FLORONA
Jurnal Ilmiah Kesehatan
Vol 1 No 1 February 2022
ISSN: 2829-3835 (Print) ISSN: 2829-3983 (Electronic)
Open Access: https://jurnal.arkainstitute.co.id/index.php/florona/index

Congenital left hemidiaphragmatic agenesis: A case report

William Wijaya[1], Yustinus Rurie Wirawan[2]
[1,2]RSUD Dr. (H.C) Ir. Soekarno Provinsi Kepulauan Bangka Belitung
[1]*williamwijaya_dr@yahoo.com*, [2]*ruriewirawan@yahoo.co.id*

Article Info :
Received :
February 18th, 2022
Revised :
February 20th, 2022
Accepted :
February 25th, 2022

ABSTRACT

Congenital diaphragmatic hernia is a congenital diaphragmatic disorder characterized by herniation of the abdominal viscera into the thoracic cavity. Inrare diaphragmatic developmental abnormality cases, there is complete diaphragmatic agenesis. This study aims to report a case of left hemidiaphragmatic agenesis with a typical postnatal course of the disease. The research method used is descriptive qualitative. The results showed that a 13-day-old baby girl experienced respiratory problems for a week. Chest radiography showed left-sided congenital diaphragmatic hernia. It can be concluded that left diaphragmatic agenesis is a rare developmental disorder of the diaphragm. Surgical intervention as early as possible can prove beneficial for the patient. Patients can be treated surgically without using prosthetic materials by mobilizing and suturing the remaining diaphragm with the intercostal muscles.

Keywords: Congenital diaphragmatic hernia, diaphragmatic agenesis, Radiograph, Repair

INTRODUCTION

A 13-day old female infant presented respiratory distress for a week and was transferred from type C to type B hospital. The child was born by vaginal delivery with respiratory distress at the primary care center.

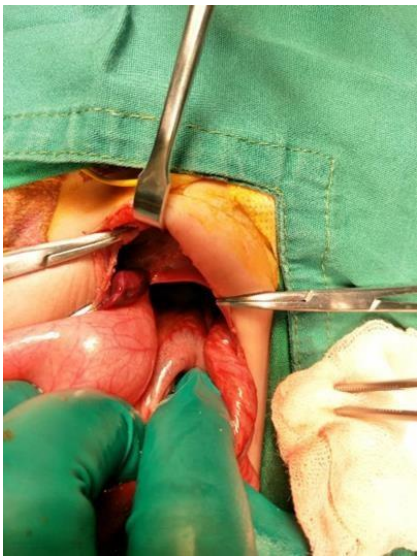

Figure 1. Operative view of the left hemidiaphragmatic agenesis. No diaphragmis seen on the lateral and anterior sides, whereas a small diaphragm edge can besutured anteriorly by stay suture to the intercostal muscles.

`

Oxygen therapy is given through the cannula. Ophthalmic prophylaxis wasperformed, and then she was referred to a type C hospital, where a nasogastric tube was applied, and antibiotic treatment with meropenem was initiated. KAEN 4B was administered with a metabolic flow of 16 drops per minute. The child was referred to a type B hospital to be managed by General Surgery.

The baby's weight was 2.6 kg during the admission, and a general physical examination revealed tachypnea (respiratory rate >50 min). Pulse was 143/min, and bloodpressure was normal. There was an overt sign of respiratory distress on respiratory systemexamination, such as tachypnea, nasal flaring, intercostal and subcostal retractions. On auscultation, there were bowel sounds in the left chest. Air entry was standard on right chest auscultation. The abdomen was soft and not much scaphoid. A chest radiograph was requested that revealed intestinal gas shadows in the left hemithorax. A diagnosis ofleft-sided CDH was made and was prepared for elective surgical intervention.

Her preoperative laboratory parameter was average. Treatment was started on parenteral antibiotics, and on the fifth day later, the patient will be planned for an electiveoperation. Parents were counseled about the surgical procedure and its postoperative complications. The operation was started with the left subcostal incision. Most of the small and large intestines and left liver were found herniating into the left hemithorax. The content was reduced back into the abdominal cavity. The diaphragm was absent on the left hemithorax's lateral and anterior sides, and only a small edge of the diaphragm was presented on the posterior sides [Figure 1].

The lung looked hypoplastic. No hernia sac was found during the operation. Thepart of the diaphragm present on the posterior aspect was mobilized and sutured to the intercostal muscles using interrupted stitches of PGA 2.0 sutures. The abdomen wound was closed by only skin closure. The postoperative recovery was good. On the fifth postoperative day, the nasogastric tube was removed. The next day was given oral nutrition, and the patient wasdischarged on the tenth postoperative day.

Congenital diaphragmatic hernia (CDH) is a congenital abnormality of the diaphragm, characterized by diaphragm defect accompanied by herniation of the gastro-intestinal and other abdominal viscera into the thoracic cavity (Chandrasekharan et al., 2017). There is also complete diaphragmatic agenesis in rare diaphragmatic developmental abnormality cases, which usually involves one side, but sometimes both sides (Mirza et al., 2012). However, it is still a management dilemma to fix these defects. The use of prosthetic materials, abdominal or intercostal muscle flaps, etc., are various techniques that have been tried and proposed to correct this defect. We report a rare case of congenital hemidiaphragmatic agenesis (CHDA), and successful repair without prosthetic materials.

RESEARCH METHODS

We report a case of left hemidiaphragmatic agenesis with usual postnatal course. A 13-day old female infant presented respiratory distress for a week. Chest radiograph showed left sided congenital diaphragmatic hernia. The research method used is descriptive qualitative, focusing on the clinical presentation, diagnostic procedures, and postnatal management of congenital diaphragmatic hernia cases. Data were collected through medical record analysis, radiological assessments, and interviews with the treating physicians to capture a comprehensive picture of the clinical pathway. The study provides insights into the challenges of managing rare congenital anomalies and discusses the potential implications for neonatal care. The infant's postnatal course was monitored over several weeks, highlighting the respiratory outcomes and recovery trajectory following surgical intervention.

RESULT AND DISCUSSION

Classical CDH named by Bochdalek, the first to identify the defect in 1848, is characterized by a posterolateral defect in the diaphragm (Mirza et al., 2012). The left- sided Bochdalek's CDH is more common and reported at approximately 80%. Left-sided CDH has Better Prognosis as compared to right-sided CDH (Partridge et al., 2016).

In the rare maldevelopment of the diaphragm cases, there is also completediaphragmatic agenesis, which usually involves one side, but sometimes both sides (Litwinska et al., 2015). Of all CDH, only 6% is reported with CHDA, and the left side is more common than the right side (Abe et al., 2018). In our case, the abnormalitywas on the left side, showing the common side of the lesion. Some bilateral CHDA caseshave been reported with incredibly high mortality, and cases such as this cannot be treated. The cause of CHDA is based primarily on speculation and assumptions and wasnot

`

well defined to date (I. Li et al., 2019). A failure of pleuroperitoneal membranedevelopment to close the pleuroperitoneal canal on the involved side is considered the probable etiology. Various drugs and environmental factors have been proposed to play a crucial role in CDH developmental (Wynn et al., 2013).

CDH patients' survival is determined by the severity of pulmonary hypertension and pulmonary hypoplasia in the first 24 hours. The more severe pulmonary hypertension and lung hypoplasia, the early the presentation will be, and the worse will be the outcomeand vice versa (Khan & Kerbel, 2018). The clinical features are usually vomiting, respiratory distress, and cyanosis. In our case, the patient could survive in the first 24 hours. That means the severity grade of pulmonary hypertension and pulmonary hypoplasia is not high (Mandell et al., 2021).

To delineate the diaphragm and its abnormalities, CT-Scan and MRI are necessary (Tavolaro et al., 2019). In our case, the diaphragmatic abnormality was clearly visible on the chest X-ray (Zefov & Almatrooshi, 2015).

Absent diaphragmatic tissue makes the surgical procedure very difficult. Although diaphragmatic tissue is present, it is still difficult to be repaired (De Coppi & Deprest, 2017). Some of the treatment options are using a prosthesis, abdominal wall, and chest wall muscle flaps, using the pre-renal fascia, suturing the chest margins with the liver, and using the pre-renal fascia (Zhou et al., 2018). However, some surgeons even suggest no repair (Van der List & DiFelice, 2017). Approximately 30-50% of CHD patients showed prosthesis-related complications and even hernia recurrence when treated with a mesh union in the defect (Bouma & Mulder, 2017; J. Li et al., 2014).

In contrast to our case, we have a small diaphragm edge. We tried to make it enough to close with the mobilization of the retroperitoneum and suturing it with the intercostal muscles. Although there is a possibility of an abdominal compartment after the liver and most of the small and large intestines return to the abdominal cavity. In ourpatient, skin closure went well.

CONCLUSION

It can be concluded that left diaphragmatic agenesis is a rare developmental abnormality of the diaphragm. Surgical intervention as early as possible proves to be beneficial for the patient. The patient can be managed surgically without incorporating aprosthetic material by mobilizing and suturing the remnants of the diaphragm with the intercostal muscles. Additionally, more research is recommended to further explore long-term outcomes of non-prosthetic repairs and to refine surgical techniques in managing diaphragmatic agenesis. Overall, this case contributes to the understanding of congenital diaphragmatic anomalies and offers insights into improving surgical management and prognosis in affected neonates.

REFERENCES

Abe, G., Li, I., Lee, S., & Ota, K. G. (2018). A novel allele of the goldfish chdB gene: Functional evaluation and evolutionary considerations. *Journal of Experimental Zoology Part B: Molecular and Developmental Evolution*, *330*(6–7), 372–383. https://doi.org/10.1002/jez.b.22831

Bouma, B. J., & Mulder, B. J. M. (2017). Changing landscape of congenital heart disease. *Circulation Research*, *120*(6), 908–922. https://doi.org/10.1161/CIRCRESAHA.116.3093

Chandrasekharan, P. K., Rawat, M., Madappa, R., Rothstein, D. H., & Lakshminrusimha, S. (2017). Congenital Diaphragmatic hernia–a review. *Maternal Health, Neonatology and Perinatology*, *3*, 1–16. https://link.springer.com/article/10.1186/s40748-017-0045-1

De Coppi, P., & Deprest, J. (2017). Regenerative medicine solutions in congenital diaphragmatic hernia. *Seminars in Pediatric Surgery*, *26*(3), 171–177. https://doi.org/10.1053/j.sempedsurg.2017.04.009

Khan, K. A., & Kerbel, R. S. (2018). Improving immunotherapy outcomes with anti-angiogenic treatments and vice versa. *Nature Reviews Clinical Oncology*, *15*(5), 310–324. https://www.nature.com/articles/nrclinonc.2018.9

Li, I., Lee, S., Abe, G., & Ota, K. G. (2019). Embryonic and postembryonic development of the ornamental twin-tail goldfish. *Developmental Dynamics*, *248*(4), 251–283.

`

https://doi.org/10.1002/dvdy.15

Li, J., Song, M., Jian, Z., Guo, W., Chen, G., Jiang, G., Wang, J., Wu, X., & Huang, L. (2014). Laboratory aspirin resistance and the risk of major adverse cardiovascular events in patients with coronary heart disease on confirmed aspirin adherence. *Journal of Atherosclerosis and Thrombosis*, *21*(3), 239–247. https://doi.org/10.5551/jat.19521

Mandell, E., Kinsella, J. P., & Abman, S. H. (2021). Persistent pulmonary hypertension of the newborn. *Pediatric Pulmonology*, *56*(3), 661–669. https://doi.org/10.1002/ppul.25073

Mirza, B., Bashir, Z., & Sheikh, A. (2012). Congenital right hemidiaphragmatic agenesis. *Lung India*, *29*(1), 53–55. https://doi.org/10.4103/0970-2113.92364

Partridge, E. A., Peranteau, W. H., Herkert, L., Rendon, N., Smith, H., Rintoul, N. E., Flake, A. W., Adzick, N. S., & Hedrick, H. L. (2016). Right-versus left-sided congenital diaphragmatic hernia: a comparative outcomes analysis. *Journal of Pediatric Surgery*, *51*(6), 900–902. https://doi.org/10.1016/j.jpedsurg.2016.02.049

Tavolaro, C., Ghaffar, S., Zhou, H., Nguyen, Q. T., Bellabarba, C., & Bransford, R. J. (2019). Is routine MRI of the spine necessary in trauma patients with ankylosing spinal disorders or is a CT scan sufficient? *The Spine Journal*, *19*(8), 1331–1339. https://doi.org/10.1016/j.spinee.2019.03.004

Van der List, J. P., & DiFelice, G. S. (2017). Primary repair of the anterior cruciate ligament: a paradigm shift. *The Surgeon*, *15*(3), 161–168. https://doi.org/10.1016/j.surge.2016.09.006

Wynn, J., Aspelund, G., Zygmunt, A., Stolar, C. J. H., Mychaliska, G., Butcher, J., Lim, F.-Y., Gratton, T., Potoka, D., & Brennan, K. (2013). Developmental outcomes of children with congenital diaphragmatic hernia: a multicenter prospective study. *Journal of Pediatric Surgery*, *48*(10), 1995–2004. https://doi.org/10.1016/j.jpedsurg.2013.02.041

Zefov, V. N., & Almatrooshi, M. A. (2015). Chest X-ray findings in late-onset congenital diaphragmatic hernia, a rare emergency easily misdiagnosed as hydropneumothorax: a case report. *Journal of Medical Case Reports*, *9*, 1–4. https://link.springer.com/article/10.1186/s13256-015-0755-9

Zhou, X., Chen, Q., Xu, G., Chen, Y., Wang, D., & Zhou, R. (2018). Anatomical characteristics of the fascial space during laparoscopic pancreaticoduodenectomy using cadaveric models. *Int J Clin Exp Med*, *11*(2), 1254–1259.