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Congenital Diaphragmatic Hernia in a Neonate, Case Report

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Abstract | **Case Report**

Congenital Diaphragmatic hernia is a condition characterized by a defect in the diaphragm leading to the herniation of abdominal contents into the thoracic cavity interfering with normal development of the lungs. It is associated with pulmonary hypoplasia, pulmonary hypertension and heart failure. We present a case report for a neonate presented with cyanosis, grunting and tachypnea after birth and diagnosed as left sided Congenital Diaphragmatic hernia. The infant run a difficult course after surgery.

Keyword: Congenital Diaphragmatic hernia, pulmonary hypoplasia, pulmonary hypertension.

INTRODUCTION

Congenital Diaphragmatic hernia is a condition characterized by a defect in the diaphragm leading to protrusion of abdominal contents into the thoracic cavity interfering with normal development of the lungs. The defect may range from a small aperture in the posterior muscle rim to complete absence of diaphragm [1]. There is a significant morbidity and mortality due to associated pulmonary hypoplasia, pulmonary hypertension and heart failure in about 30% of CDH patients [2].

CASE REPORT

A male infant of healthy non consanguineous parents was born at 38 weeks by normal spontaneous vaginal delivery to a 27 years old primigravida mother. Pregnancy and delivery were uncomplicated. Apgar score was 5 at first mint and 7 at fifth minutes. Birth weight was 3.1kg. After birth the infant suddenly developed cyanosis, grunting tachypnea and diminished air entry on the left side, the heart was present on the right side of the chest. The abdomen was scaphoid. The infant was intubated immediately, and admitted to neonatal intensive care unit for further evaluation. The infant was transfered to our hospital at age of 6 days for surgery from privet hospital.

The mother was booked and had normal antenatal U/S scans. She had no history of diabetes mellitus, hypertension, fever, and hospital admission or medication intake.

Vital Signs

Temp: 36.5 C°, Heart rate: 170 beat/minute, Respiratory rate: 45 breath/minutes, Pre and post ductal SPO_2: 95 % & 98%, Blood pressures: 63/41 mmHg.

INVESTIGATIONS

- Complete Blood Count: WBC: 10.2, hemoglobin: 14.1, platelets 203.
- Kidney function tests: Urea: 3.3mmol/L, Creatinine: 76.6 mmol/L, Sodium 136 mEq/L, Potassium: 4.7mEq/L, Calcium: 2.39 mmol/L.
- Liver function tests: aspartate aminotransferase: 37 u/l, alanine aminotransferase: 16, alkaline phosphatase: 190, total.billrubin:192.3, direct.bilrubin:3.38, albumin: 21.6g/l.
- Coagulation profile, prothrombin time: 15sce, INR: 1.16, partial thromboplastin time: 40.3.
- CBG: PH: 7.31, PCO_2:55.7 mmHg, HCO_3:22.9 mmol/L.
- Chest x-ray (Figure1) showed left side diaphragmatic hernia with bowel loops inside the thoracic cavity, Right sided lung field shows the homogeneous opacity with some linear reticular margins, Normal right costophrenic angle

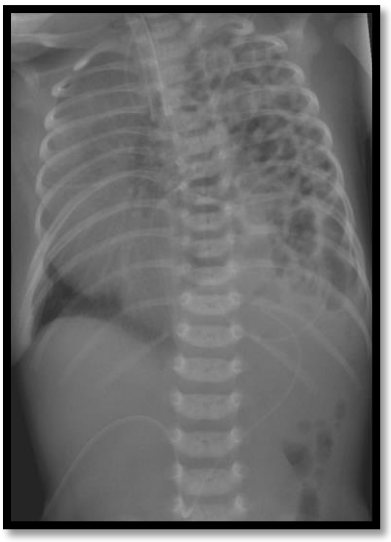

Fig-1: Chest x ray showed left side diaphragmatic hernia, with bowel loops inside the thoracic cavity

- Echocardiography: showed patent foramen ovale, persistent pulmonary hypertension (Right ventricle pressure 85mmHg).
- Abdomenal and Brain US: were normal.
- Blood Cultures: No growth.

The infant was started on intravenous fluids then total parenteral nutrition (TPN), intravenous antibiotics, mechanical ventilation, analgesia, and inotropes. Surgical consultation was done and booked for surgery after stabilization. Surgery was done at age of 8 days of life

Intraoperatively

Spleen, small bowel, and large bowel till splenic flexure were in the Left side of the chest. The diaphragm was deficient anteriorly and posteriorly. All abdominal contents were withdrawn from the chest cavity. Left lung was not developed, and chest drain was placed (Figure 2).

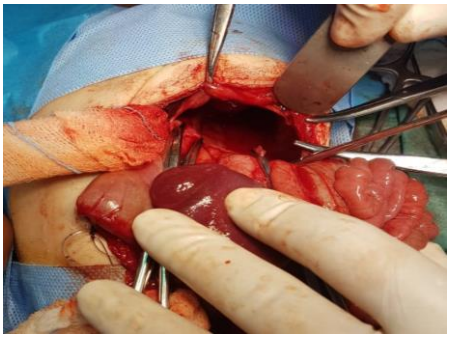

Fig-2: Intraoperatvely spleen, small bowel and large bowel till splenic flexure were in the Left side of the chest

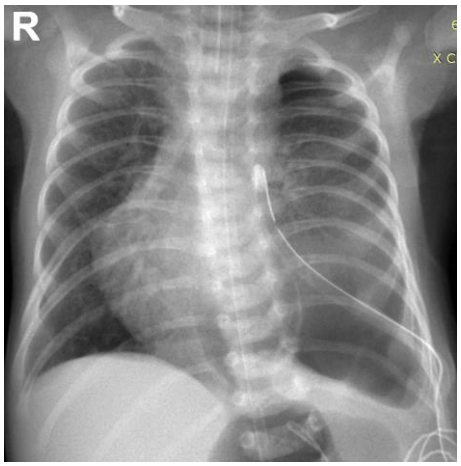

Fig-3: Showed improved chest x ray postoperatevly, however patient was still critically ill on high freqncy ventilator

Postoperative the infant was critical; so shifted to HFOV and sedated with midazolam and fentanyl infusions. He developed severe hypotension so dopamine and dobutamine infusions were commenced. He developed fever on day11; so septic screen was taken and antibiotics changed. Blood culture grew gram positive cocci in clusters. He developed severe PPHN and nitric oxide started. Condition continued critical and he got cardiopulmonary arrest not responded to active resuscitation; and the baby was declared dead at age of 16 days.

DISCUSSION

Congenital Diaphragmatic hernia (CDH) is characterized by a defect in the diaphragm leading to the protrusion of abdominal contents into the thoracic cavity affecting the normal development of the lungs. The condition may present as an isolated lesion or as part of a syndrome. The incidence of CDH based on the available literature ranges from approximately 0.8 - 5/10,000 births and varies across the population [1-4].

The etiology of CDH largely remains unclear and currently is thought to be multifactorial. The majority of the cases have an isolated diaphragmatic defect presenting with pulmonary hypoplasia and persistent pulmonary hypertension of newborn (PPHN). CDH can be associated with cardiac, gastrointestinal, genitourinary anomalies or with chromosomal aneuploidy such as trisomies. Multiple genetic factors along with environmental exposures and nutritional deficiencies have been proposed to be the possible etiologies for CDH [5-7]. Prenatal diagnosis by ultrasound detects more than 50% of CDH cases at a mean gestational age of 24 weeks [8].

Most common associated chromosomal abnormalities are the trisomies 18, 13 and 21[9]. Deliveries should be conducted at centers with capabilities of managing an infant with CDH and associated complications. Resuscitation in the delivery room is based on neonatal resuscitation program (NRP) guidelines [10].

Ultrasound measurement of observed to expected lung/head ratio should be used between 22 and 32 weeks of gestational age to predict the severity of pulmonary hypoplasia in isolated CDH[11]. Fetal magnetic resonance imaging (MRI) should be used (where available) for the assessment of lung volume and liver herniation in moderate and severe CDH [11].

The neonatal resuscitation guideline from the American Heart Association and the American Academy of Pediatrics supports immediate endotracheal intubation for neonates with a known diagnosis of CDH, and thus the strict avoidance of bag-valve-mask ventilation for these patients [12].

Two standardized echocardiograms, one within 48 hours of birth and one at 2 to 3 weeks of life, are needed to assess pulmonary vascular resistance, as well as left ventricular and right ventricular function. Additional studies may be conducted as clinically indicated.

Open surgical repair should generally be delayed until the infant is “stable.” Long-term disability surveillance is essential, especially in the high-risk cohort, and should be managed by interdisciplinary teams of primary care physicians, pediatricians, pediatric subspecialists, pediatric surgeons and other allied health providers [11].

CONCLUSION

Congenital diaphragmatic hernia is rare neonatal entity and should be considered in neonate with respiratory distress and cyanosis in the first minutes or hours of life, although a later presentation is possible. High index of suspicion, early diagnosis and management can save patient from serious complications.

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