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Case Report

Congenital diaphragmatic hernia in neonate with migrating spleen

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ABSTRACT

A case of full-term neonate with congenital diaphragmatic hernia (CDH) is a potentially severe anomaly wherein a diaphragmatic defect allows intrathoracic herniation of intra-abdominal contents. CDH is usually accompanied by pulmonary hypertension which worsens the prognosis. In our case, the newborn doesn't develop pulmonary hypertension and timely surgical intervention and accurate post-operative care gave the expected outcomes.

Keywords: CDH, Migrating spleen, Open repair

INTRODUCTION

CDH is a congenital defect of the diaphragm, where all or part of the diaphragm fails to form, that permits herniation of abdominal viscera into the thoracic cavity.[1,2] Its incidence is 1/3500 live births with a male predominance.[2] Postero-lateral hernias, also known as 'Bochdalek' hernias, account for 70-75% of all CDH cases. In Bochdalek hernias, 85% are left sided, 13% are right sided and 2% are bilateral. Anterior hernias (Morgagni hernias) and central hernias are relatively rare.[4] CDH patients often have pulmonary hypoplasia and abnormal bronchial and vascular branching patterns resulting from prenatal compression of the lung and abnormalities in lung development. There is a controversy over the survival rate of CDH, but it seems to have improved from 50% to 80% over past 3 decades owing to advances in neonatal care.[1]

CASE REPORT

A full-term female newborn, born through vaginal delivery with no maternal comorbidities, who became dyspneac and cyanosed with air entry reduced on the left side. Chest X-ray reveals bowel loops present in the thoracic cavity with collapsed left sided lung and mediastinal shift (Figure 1). A detailed evaluation of the newborn was done by neonatologists, surgeons, and anesthetists, and found the newborn fit for the surgery with no pulmonary hypertension. The newborn underwent an open repair in which small bowel, transverse colon, and spleen were found in the thoracic cavity. Herniated structures were reduced into the abdominal cavity and diaphragmatic defect closed with prolene and closure was done. During initial post operative course patient was monitored for abdominal compartment syndrome and respiratory distress in neonatal ICU. The post-operative course was uneventful and the patient was discharged after suture removal on 12th post-operative day (Figure 2).

Figure 1: Pre-operative chest X-ray showing bowel loops within the thoracic cavity with collapsed lungs.

Figure 2: Post-operative chest X-ray showing normal lungs with no bowel loops within thoracic cavity.

DISCUSSION

CDH is a malformation that results from a defect of the muscular or the tendinous portion of the diaphragm leading the abdominal viscera to herniate into the thoracic cavity and compete for space with the developing lungs. Clinically, CDH occurs either as an isolated malformation in approximately half of cases/in association with other malformations. Cardiovascular defects by far most common associated malformations. Less frequently, CDH is associated with musculoskeletal, urogenital/ brain malformations and is rarely associated with ocular defects that affect about 1% of cases.[3]

One of the largest advances in the care of CDH patients in the modern era comes from the advent of improved prenatal imaging and access to that imaging. CDH, having abdominal contents herniating into the thorax, is often easily detected by ultrasound. Many fetal centers proceed to fetal MRI and fetal echocardiography after the detection of a potential diaphragm defect on ultrasound.[5] CDH is incredibly challenging to treat because of the variety of anomalies within the chest. There is the obvious diaphragmatic defect that needs to be repaired for proper pulmonary mechanics and gastrointestinal function but more importantly, there is physiologic dysfunction of the pulmonary parenchyma that causes severe morbidity in some patients.[5] Neonates with CDH often suffer from severe respiratory failure. Therefore, respiratory supports such as the use of exogenous surfactant, inhaled nitric oxide (NO), high frequency oscillatory ventilation (HFOV), and extracorporeal membrane oxygenation (ECMO) may increase the survival.[4] Pulmonary hypertension is now recognized as the major cause of morbidity and mortality in newborns with CDH.[5] From treating CDH as an operative emergency to stabilizing the patient and treating pulmonary hypertension and delaying surgery for optimization is the key to successful management.[5] The management of CDH has seen steady progress over the last 20 years. Although the accuracy of these outcome predictions remains a matter of debate, it seems important that all teams in charge of those fetuses use the same prognostic factors to be able to improve and compare their practice.[7] Today, overall survival has reached 80% in live-born infants as a direct result of changes in medical and surgical management. Preoperative physiologic stabilization and subsequent elective repair have become the cornerstones of management.[6]

CONCLUSION

CDH diagnosis and antenatal management essential to reduce patient mortality and morbidity. Prognosis is poor and depends on presence and degree of pulmonary hypoplasia and association with congenital malformations. The prenatal evaluation of the postnatal prognosis of fetuses displaying CDH has improved over the past five years. It is important to have a multidisciplinary approach with a team of experienced maternal-fetal medicine experts, neonatologists, surgeons, and anesthetists working in concert to get the best outcomes.

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