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Research Article

Newborn female with congenital diaphragmatic hernia (bochdalek hernia): A case report.

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ABSTRACT

We reported a case of a newborn baby girl with congenital diaphragmatic hernia. This caused by a defect of posterolateral diaphragm during the embryological period, resulting in discontinuity of the diaphragm. The mortality of infants with congenital diaphragmatic hernia remains high, despite of optimal perinatal care. This case report presents a successful postnatal diagnosis and treatment of a newborn girl patient with congenital diaphragmatic hernia. Treatment is by intubation and surgical repair of the diaphragm. The patient was then discharged from the hospital, no additional symptoms were found after 3 months.

Keywords: *Congenital, Diaphragmatic, Hernia, Bochdalek, Radiograph*

INTRODUCTION

Congenital diaphragmatic hernia (CDH) is a developmental closure defect, resulting in discontinuity of the diaphragm. This allows abdominal viscera to herniate into the chest. There are several types of diaphragmatic hernia, in which 95% of the defect is located in the posterolateral diaphragm which known as Bochdalek hernia. The defect of the posterolateral left side of the diaphragm accounts for 75%-90% of the cases, but it could also occurred on the right side.(1)

The mortality rate of infants with congenital diaphragmatic hernia remains high, despite of optimal perinatal care, in which pulmonary hypoplasia and associated persistent pulmonary hypertension also has a big role. Management strategies such as permissive hypercapnia, high frequency oscillatory ventilation (HFOV), inhaled nitric oxide (NO), extracorporeal membrane oxygenation (ECMO) and delayed surgical repair have emerged in the care of high-risk patients which offer some hope and improving overall survival.(1)

CASE REPORT

We present a female patient, born at term from a 25-year old mother by caesarean section upon request. Her mother completed antenatal care as directed by an obstetrician and no problems were recorded during her examination. After delivery the baby was born well, cried immediately but then in just a few moments looked cyanotic and had difficulties in breathing. The baby then transferred to Neonatal Intensive Care Unit (NICU) and Chest x-ray was done for diagnosis. According to figure 1, right congenital diaphragmatic hernia, mediastinal shifting to the left, compressed lung due to bowel herniation was observed. Structure of the liver was shown vaguely on the right upper side of the abdomen.

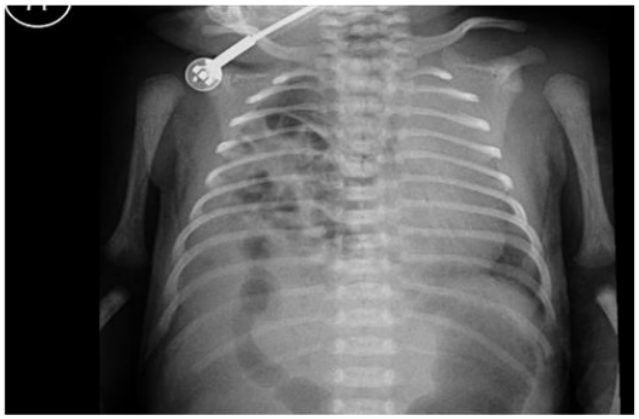


Fig.1: Antero-Posterior Chest X-ray After Birth

The next examination was done. Figure 2 was taken 30 minutes after contrast oral agents were given.

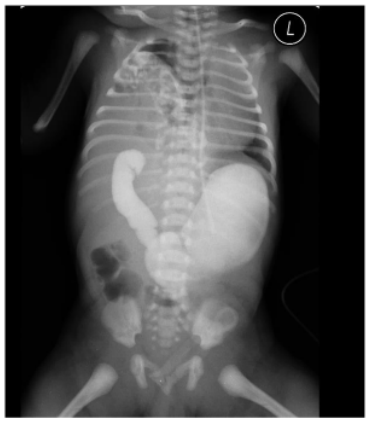
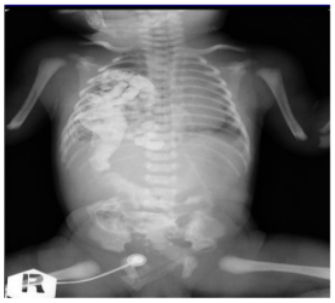


Fig.2: After contrast agents



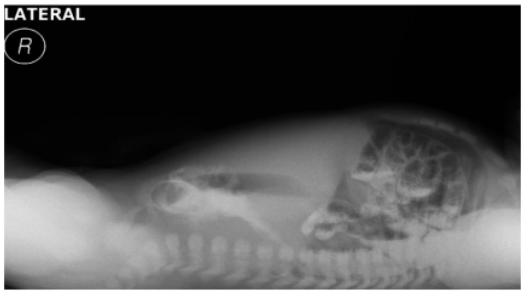


Fig.3, Fig.4: Six hours after Figure 2

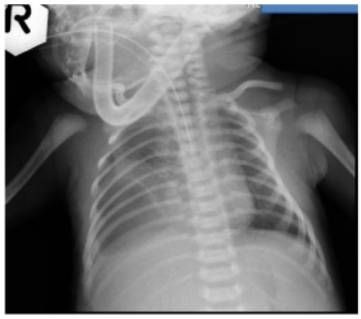


Fig.5: After the surgery at two day old

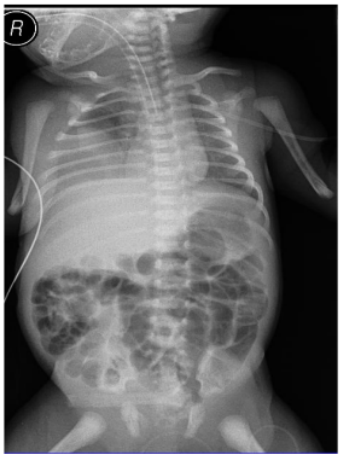


Fig.6: At five-days old

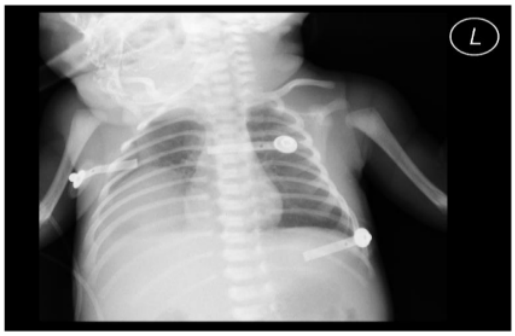
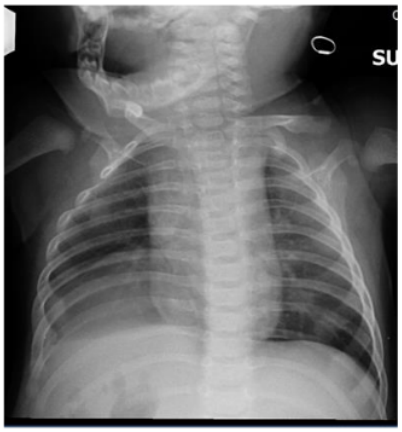


Fig.7: At seven-days-old. Fifth post operation day.



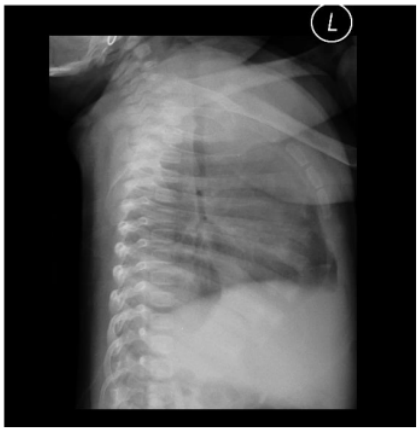


Fig.8, Fig.9: Three months after the discharge from the hospital.

DISCUSSION

The diaphragm begins to develop at approximately 4 weeks of gestation and is fully formed by 12 weeks. It is derived from the septum transversum, the pleuroperitoneal folds, derivatives from the body wall, the dorsal mesentery, and a pair of pre-muscle masses lying opposite the fourth cervical segment of the 9 mm embryo. The septum transversum originates around day 22 at a cervical level, but caudal to the developing heart. In the normal conditions, the pleuroperitoneal folds fuse with the septum transversum, the esophageal mesentery and the muscular ingrowth from the body wall invade the folds, forming the muscular part of the diaphragm.(1,2) The embryologic basis of congenital diaphragmatic hernia remains controversial. It was thought initially that the defect happened secondary to failure of different parts of the diaphragm to fuse resulting in a patent pleuroperitoneal canal. This in turn, allows the gut to enter the thoracic cavity when it returns from the extraembryonic coelom of the umbilicus. Another opinion is that lung hypoplasia may be the primary causal factor in the pathophysiology of diaphragmatic hernia.(2)

Both environmental and genetic factors are thought to contribute to the etiology of CDH. To date, genetic causes have been identified in 30% of cases. The most frequent CDH-associated aneuploidies include trisomy 19, trisomy 13, trisomy 21, and less frequently trisomy 9, trisomy 16, trisomy 22, mosaic trisomy 2, Turner syndrome, and trisomy X.(3) In this case the patient has no physical anomalies and no genetic screening was done.

Approximately 50% of cases are diagnosed antenatally as a result of routine ultrasound screening. Sensitivity of screening improves with advancing gestation, the presence of associated abnormalities, and with an experienced sonologist. Once a CDH has been identified, high resolution ultrasound should be used to grade severity or in order to aid parental counselling. This usually includes measurements of lung volume and the position of the liver as well as looking for significant comorbidity.(4) Most newborn infant present with severe respiratory compromise in the neonatal period. Late presentation is rare and variable leading to misleading radiologic assessment, delay in treatment, and fatal consequences.(5) In this case CDH is not detected during antenatal period.

Different imaging modalities can be useful in identification of thoracic hernias, considering their advantages and disadvantages including radiographs, upper gastrointestinal series, ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI). The roles of imaging in the evaluation of hernias include establishing the diagnosis, differentiating the type, delineating the extent, identifying the contents, detecting complications and providing a roadmap for intervention/surgery.(6) A chest radiograph is often the first imaging study performed in children with CDH, particularly those with respiratory symptoms, however in patients with CDH who present with gastrointestinal symptoms, abdominal radiographs may be obtained as a first study. The diagnosis of CDH is clear when gas-filled loops of bowel are seen in chest, with or without associated mediastinal shift. Radiologically CDH

can mimic congenital lung cyst, infective lung diseases, and pneumothoraces.(5,7) In this case, we found CDH by plain radiograph and upper gastrointestinal series.

A literature review conducted by Baglaj et al suggests that an upper gastrointestinal series is the best modality for diagnosis of CDH presenting postnatally. A CDH may contain only stomach, only small bowel, or only large bowel. As such if the index of suspicion is high, a complete small bowel follow-through and/or contrast enema should be performed if the stomach is found to be within the abdomen on the upper gastrointestinal study. In a very small percentage of children with CDH, the hernia may contain only solid organs.(7)

Newborns with CDH who have immediate respiratory distress should be preferentially intubated at birth; bag and valve ventilation should be avoided. The goal is to achieve preductal saturation levels of 80-95%. FiO2 is started at 1.0 and then adjusted downwards to achieve targets. Immediate placement of oro or nasogastric tube with continuous or intermittent suction will help to decompress the bowel and facilitate lung expansion. A central or peripheral venous line should be inserted. Arterial blood pressure to be maintained at acceptable levels for gestational age.(8,9,10,11,13,14,15)

The standard surgical approach to repair the diaphragmatic defect consists of the subcostal incision with removal of the abdominal contents from the thorax and complete closure of the defect. For defects that are too large and to be closed by primary repair, a number of reconstructive techniques such as prosthetic patches have been evolved to close the gap, and this can be accomplished by synthetic non-absorbable material or by natural absorbable patch. The use of minimally invasive surgery techniques for repair of CDH is gaining popularity; however they have a significantly higher recurrence risk of herniation.(8,12,13,16,17,18,19)

The postnatal diagnosis and management of Congenital Diaphragmatic Hernia remains challenging. It is very important to detect the anomaly during antenatal period for better preparation and prognosis. Congenital Diaphragmatic Hernia requires careful examination followed by multiple imaging techniques, detecting complications, then providing roadmap for surgical intervention.

CONCLUSION

Deaths of infants with congenital diaphragmatic hernias remain high, despite optimal perinatal care. It is important to detect anomalies during the antenatal period for better preparation and prognosis. Successful postnatal diagnosis and treatment of female patients with congenital diaphragmatic hernias by means of intubation and surgical repair show better outcomes

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