microscopy findings revealed extensive lung hypoplasia, which indicates that the herniation of abdominal contents could have been there for a long period. Bowel pathologies like intestinal malrotation were also absent in this case.

There are two clinical scenarios in late-onset CDH in older children: presenting as an incidental finding on plain radiographs or presenting with symptoms develop due to incarceration, strangulation, and

visceral rupture inside the chest cavity.[12,13] In this case the child had presented with vague gastrointestinal symptoms like upper abdominal pain, nausea, and vomiting. Most gastric perforations are secondary to peptic ulcer disease. Gastric ulcers on top of the increasing luminal pressure inside the stomach may be the reason which leads to full-thickness stomach wall perforations. These perforations communicate between the gastric lumen with the peritoneal cavity. The gastric content is then free to enter the peritoneal cavity causing peritonitis. In this case, we found multiple perforations over the posterior wall gastric ulcers opening into the lesser sac causing chemical peritonitis which is usually confined and presents with less marked symptoms.

Gastritis and acute gastroenteritis are the most common differential diagnosis which presents to general practitioners in a primary care setting. As in this case, the chances of missing the diagnosis at a single visit to a general practitioner is fairly high due to the relatively uncommon and rare diagnosis presenting with vague clinical symptoms. Although the absence of respiratory symptoms like dyspnea or tachypnea may easily mask the conditions still there is possible medical negligence. But the late presentation and having limited investigation facilities may also lead to missing the diagnosis until the death. But using simple clinical examination techniques can elicit poor air entry and reduced chest expansion that lead to clinical suspicion of rare conditions like CDH. Although the chances of saving the life of the child are low, still there will be a possibility with prompt diagnosis and timely medical care.

Conclusions

This case highlights the importance of higher clinical vigilance and the value of early diagnosis and timely medical attention to fairly uncommon but surgically correctable conditions like CDH

Disclosure statement

Conflicts of interest: The author declares that she has no conflicts of interest.

Funding: None

References

1. Solomon N, Hayes JA Sudden Infant Death Due to Congenital Diaphragmatic Hernia. Acad Forensic Pathol. 2016; 6(4):720-30. doi:10.23907/2016.068
2. Phatak SV, Ghodke K. Antenatal diagnosis of congenital diaphragmatic hernia: A case report. Indian Journal of Radiology and Imaging. 2006; 16(4):793-5.
3. Sandra M, Nana N, Prem P. Congenital diaphragmatic hernia and retinoids: searching for an etiology. Pediatr Surg Int 2008; 24:755-61.
4. Kim, Jennifer M, Couluris M, Schnapf BM, Celi A, Juffermans, NP, Altraja. A. Late-presnting Left-Sided Morgagni Congenital Diaphragmatic Hernia in a 9-Year-Old Male. New York: International Scholarity Research Network, ISRN Pulmonology, 2011; 4(2):1-4.
5. Losty PD, Davenport M, Pierro A. Congenital diaphragmatic hernia. In: Davenport M, Pierro A, editors. Paediatric Surgery. New York: Oxford Press; 2009; 2(1):132-5.
6. Ameet K, Vikas M, Ramakrishnan TS, Samaresh S. Caecal perforation with faecal peritonitis – unusual presentation of Bochdalek hernia in an adult: a case report and review of literature. World Journal of Emergency Surgery 2009; 4(2):16.
7. Javier AA, Bindi NM, Oluyinka O, Olutoye. Delayed presentation of congenital diaphragmatic hernia manifesting as combined-type acute gastric volvulus: a case report and review of the literature. Journal of Pediatric Surgery 2008; 43(3):35-9.
8. Subrato J Deb. Massive right-sided Bochdalek hernia with two unusual findings: a case report. Journal of Medical Case Reports.2011; 5(2):519.
9. Salacin S, Alper B, Cekin N, Gulmen MK. Bochdalek hernia in adulthood: a review and an autopsy case report. J Forensic Sci 1994; 39(3):1112-6.
10. Samadhi DAH., Ruwanpura, PR. (2021). Death Due to Late Onset Diaphragmatic Hernia: Death Due to Diaphragmatic Hernia. Iranian Journal of Pediatric Surgery, 2021; 7(2):126-32. https://doi.org/10.22037/irjps.v7i2.27473.
11. Delaney CA, Rozance PJ, Sandoval JA, Bealer JF, Kinsella JP. Late onset diaphragmatic hernia complicated by intestinal strangulation. Curr Opin Pediatr. 2012; 24(2):274-6.
12. Blackstone MM, Mistry RD. Late-presenting congenital diaphragmatic hernia mimicking bronchiolitis. Pediatric Emergency Care. 2007; 23(9):653-6.
13. Ojo BA, Ngbea JA, Mohammed H, Ekeh BA, Ugwu VI, Otene B, Nyaga J, Umobong EO, Abayol P. Incidental Asymptomatic Diaphragmatic Hernia in An Adult At Postmortem. American Journal of Medical Sciences and Medicine. 2014; 2(5):96-8.