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(CASE REPORT)

Right sided congenital diaphragmatic hernia

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Abstract

Introduction: Congenital diaphragmatic hernia is a rare affection, has been classified as posterolateral, anterior or central. The posterolateral defect occurs on the left side (85%), although it may involve the right side (13%) or be bilateral (2%).

Patients and methods: We report the evolution after surgical treatment of right diaphragmatic hernia in one case.

Results: A male neonate, born via uncomplicated vaginal delivery at a gestational age of 10 days, weighed 3.8 kg at birth and initially presented with weak crying and a dusky skin tone. He required intubation and was placed on a ventilator. Subsequent imaging studies including chest radiography and CT scan revealed a right congenital diaphragmatic hernia, while ultrasound confirmed the presence of pulmonary hypertension.

Following stabilization, the newborn underwent thoracotomy surgery, during which intraoperative exploration revealed the entire liver within the thoracic cavity along with intestinal loops. The liver was challenging to maneuver but was successfully relocated into the abdomen, and the intestinal loops were reduced. Repair of the diaphragmatic defect was accomplished using non-absorbable sutures.

Following surgery, the neonate was transferred to the neonatal intensive care unit (NICU), where a postoperative chest radiograph on the fourth day showed expansion of the right lung. Over the subsequent two years, the patient's condition remained stable without the development of respiratory complications or worsening symptoms.

Conclusion: Success in this difficult case is optimized by close cooperation between the neonatologist, anesthesiologist, and pediatric surgeon. Although the hospital stay was prolonged, the survival outcome is excellent in this high-risk case.

Keywords: Congenital diaphragmatic hernia; Pulmonary hypertension; Liver intra-thoracic; Neonatal intensive care unit

1. Introduction

Embryologically, the diaphragm develops between the 8th and 12th week of gestation. The septum transversum separates the thoracic cavity from the abdominal cavity, and muscle fibers migrate into this membrane. Left-sided anomalies are more common because of the late closure of this membrane. The loss of lung development may be explained by the loss of space to develop, the diagnosis of CDH can be made before birth by ultrasound in 90% of cases. Fetal ultrasound findings include polyhydramnios, intestinal loops in the chest, echogenic chest mass.

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Initial postnatal treatment is aimed at resuscitating and stabilizing the infant in cardiopulmonary distress. Pulmonary hypertension and associated cardiac abnormalities are assessed by echocardiography. The timing of surgery is based on the clinical judgment and discretion of the surgeon.

In this case, we aimed to share our experience in the management of this affection.

2. Case report

A male newborn, 10 days old gestational age, was delivered via uncomplicated vaginal birth. He weighed 3.8 kg, exhibited faint crying, and had a dark skin tone upon birth. He was immediately intubated and placed on a ventilator (HFO ventilation with a mean pressure of 13, FiO2 100%). A chest radiograph (see figure 1) and CT scan confirmed the presence of a right congenital diaphragmatic hernia. Ultrasound also detected pulmonary hypertension, while other examinations yielded normal results. Following stabilization, the newborn underwent surgical intervention.

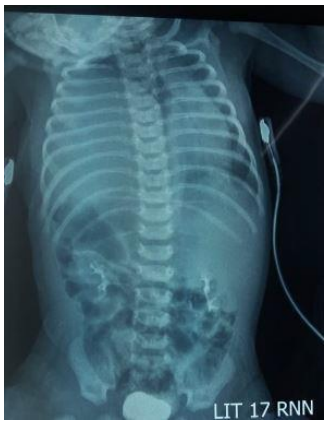

Figure 1 Homogeneous pulmonary opacity (shriveling the lung at the top and pushing back the trachea on the left) with image of colon filling the base on the right

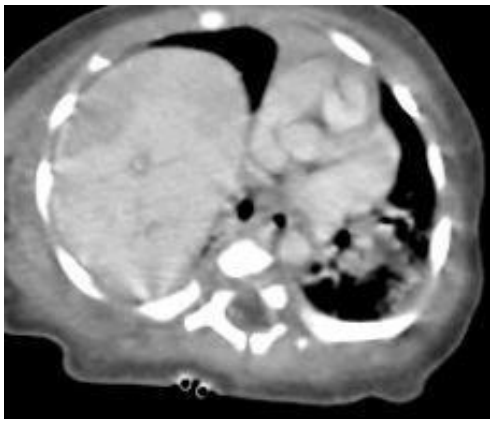

Figure 2 CT scan of chest showing liver at the level of heart

Surgery was performed by thoracotomy at the level of the 5th right intercostal space. The entire liver was intra-thoracic with the intestinal loops. The liver was twisted at 90 degrees in the thoracic cavity. The stomach and spleen were found in the abdomen. Only a thin rim of the diaphragm on the anterior aspect was present, and the posterior rim was absent. (Figure 3). The right lung was very small (Figure 4).

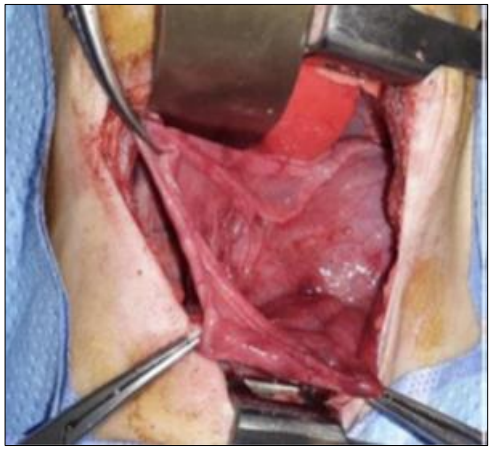

Figure 3 Surgical image showing the appearance of the diaphragm with absence of the posterior border

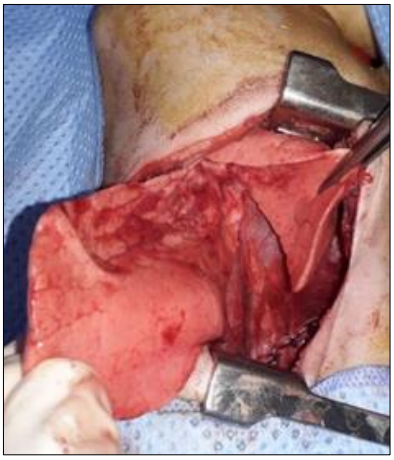

Figure 4 Surgical image showing pulmonary hypoplasia

The liver could be lowered into the abdomen with difficulty, and the intestinal loops were reduced. Diaphragm repair was performed with 2-plane with non-absorbable sutures through the anterior border and pleural flap. (Figure 5)

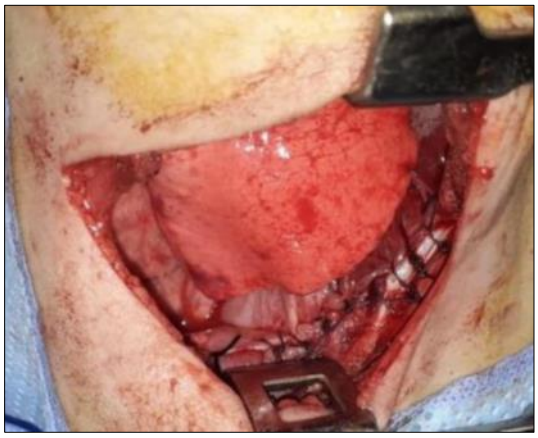

Figure 5 Appearance of the diaphragm after closure

The abdomen was primarily closed without significant tension. Following surgery, the baby was transferred back to the neonatal intensive care unit and maintained on high-frequency oscillatory ventilation. On the fifth day after surgery, the baby began nasogastric feeding, initially at half strength and then progressed to full strength. A chest radiograph on the fourth postoperative day revealed expansion of the right lung, and the baby was successfully weaned off ventilation after two weeks. Remarkably, the baby experienced a smooth recovery and was discharged from the hospital on the twenty-ninth postoperative day. Regular follow-up appointments have been scheduled.

With a hindsight of two years, the baby's progress has been favorable, with no exacerbation or onset of respiratory complications.

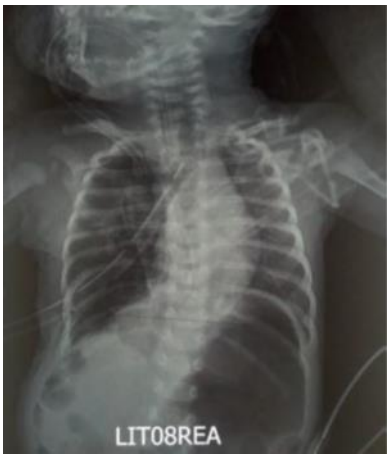

Figure 6 Chest X-ray in the immediate postoperative period

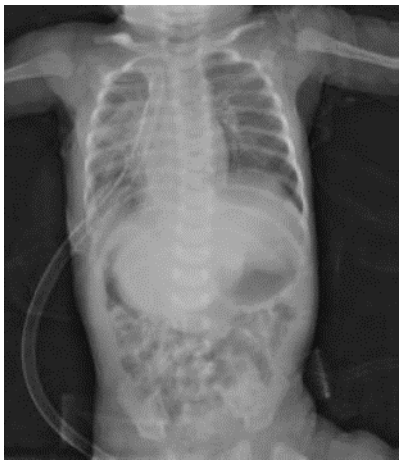

Figure 7 Postoperative day four, showing lung expansion without deviation of the mediastinum

3. Discussion

Right-sided congenital diaphragmatic hernia (CDH) have been associated with higher morbidity and mortality than left-sided defects [1, 2]. Whether the side of the anomaly has prognostic value remains controversial, as other groups have reported no difference [3]. Or better survival [4]. For those who were managed before birth.

It is clear that the size of the anomaly is inversely correlated with survival and is the strongest predictor of outcome, both in terms of survival and morbidity [5, 6].

The surgical repair was difficult in this right sided CDH because the size of the defect. Was more than four centimeters. Only a thin rim of diaphragm was present anteriorly and the posterior rim was absent completely. The reduction of liver posed a difficult problem. Liver replacement in the abdomen can be complicated by kinking of hepatic veins causing profound hypotension. Potential anatomic anomalies such as possible hepatopulmonary fusion [7, 8] anomalous venous drainage uniquely associated with right sided defects.

Survival based on liver herniation alone is 43% as compared to 93% survival without liver herniation [9]. The series published by Fischer et al [10]. Has shown the survival rate (right CDH 55% to left CDH 77%) ECMO requirement (right CDH 40% Vs left CDH 15%) prosthetic material in R CDH Vs L CDH (76% Vs 41%) and abdominal wall (38% Vs 19%) repairs. These data support that right side CDH carries a high mortality and morbidity.

The repair of a CDH may be as variable as clinical management. A thoracic approach is our attitude. While other surgeons prefer a transverse incision on the ipsilateral side, the type of repair is dependent on the size of the defect. If the defect is small, a tension free primary surgical closure should be performed with non-absorbable sutures. If the defect is wide primary closure may be attempted by one of the patch methods (Prosthesis, Muscle Flap [11, 12, 13]. Bioactive Material [14].)

4. Conclusion

Preoperative physiologic stabilization and subsequent elective repair have become the cornerstone of CDH management as in our case. The introduction of a treatment protocol, using high-frequency oscillatory ventilation from birth, a vasodilator, and delayed surgical repair after stabilization, improved the survival of this neonate with CDH. Success in this difficult case is optimized by close cooperation between the neonatologist, anesthesiologist, and pediatric surgeon. Although the hospital stay was prolonged, the survival outcome is excellent in this high-risk case.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

There are no conflicts of interest or acknowledgements.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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