

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/radcr

Case report

Rare late-presentation congenital diaphragmatic hernia mimicking a tension pneumothorax ☆☆☆

Muhammad Zahid Abdul Muien, MD, DrRad^a, Kohila Jeyaprahasam, MBBS^b,
Thiagu Krisnan, MBBS, MMed^b, Chiak Yot Ng, MBBS, MMed(Rad)^a, Yong Guang Teh, MD,
DrRad^{a,b,*}

^a Faculty of Medicine & Health Sciences, Universiti Malaysia Sabah, 88400 Kota Kinabalu, Sabah, Malaysia

^b Department of Radiology, Sabah Women & Children's Hospital, 88400 Kota Kinabalu, Sabah, Malaysia

ARTICLE INFO

Article history:

Received 6 June 2021

Revised 8 June 2021

Accepted 8 June 2021

Keywords:

Congenital diaphragmatic hernia

Pneumothorax

Chest radiograph

CT

ABSTRACT

Congenital Diaphragmatic Hernia (CDH) is due to a defect in the diaphragm and is usually detected soon after birth. However, in rare cases, asymptomatic CDHs can be missed and present later in life. Late-presentation CDH can be misdiagnosed as tension pneumothorax leading to iatrogenic complications. We report a case of a 10-year-old boy who presented with non-specific symptoms of vomiting and occasional breathlessness, but was subsequently diagnosed as late-presentation CDH. This case highlights the role of imaging in the diagnosis and management of late-presenting CDH. The role of CT imaging as an invaluable tool to further evaluate equivocal radiographic findings in CDH is discussed.

© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

Congenital diaphragmatic hernias (CDHs) are caused by a defect in the diaphragmatic leaflet that permits the herniation of bowel loops, solid organs, and mesentery into the thoracic cavity. CDHs are usually diagnosed soon after birth, especially when a child is symptomatic. Patients usually present with vomiting, tachypnea, and cyanosis with reduced air entry in the affected lung on auscultation [1]. This constellation of symptoms would prompt the clinician to request a chest radiograph which would reveal the characteristic finding of bowel loops in one of the hemithorax.

Although most CDHs are usually detected soon after birth, in rare cases, asymptomatic CDHs can be missed and present much later in life. CDHs that are detected later than 30 days are considered late-onset CDH [2]. Later presentation of CDH in children ranges between 2 months and 12.5 years old. Misdiagnosis of a CDH can cause significant morbidity as they can be treated as tension pneumothorax leading to further complications due to inappropriate interventions [3]. Distinguishing between tension pneumothorax and CDH is particularly crucial as the management of these two diagnoses differs greatly.

☆ Funding: All authors have no funding source to declare.

☆☆ Acknowledgments: We would like to thank the Director General of Health Malaysia for permission to publish this report.

* Corresponding author.

E-mail address: tehyongguang@ums.edu.my (Y.G. Teh).

<https://doi.org/10.1016/j.radcr.2021.06.024>

1930-0433/© 2021 The Authors. Published by Elsevier Inc. on behalf of University of Washington. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

We present a case that highlights the role radiologic investigations have in distinguishing CDH and tension pneumothorax.

Case report

A 10-year-old boy, who was previously well, presented with a sudden onset of unprovoked postprandial vomiting of clear fluid for 3 days before admission. There was associated epigastric pain and a history of reduced appetite for several weeks. He also complained of occasional breathlessness for the past three months; otherwise, there was no other significant systemic complaints. According to his mother, he was active in sports and thriving at school. The child was initially assessed at a rural clinic and was unsuccessfully treated as gastritis. The symptoms persisted and the worried mother brought the child to a local District Hospital during the COVID-19 pandemic. The non-specific nature of the patient's symptoms did raise concerns of atypical COVID-19 infection. His COVID-19 polymerase chain reaction test was negative. The patient was subsequently transferred to our center for further management.

On examination, the boy was comfortable and not in respiratory distress. His blood pressure, heart rate, and respiratory rates were normal. A temperature of 37°C and a pain score of 0/10 was recorded. Reduced air entry over the left lung was heard on auscultation and dullness on percussion was elicited over the left lower zone. An erect chest radiograph revealed a hyperlucent left hemithorax. A curvilinear opacity within the left hemithorax and air-fluid level was also noted in the chest radiograph. The left costophrenic angle was blunted. Lung markings were seen at the left upper zone between this linear opacity and the thoracic wall. Tracheal deviation and mediastinal shift to the right was also seen on the radiograph (Fig. 1).

The absence of clinical signs of respiratory distress and the unusual radiographic findings prompted the attending paediatric team to request a CT thorax for further assessment. Despite being asymptomatic during admission, the mediastinal shift was a strong clinical concern that needed to be resolved promptly. The CT scan confirmed the mediastinal shift to the right (Fig. 2B) and revealed a Bochdalek hernia - a large defect in the posterior leaflet of the left hemidiaphragm - with a diameter of 4.8 cm. There was herniation of the stomach, omentum, and splenic flexure into the left hemithorax via this defect. The stomach was markedly distended, however, no evidence of perforated viscus or pneumothorax was detected in the CT images (Figs 2 and 3).

The patient underwent surgical repair a few days after diagnosis. Intraoperative findings confirmed the diagnosis of a left Bochdalek hernia with posterolateral diaphragmatic defect containing the dilated stomach, omentum, large bowel, and part of the spleen. Post-operative radiograph demonstrated normal expanded left lung with a clearly delineated left cardiac border and resolution of the mediastinal shift (Fig. 4). The patient recovered and was subsequently discharged well a week after the surgery. Follow-up assessment 3 months later documented that the boy was back to his normal daily routine with no active complaints.

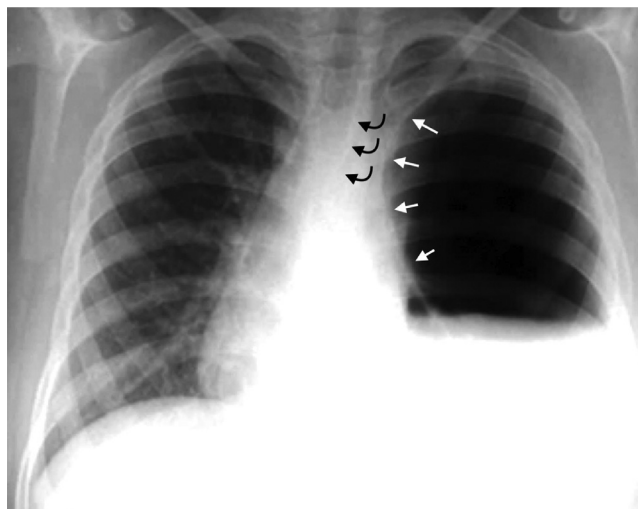


Fig. 1 – Hyperlucent left hemithorax with air fluid level. The curvilinear opacity outlining the hyperlucent area (white arrows) represents part of the herniated stomach. Lung markings are present at the periphery of the curvilinear opacity. Mediastinal shift and tracheal deviation to the right is evident (black curved arrows). The air-fluid level was formed by fluid in the stomach.

Discussion

CDH is usually diagnosed in neonates with an incidence of 2.3 in every 10,000 births [4]. Late-presenting CDHs are those that present after 30 days of life, and are rare, constituting only 2.6% of CDH cases [2]. Diagnosis is often challenging and misdiagnosis occurs up to 62% [5]. Depending on the size, different organs can herniate through the defect including the stomach, spleen, liver, and small bowels.

In patients with a large CDH, clinical signs at presentation may include respiratory distress, rapid heart rate, cyanosis, or even abnormal chest development [3]. In the case of asymptomatic patients or patients with intermittent symptoms, a diaphragmatic hernia may be missed and is commonly diagnosed incidentally on imaging studies much later on.

Close analysis of radiographic findings will help direct subsequent investigations. In our patient's radiograph, the presence of a thin curvilinear opacity seen adjacent to the mediastinum was very unusual for a pneumothorax. This is because in the case of pneumothorax, collapsed lung tissue is typically visualized as a dense homogeneous opacity at the hilar region. Critically, the incongruence between the clinical signs and the radiographic findings raised the suspicion of a congenital diaphragmatic hernia. However, the diagnosis of tension pneumothorax still needed to be ruled out as a mediastinal shift to the right was noted on the chest radiograph. This was the indication for performing a CT scan. The axial CT images demonstrated the presence of a mediastinal shift to the right (Fig. 2B) and revealed that the cause of the shift was due to a late-presenting CDH instead of a tension pneumothorax (Fig. 3).

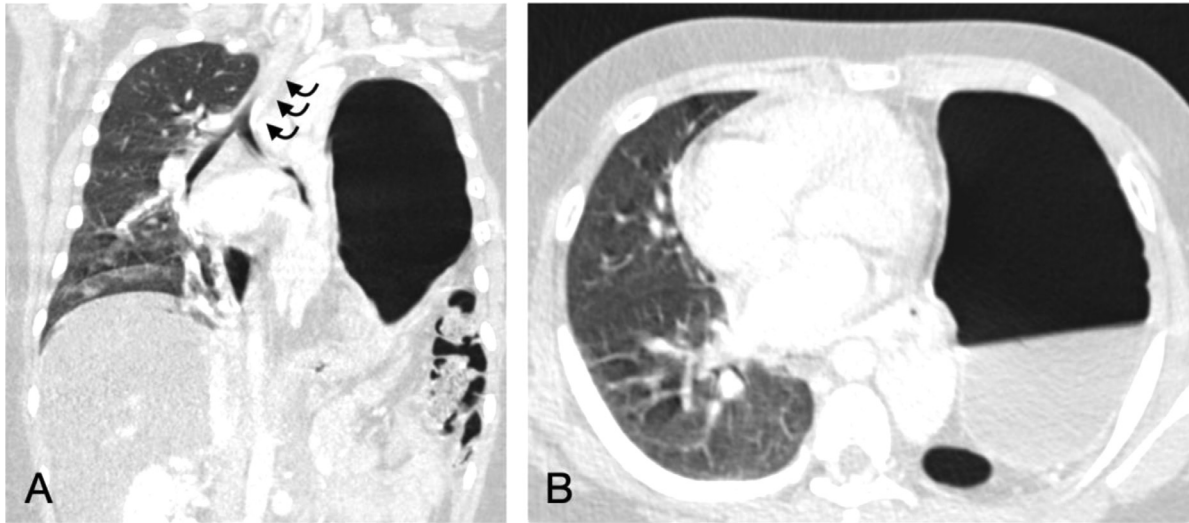


Fig. 2 – (A) Coronal reformatted CT image with lung windowing shows an air-filled cavity occupying the entire left hemithorax with tracheal deviation to the right (black curved arrows). (B) Axial CT image with lung windowing clearly demonstrates the mediastinal shift to the right.



Fig. 3 – Sagittal reformatted CT image with soft tissue windowing shows a defect in the posterolateral wall of the left hemidiaphragm (white curved arrows) with herniated stomach and bowel loops. An air-fluid level is seen within the distended stomach cavity.

Tension pneumothorax is a potentially fatal condition in which a one-way valve causes progressive hyperinflation of the hemithorax. The resultant mediastinal shift, if severe

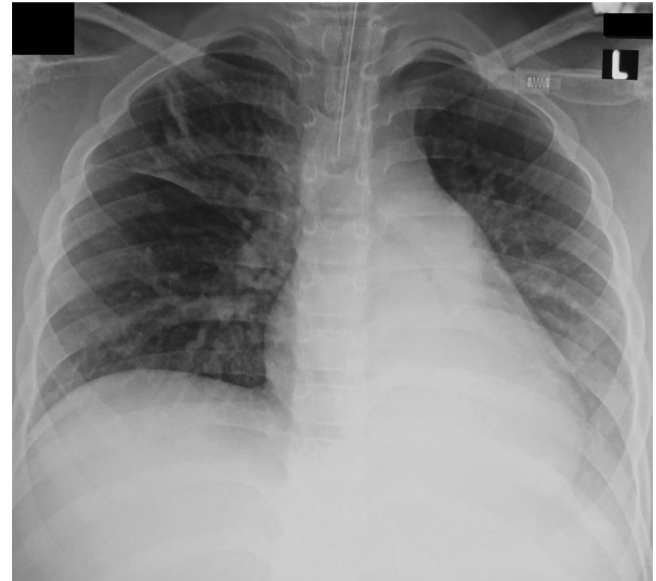


Fig. 4 – Postoperative portable radiograph showing an aerated left lung and visible left cardiac margin. The child was still intubated in the Intensive Care unit at that time.

enough, can precipitate cardiorespiratory collapse when the venous return is impeded by superior vena cava obstruction. Treatment of tension pneumothorax is immediate decompression followed by chest tube insertion. In our case, erroneous insertion of a chest tube into a dilated stomach or bowel loop would have significantly increased a patient's morbidity [3].

Although CT scans carry a relatively high radiation burden, it is a very useful modality for clarifying equivocal radiographic information as well as provides detailed anatomical information for pre-operative planning. In this case, the large

defect in the posterior leaflet of the left hemidiaphragm was clearly demonstrated on cross-sectional imaging. The CT scan images also clarified the radiographic findings observed on the chest radiograph and showed that the mediastinal shift was due to the mass effect from the herniated organs rather than a tension pneumothorax. In particular, the CT scan showed that the stomach was markedly distended and that the stretched stomach wall mimicked the appearance of pleural lining on the chest radiograph.

Surgical repair is the definitive treatment for CDH and favorable outcomes have been documented [6]. The goal of the operation is tension-free closure of the diaphragmatic defect [7]. Minimally invasive congenital diaphragmatic repair has been carried out via laparoscopy or thoracoscopy. The laparoscopy technique has been reported to be more technically challenging with risk of recurrence [8].

Conclusion

High clinical suspicion of late-presentation CDH is needed to diagnose this rare condition. Imaging findings of CDH may mimic that of tension pneumothorax, but close analysis of radiographic information is important to help clinch the diagnosis and guide subsequent investigations. This case has shown that correlation between imaging and clinical findings is crucial if the misdiagnosis of CDH is to be avoided. Although CT scans are not routinely performed in the paediatric population to diagnose CDH, owing to their significant radiation burden, they should be judiciously used in cases where radiographic data is equivocal.

Ethical approval

Not required.

Patient consent

Written informed consent was obtained from the patient's next of kin for the publication of this case report.

Guarantor

Dr Yong Guang Teh, Faculty of Medicine & Health Sciences, Universiti Malaysia Sabah

Declaration of Competing Interest

All authors have no conflict of interest to declare.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2021.06.024](https://doi.org/10.1016/j.radcr.2021.06.024).

REFERENCES

- [1] Bağlaj M. Late-presenting congenital diaphragmatic hernia in children: a clinical spectrum. *Pediatr Surg Int* 2004;20(9):658–69.
- [2] Kitano Y, Lally KP, Lally PA. Late-presenting congenital diaphragmatic hernia. *J Pediatr Surg* 2005;40(12):1839–43.
- [3] Mei-Zahav M, Solomon M, Trachsel D, Langer JC. Bochdalek diaphragmatic hernia: not only a neonatal disease. *Arch Dis Childhood* 2003;88(6):532–5.
- [4] Paoletti M, Raffler G, Gaffi MS, Antounians L, Lauriti G, Zani A. Prevalence and risk factors for congenital diaphragmatic hernia: a global view. *J Pediatr Surg* 2020;55(11):2297–307.
- [5] Berman L, Stringer D, Ein SH, Shandling B. The late-presenting pediatric Bochdalek hernia: a 20-year review. *J Pediatr Surg* 1988;23(8):735–9.
- [6] Hollinger LE, Harting MT, Lally KP. Long-term follow-up of congenital diaphragmatic hernia. *Seminars in Pediatric Surgery* 2017;26(3):178–84.
- [7] Dingeldein M. Congenital diaphragmatic hernia: management & outcomes. *Adv Pediatr* 2018;65(1):241–7.
- [8] Obata S, Souzaki R, Fukuta A, Esumi G, Nagata K, Matsuura T, et al. Which is the better approach for late-presenting congenital diaphragmatic hernia: laparoscopic or thoracoscopic? a single institution's experience of more than 10 years. *J Laparoendosc Adv Surg Tech* 2020;30(9):1029–35.