Fetal intrapericardial Morgagni hernia presenting with hydrops fetalis: A case managed by thoracoamniotic shunting and fetal endoscopic tracheal occlusion (FETO) and review of the literature

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Introduction

A rare type of congenital diaphragmatic hernia is the Morgagni hernia (about 2-5%), which is characterized by an anterior mainly right-sided defect of the diaphragm, infrequently combined with a herniation of the liver into the pericardial cavity, resulting in pericardial effusion and subsequently lung hypoplasia. We report a case of Morgagni hernia with pericardio-diaphragmatic aplasia, complicated by massive pericardial effusion and fetal hydrops. The case was successfully managed in utero with thoraco-amniotic shunting and a late tracheal occlusion, followed by corrective surgery after birth.

Case report

Referal at 23 weeks’ gestation for detailed ultrasound evaluation because of hydrothorax. Ultrasound revealed a fetus with concordant growth, mild ascites and polhydramnios and what was thought, at that time, severe bilateral pleural effusions. Both lungs were atypically compressed into an extremely posterior position (a,b). There was no evidence of further fetal abnormalities and the karyotype (46,XY) was normal. Due to suspected pulmonary hypoplasia and pronounced hydrops, thoraco-amniotic shunting (Somatex Intra-Uterine-Shunt) was performed and resulted in the complete drainage of the effusions. Follow-up scans demonstrated a progressive, right-sided protrusion of the liver into the thorax (c) with a mediastinal shift to the left and extremely hypoplastic lungs. The initial diagnosis was revised and the presumptive diagnosis of an anterior diaphragmatic hernia (Morgagni) with pericardio-diaphragmatic aplasia was made. This was also supported by fetal MRI at 32 weeks’ (d). Due to persistant hypoplastic lungs, a late tracheal occlusion was performed at 32 weeks.

Delivery, Surgery and Discharge

The child was delivered at 38 weeks’ by cesarean section and was admitted to the intensive care unit for intubation and mechanical ventilation. Corrective surgery was done at day 6, with relocation of the hepatic lobes into the abdominal cavity and closure of the anterior diaphragmatic hernia. Reconstruction of the pericardium was achieved with the small rim of the diaphragmatic muscle and the doubled hernial sac. Discharge was after 3 months without any breathing support.

Discussion

We present a rare form of Morgagni hernia with intrapericardial liver herniation, severe pericardial effusion and subsequent fetal hydrops. Assessment of fetal lung position in the thorax is essential in order to distinguish pericardial from pleural effusion. Differential diagnoses of pericardial effusion should include diaphragmatic evagination, cardiac diverticulum or pericardial teratoma.

Prenatal intervention with firstly thoraco-amniotic shunting, followed by late tracheal occlusion was considered to possibly improve the perinatal outcome. Review of the literature revealed 13 other cases of Morgagni hernia diagnosed during fetal life. Only 4/13 cases presented additional structural abnormalities. Postnatally, respiratory distress was common and corrective surgery was always needed. However, the reported outcome after corrective surgery showed to be excellent.