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Case Report

A Rare Case of Right-Sided Congenital Diaphragmatic Hernia with Two Defects

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Abstract

We here in report an unusal case of a patient with right Congenital Diaphragmatic Hernia (CDH) with two defects, which were noted during surgical repair, to highlight this rarely documented anatomical anomaly that warrants further investigation.

Keywords: Congenital diaphragmatic hernia; Neonate; Multiple defects; Chest radiography

Introduction

Congenital Diaphragmatic Hernia (CDH) represents approximately 8% of all congenital malformations, with an incidence of 0.8 per 10,000 live births [1]. The pathology underlying CDH includes a diaphragmatic defect that predisposes the patient to the herniation of abdominal contents into the thoracic cavity. The complex interplay among genetic, cellular, mechanical, and environmental mechanisms has been proposed to impact the integration of the muscles, connective tissues, and nerves responsible for the embryogenesis of the diaphragm [2]. There are three well-recognized types of CDH: posterolateral (Bochdalek), anterior (Morgagni), and hiatal hernias. However, a greater variability can be noted in the clinical presentations and outcome of various CDH types owing to the complex interactions among the above mentioned mechanisms [3]. We herein report an unusual case of a patient with right-sided CDH comprising two ipsilateral defects. Proper understanding and identification of this congenital anomaly during surgery is essential as overlooking this rare presentation may lead to postoperative confusion and increased morbidity.

Case Presentation

A full-term female neonate weighing 3.3 kg was referred to our hospital with a suspected diagnosis of right-sided CDH. Her 32-year-old mother (gravida, 3; para, 2) underwent regular antenatal follow-up examinations, the results of which were normal, and delivered through emergency caesarian section because of fetal distress. The APGAR scores were 5, 8, and 8 at 1, 5, and 10 min, respectively. The baby developed respiratory distress immediately after birth and required intubation and conventional ventilation with minimal settings. The venous cord blood gases were within normal range (pH, 7.36; pCO_2, 38 mmHg; and HCO_3, 21.8 mmol/L). Chest radiograph revealed right-sided diaphragmatic hernia. The neonate was referred to our hospital for further management at the age of 36 hours.

The examination performed at the time of admission revealed no apparent dysmorphic features. The patient required minimal ventilation settings with FiO_2 of 30% to achieve an oxygen saturation of 90% to 95%. The patient's vital signs were as follows: heart rate, 140 beats/min; blood pressure 66/32 mmHg; and mean arterial pressure 43 mmHg. The venous blood gas measurements showed the following results: pH: 7.26; pCO_2, 55 mmHg; HCO_3 22 mmol/L; and base deficit 1.3.

Chest radiography revealed intestinal loops with liver herniation into right hemi-thorax (Figure 1), accompanied with a contralateral mediastinal shift. Echocardiography revealed good cardiac function, severe pulmonary artery hypertension, Tricuspid Regurgitation (TR) of 58 mmHg, a small atrial septal defect, and patent ductus arteriosus with a left-to-right shunt. There was no difference between pre- and post ductal oxygen saturation. The patient did not require ionotropic support, and blood pressure remained within the normal range during the entire preoperative period.

On the third day of her life, the patient underwent right posterolateral thoracotomy through the sixth intercostal space. Intraoperatively, two defects, both having intact sacs were found. One was a large anterolateral diaphragmatic defect with the sac containing the liver, whereas the other was a smaller posterolateral diaphragmatic defect containing bowel loops (Figure 2).

The right lung was hypoplastic. The edges of both defects were surrounded with strong diaphragmatic muscles; therefore, closure of both defects was performed using silk 2/0 sutures after excision of sacs. Postoperative chest radiographs showed adequate expansion of both lungs and good repair of the right diaphragm (Figure 1). The patient required high-frequency oscillatory ventilatory support in the postoperative period because of respiratory acidosis. The patient was gradually weaned from the ventilator support and feeding was initiated, which was tolerated. The histopathological examination of

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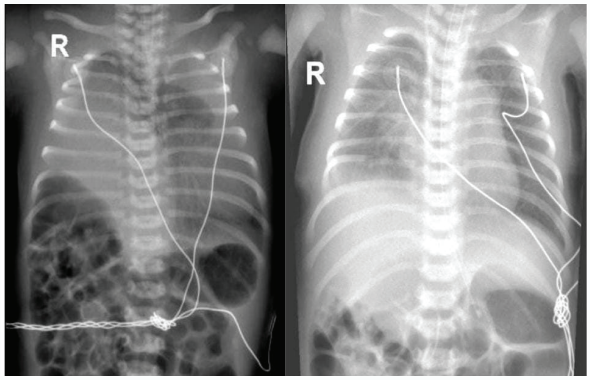

Figure 1: Plan radiograph showing: (a) right sided diaphragmatic hernia (b) Post-operative outcome.

the sac indicated the presence of fibrous tissue alone. The patient was discharged with follow-up scheduled in the outpatient clinic.

Discussion

Right-sided CDH is a rare condition representing 10%-15% of all CDH cases compared with left-sided CDH, which represents 75% to 90% of all cases [1]. Excluding hiatus hernia, the most common anatomic defect encountered during the surgical repair of CDH are posterolateral (Bochdalek) and retrosternal (Morgagni) hernias [4]. Other rarer defects can involve the anterolateral area and the central tendon or the entire hemidiaphragm (agenesis). Although rare, bilateral diaphragmatic defects have been reported in 1% ± 10% of all cases. A systematic analysis of patients with lethal diaphragmatic defects has revealed the presence of additional phenotypic variants such as multiple ipsilateral simultaneous defects [5,6]. However, there is a paucity of literature regarding the true incidence and underlying pathophysiology of these rarer phenotypic variants.

Timing of the complex embryologic development of the diaphragm has a direct bearing on the development of CDH pathologies [6]. Membranous closure of the diaphragm is complete by the eighth week of gestation. Thereafter, muscular fibers from the third, fourth, and fifth cervical myotomes reinforce the membranous folds, completing the closure by the ninth week of gestation. Normally, the midgut returns from its extracelomic herniation at approximately at the tenth week of gestation. Evidence suggests that the completion of this return before the closure of pleuroperitoneal canal might lead to its herniation into the pleural cavity in the absence of a sac. If the herniation occurs after the membranous closure of pleuroperitoneal canal but before muscular in growth, a sac covering the herniated viscera might be present [7]. Our patient had two defects with herniated intestines and liver covered with separate sacs, and a strong diaphragmatic muscle intervening the two defects.

Multiple unilateral defects in the diaphragm are extremely rare [5,6]. The presence of multiple small defects can be attributed to muscle degeneration during diaphragm development, and the resultant diaphragm is termed cribriform [8]. Another hypothesis to explain multiple unilateral diaphragmatic defects is the failure of complete muscular buttressing, which results in a diaphragmatic defect [6].

Compared to those with left-sided CDH, proportionally more patients are diagnosed with right-sided CDH after birth. Studies suggest that right-sided CDH is more likely to be missed in the antenatal period, likely because of the resemblance of the liver to pulmonary parenchyma on ultrasound and the presence of fewer intestinal loops in the thorax [9]. This finding was similar to that observed in our patient, in whom the antenatal follow-up failed to detect the pre-existing condition. Previous studies have reported favorable outcomes in neonates with CDH comprising an intact hernial sac because of better fetal lung growth, lower probability of herniation of abdominal contents, and better postnatal outcome [10].

In the present case, intraoperatively, the CDH appeared as a single defect; however, the second defect was identified during attempt to reduce the contents of the first defect. Therefore, proper identification of multiple defects is essential during the surgical repair of diaphragmatic hernia.

Evidence confirm that right-sided defects are associated with worse outcomes than left-sided defects owing to the larger defect size; position of the liver; and severity of pulmonary hypoplasia, pulmonary hypertension, and associated cardiac anomalies. These factors can predict the possible challenges in surgical repair [11]. In our patient, one of the defects contained the liver and could be reduced to abdominal cavity without an increase in the intra-abdominal pressure; therefore, tension-free repair was successfully achieved despite the presence of two defects. The utility of various patch methods have been described in larger defects [12]. Hepatopulmonary fusion, a potential

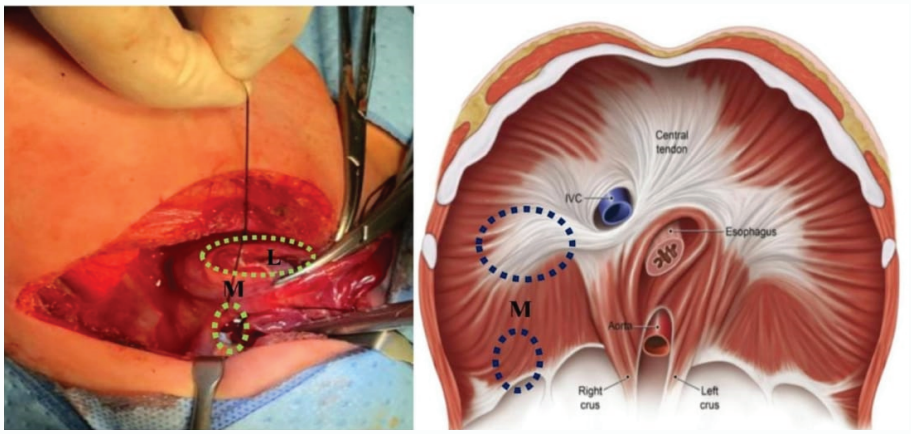

Figure 2: Two defects in diaphragm (dotted lines) seperated by thick muscle tissue (M), Liver (L) also visible.

anatomic anomaly that has been uniquely associated with right-sided defects [13], was not observed in the present case. Previous studies have reported the prolonged requirement of respiratory support, including high-frequency ventilation and extracorporeal membrane oxygenation, in the postoperative period because of pulmonary morbidity in patients with right-sided CDH [14]; this approach was not used for our patient who required high-frequency ventilation for 48 hours and gradually weaned off to a conventional settings with subsequent extubation.

In conclusion, our rare case of a patient with right-sided CDH and two defects highlights the importance of careful surgical inspection of the diaphragmatic anatomy and aims to raise awareness regarding potential multiple defects in CDH.

References

1. Kirby E, Keijzer R. Congenital diaphragmatic hernia: current management strategies from antenatal diagnosis to long-term follow-up. Pediatr Surg Int. 2020;36(4):415-29.
2. Kardon G, Ackerman KG, McCulley DJ, Shen Y, Wynn J, Shang L, et al. Congenital diaphragmatic hernias: from genes to mechanisms to therapies. Dis Model Mech. 2017;10(8):955-70.
3. Longoni M, Pober BR, High FA. Congenital diaphragmatic hernia overview. In: Adam MP, Ardinger HH, Pagon RA, editors. Gene Reviews. USA: Seattle University of Washington; 2020.
4. Chandrasekharan PK, Rawat M, Madappa R, Rothstein DH, Lakshminrusimha S. Congenital diaphragmatic hernia - a review. Matern Health Neonatol Perinatol. 2017;3:6.
5. Akel S, Nasr W. Multiple ipsilateral congenital diaphragmatic pathologies: rarities to consider. Eur J Pediatr Surg. 2001;11(3):200-3.
6. Urushihara N, Nakagawa Y, Kawashima K, Fukazawa T, Watanabe Y, Todani T. Congenital right diaphragmatic hernias through posterolateral and anterolateral defects associated with extralobar pulmonary sequestration: a case report. J Pediatr Surg. 1997;32(1):101-2.
7. Heiwegen K, van Heijst AF, Daniels-Scharbatke H, van Peperstraten MC, de Blaauw I, Botden SM. Congenital diaphragmatic eventration and hernia sac compared to CDH with true defects: a retrospective cohort study. Eur J Pediatr. 2020;179(6):855-63.
8. Appelquist E, Høier-Madsen K. Cribriform diaphragm: A variant of congenital diaphragmatic herniation. A case report. Scand J Thorac Cardiovasc Surg. 1986;20(2):185-7.
9. Burgos CM, Frenckner B, Luco M, Harting MT, Lally PA, Lally KP, et al. Right versus left congenital diaphragmatic hernia - What's the difference? J Pediatr Surg. 2017;S0022-3468(17):30649-8.
10. Zamora IJ, Cass DL, Lee TC, Welty S, Cassady CI, Mehollin-Ray AR, et al. The presence of a hernia sac in congenital diaphragmatic hernia is associated with better fetal lung growth and outcomes. J Pediatr Surg. 2013;48(6):1165-71.
11. Abramov A, Fan W, Hernan R, Zenilman AL, Wynn J, Aspelund G, et al. Comparative outcomes of right versus left congenital diaphragmatic hernia: A multicenter analysis. J Pediatr Surg. 2020;55(1):33-8.
12. Saxena AK. Surgical perspectives regarding application of biomaterials for the management of large congenital diaphragmatic hernia defects. Pediatr Surg Int. 2018;34(5):475-89.
13. Ferguson DM. Congenital Diaphragmatic Hernia Study Group. Hepatopulmonary fusion: A rare variant of congenital diaphragmatic hernia. J Pediatr Surg. 2020;55(9):1903-7.
14. Partridge EA, Peranteau WH, Herkert L, Rendon N, Smith H, Rintoul NE, et al. Right- versus left-sided congenital diaphragmatic hernia: a comparative outcomes analysis. J Pediatr Surg. 2016;51(6):900-2.