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Case Report

Hepatopulmonary Fusion in a Newborn Diagnosed during Surgical Repair of Right Congenital Diaphragmatic Hernia

Rechid khemakhem[1]*, Mohamed Raiz elhaq Rao[1], Refeat Mohamed Sadek[1], Adel Metwali Fadhl[1], Majdi Gueessa[2], Mohamed yousef[2] and Houda Rahay[2]

[1]*Department of Pediatric Surgery, King Faycal Medical Complex, Taif, KSA*
[2]*Department of Neonatology, Pediatric Hospital, Taif, KSA*

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***Corresponding author**

Rechid Khemakhem, Department of pediatric surgery, King Faycal medical complex, Taif, KSA, Tel: +966549514087, Email: kakoum_rochdi@yahoo.fr

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Abstract

Hepatic Pulmonary Fusion (HPF) is a rare malformation in which adhesion between the liver and the right lower lobe of the lung occurs. This situation is frequently associated with right Congenital Diaphragmatic Hernia (CDH), and often only discovered during surgical repair of the defect. Few cases have previously been reported in the literature [1].

In this paper we are reporting a case of HPF diagnosed fortunately in a tow days old female baby, at time of surgical repair of right side CDH and we discuss the modalities of diagnosis and the difficulties encountered during surgery.

Introduction

Congenital Diaphragmatic Hernia (CDH) is a rare malformation that consists on a defect in the development of part of the diaphragm with herniation of abdominal content in the thoracic cavity. It is usually responsible of different degree of lung hypoplasia due to compression of the developing lung during fetal period. Right side CDH represents only 10 to 15% of cases and it is usually limited to a lobe of the liver that herniate into the thoracic cavity [1].

Hepatic Pulmonary Fusion (HPF) is a rare congenital anomaly in which different degree of adhesion between the liver and the right lower lobe of the lung occurs. This varies from some fibro-vascular communications to complete parenchymal fusion between lungs, liver and diaphragmatic sac with abnormal systemic venous drainage and arterial supply [2].

Babies with Hepatic pulmonary fusion and CDH are subject of associated malformations which constitute an important cause of mortality; most important of them are cardiac malformations. Mortality in CDH depends also on the severity of the pulmonary hypoplasia and pulmonary hypertension which is caused by a decrease in cross-sectional area of pulmonary vasculature and dysfunction of the surfactant system that will jeopardize gaseous exchange [3]. HPF is usually discovered in occasion of surgical repair of right CDH and attempt to separate this different structures can have dramatic consequences.

Case Report

We describe a case of a full term female newborn, born by normal vaginal delivery, how presents soon after birth respiratory distress with tachypnea, cyanosis and intercostal retraction. Chest auscultation show diminution of breath sounds in the right hemi-thorax without added sounds. Patient was intubated, sedated and mechanically ventilated; A naso-gastric tube was placed for aspiration. Chest x ray shows a part of the liver occupying the lower half of the right hemi-thoracic. Patient was than keep NPO, on mechanical ventilation with 100% Fio2, inotropes and deep sedation. Cardiac echography shows moderate pulmonary hypertension. After a stabilization of 48 hours, patient was operated. Laparotomy confirms the diagnostic of right CDH with large defect and herniation of the liver. Reduction of herniated lobe was difficult because of strong adhesion between the herniated liver and right lower lobe of the lung. We try to progressively divide this adhesion with diathermia but it was difficult and too hemorrhagic. Surgicell was applied than defect partially closed around this strong adhesion with double face mesh. A sub diaphragmatic drain was placed and incision closed in layers.

In post operatory, patient develop bleeding from the drain with decrease of hemoglobin and was transfused with PRBC and frozen plasma. She deteriorates his respiration because of increase in pulmonary hypertension and didn't respond to aggressive resuscitation.

Discussion

Hepatic pulmonary fusion is a rare congenital anomaly in which different degree of adhesion between the liver and the right lower lobe of the lung occurs. This varies from some fibro-vascular communications to complete parenchymal fusion between lungs, liver and diaphragmatic sac with abnormal systemic venous drainage and arterial supply [2].

The lung on the fused side is usually hypoplastic and systemic arterial and venous vessels will often be seen supplying and draining the fused lung tissue. Because of this observation, some authors have suggested that hepatic pulmonary fusion may have a relationship to pulmonary sequestration [4].

Hepatic pulmonary fusion is usually associated with right sided CDH [1]. Development of hepatic pulmonary fusion in-utero is not well understood but these two malformations may be caused by similar development abnormalities. In fact, failure of formation of the embryonic diaphragm allows the liver to herniate into the thorax during the 10th week of gestation. Later in development, the liver may then fuse to the primitive lung. Conversely, hepatic pulmonary fusion may be a primary anomaly which then "prevents" the diaphragm from forming properly [4].

The diagnosis of CDH can be made prenatally by Ultrasonography in 90% of cases. Typically this is diagnosed at the 24th week of gestation, but some have reported diagnosing it as soon as 11th week. Fetal ultrasound finding include polyhydramnios, intra-thoracic stomach or intestinal loops. Liver herniation might be associated with poorer prognosis when detected by antenatal ultrasound [5].

With increased antenatal diagnostic and use of fetal MRI for CDH, hepatic pulmonary fusion may be identified prior to delivery. When a CDH is discovered on a prenatal ultrasound study, it is now more common that a fetal MRI is also performed. Perhaps increased use of this modality will be able to identify instances of hepatic pulmonary fusion prior to postnatal operative exploration. The finding of hepatic veins draining directly into the right atrium as seen on echocardiography may also be diagnostic of hepatic pulmonary fusion [4]. In this circumstance, delivery must be planned to be in tertiary health center with available neonatal ICU and expert pediatric surgery team.

In most instances CDH is diagnosed in newborns that have respiratory symptoms. Sometimes it can be discovered beyond the neonatal period when the patient presents with delayed onset of symptoms [2]. In this situation, the diagnosis of HPF is difficult to make using imaging alone and many cases are encountered at the time of surgery. This diagnosis should be suspected in case of right side CDH without a contralateral mediastinal shift. In this case, CT chest or better MRI can confirm the diagnostic [4].

Repair of CDH can be done by sub-costal incision or even by thoracotomy. Surgical correction may be challenge due to the relative small size of the abdominal cavity and the difficulty to reduce all organs in to abdominal cavity without pressure. At operation, the posterior rim of the diaphragm is first separated from the overlying peritoneum and then a two-layer closure of the canal is done. Synthetic patch may be needed where the diaphragm is insufficient [4].

In case of HPF, operative exploration with intent to separate these organs may result in bleeding and injury to pulmonary parenchyma. Reduction of bowel and creation of 2 separate cavities is the primary surgical goal when separation of the fused tissues cannot be achieved without undue risk [2,4]. The surgeon should be aware that division of hepatic pulmonary fusion to achieve complete separation of the thoracic and abdominal cavity adds risk to the procedure and may not be necessary. Reduction of bowel and approximation of the diaphragmatic defect around the fused liver and lung was successfully employed in the present case. However, long term follow up is necessary to be sure that this technique is enduring [2,4].

Conclusion

HPF is a rare condition frequently associated with right side congenital diaphragmatic hernia. This malformation is difficult to diagnose with imaging alone and must be evocated in case of non-shifting of the mediastinum to the opposite side on chest x-ray. In this situation chest CT scan or better MRI is obtained to confirm this diagnostic.

Treatment is surgical and consists of reduction of the herniated liver, release of adhesions and closure of the diaphragmatic defect. Partial closure of the defect around the fused liver and lung is an option that must be considered in some cases. Attempt to completely release this adhesions can be very dangerous.

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