Original Research Paper

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ANTENATAL DIAGNOSIS OF BOCHDALEK HERNIA

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ABSTRACT Bochdalek hernia, the most common fetal congenital diaphragmatic hernia, presents most commonly on the left posterolateral side of diaphragm and is associated with a poor outcome. We report an antenatal case of bochdalek hernia in a 21 year old G1P0 female at 22w2d of gestation.On ultrasonography, the fetal chest and abdomen sections showed the presence of fetal stomach in thorax, adjacent and posterior to fetal heart. The objective of this report is to create awareness about the presenting ultrasonographic picture of bochdalek hernia, discuss its existing prognostic factors(O/E LHR- observed/expected lung area to head circumference ratio and liver herniation being the most important ones) and therapeutic interventions(Fetoscopic Endotracheal Occlusion being the only clinically applied intervention) which can henceforth help formulate an adequate management plan and hence improve the overall prognosis.

KEYWORDS :

INTRODUCTION

An absent or partially formed diaphragm results in an abnormal opening (hernia) that allows the stomach and intestines to move into the chest cavity and crowd the heart and lungs. This crowding can lead to underdevelopment of the lungs(pulmonary hypoplasia), potentially resulting in life threatening difficulties that are apparent from birth.[1] Congenital diaphragmatic hernia(CDH) occurs in 1 of every 2000-3000 live births and accounts for 8% of all the major congenital anomalies.[2]Bochdalek hernia is the most common fetal CDH, which is present more commonly on the left posterolateral side and is associated with a poor outcome.[3] Prenatal diagnosis of diaphragmatic hernia makes it possible to define the natural history of this lesion, determine the pathophysiologic features that affect clinical outcome and formulate management based upon prognosis.[4]We report an antenatal case of Bochdalek hernia in a 21 year old female at 22w2d of gestation.

CASE REPORT

A 21 year old G_1P_0 female presented to us at the gestational age 22w2d for a routine antenatal Level II ultrasound scan. The patient was relatively healthy and had no complaints. On ultrasonography, the fetal chest and abdomen sections showed the presence of fetal stomach (anechoic) in the left posterolateral part of thorax adjacent to fetal heart. Figure 1, Figure 2 and Figure 3 are the axial, coronal and sagittal ultrasonographic sections of the fetal thorax respectively showing the stomach marked with a white arrow. The fetal stomach appears over-distended. These findings suggest the likely possibility of a diaphragmatic hernia in the postero-lateral region (commonly called Bochdalek hernia). Fetal situs is normal and the liver is not seen herniated in the thorax. Other fetal parameters including fetal brain structures (cistern magna, cavum septum pellucidum), trigone, anterior abdominal wall, kidneys, urinary bladder, lung, bowel and extremities all appeared normal with their echogenecities within normal limit. The patient was counselled regarding the disease, its complications and management, and was advised follow-up every 2 weeks in order to devise an appropriate post-natal management plan based on the subsequent measurement of LHR of fetus.

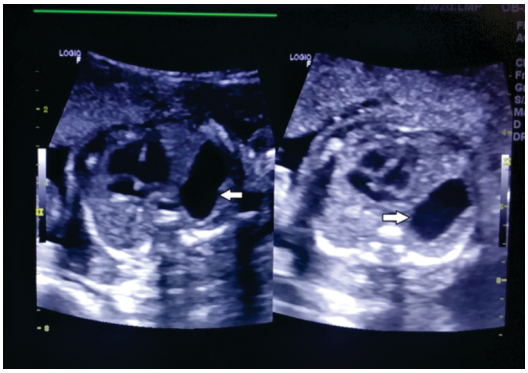

Figure 1

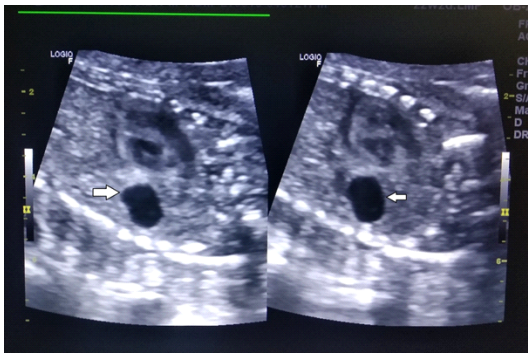

Figure 2

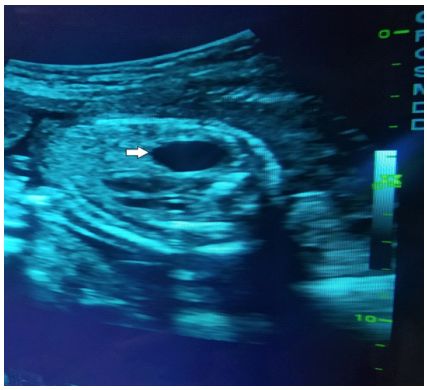

Figure 3

DISCUSSION

CDH does not designate one single clinical entity and outcomes are diverse. 84% of lesions are left sided, 13% right sided and 2 % bilateral. While the name essentially points to a defect in the diaphragm, it is the abnormal lung development that accompanies the condition that gives it its clinical relevance.[5]

Left sided CDH is most common and typically characterised by shift of the heart and mediastinum to the right, caused by herniation of the stomach and intestines. The stomach is easily recognised because of its fluid contents.[5]

The morphologic changes become obvious only when the lung becomes functional at birth. These lead to variable degree of respiratory insufficiency and pulmonary arterial hypertension (PAH). PAH is increasingly treated by early administration of inhaled nitric oxide (iNO).[5]

The best validated prognostic indicator is the lung area to head circumference ratio. Ultrasound is used to measure the lung area of the index case, which is then expressed as a proportion of what is expected normally (observed/expected LHR). When O/E LHR < 25%, survival chances are 15%.[5]

Liver herniation has been long recognised as a prognostic indicator. Other less used prognostic indicators are position of the stomach, fetal lung vascularisation and its potential for vasorelaxation.[6]

As CDH is a developmental problem, the ideal therapeutic window of opportunity is the prenatal period. Today the only clinically applied intervention is Fetal Endoluminal Tracheal Occlusion (FETO). TO prevents egress of lung fluid, which in turn causes increased pulmonary stretch, hence accelerated lung growth.[6]

In severe cases, the FETO task force initially proposed insertion of balloon at 26-28 weeks and for moderate cases at 30-32 weeks. Reversal of occlusion is proposed at 34 weeks[6]

For babies born with a known CDH there is significant debate about delivery time and mode, the use of extracorporeal membrane oxygenation (ECMO) and post-surgical treatment. In cases with mild to moderate CDH detected prenatally, the current treatment is aggressive respiratory support including mechanical ventilation, medical stabilization and surgical closure of the defect.[7]

The main differential diagnosis are other pulmonary pathologies, such as cystic pathology (cystic adenomatoid malformation, bronchogenic, enteric and neurenteric cysts, mediastinal teratoma and thymic cysts) or bronchopulmonary sequestration as bronchial atresia.[5]

CONCLUSION

Signs suggesting a poor prognosis in CDH include large hernia size, early gestational age at diagnosis, intrathoracic liver, small contralateral lung, the presence of associated abnormalities, bilateral CDH and unfavourable L: H Ratio.[3]

The early antenatal detection is very important, as it is the key to help in the obstetric management as well as newborn care. Antenatal diagnosis allows prenatal management (e.g., FETO) that may be indicated in cases with severe lung hypoplasia. It is therefore critical that intensive training be offered to all sonographers and training doctors, in order to maximise the detection rates of congenital diaphragmatic hernia and provide patients with the best care possible. It is also crucial to have a comprehensive evaluation, including imaging and (high-resolution) genetic studies. The primary purpose of this evaluation is to rule out associated anomalies and to assess the severity of pulmonary hypoplasia in order to offer parents an individualized prognosis. The latter can be done on the basis of the dimensions of the lung, its vascularisation and liver position. Based on this complete evaluation and after extensive counselling, parents should make an informed choice out of the available management options, which includes expectant management with prenatal referral to a high volume center for carefully timed delivery, termination of pregnancy, or fetal intervention for selected patients .[7]

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