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Case Report

Congenital Diaphragmatic Hernia: An Unusual Cause of Obstructive Jaundice

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Abstract

Congenital diaphragmatic hernia is exceptional in children. It is defined by the presence of an abnormal diaphragmatic defect causing the abdominal cavity and the thoracic cavity to communicate. It usually manifests itself at birth through respiratory distress, or sometimes through sepsis requiring urgent treatment. We report the rare case a girl with cholestatic jaundice secondary to a left antero diaphragmatic hernia with visceral content (hail and colon).

Keywords: Congenital diaphragmatic hernia; Obstructive jaundice; Bochdalek hernia.

INTRODUCTION

Congenital diaphragmatic hernia (CDH) has been defined as a congenital malformation of the diaphragm resulting from the persistence of the pleuroperitoneal canal owing to the non fusion of the pleuroperitoneal folds during the eighth week of gestation; it is a birth defect of the diaphragm which allows the abdominal organs to push into the chest cavity, hindering proper lung formation [1, 2]. The CDH is almost prounouced an emergency as a severe respiratory distress due to both pulmonary hypertension and pulmonary hypoplasia and sepsis [3]. We present the case of a patient with a left antero intern congenital diaphragmatic hernia associated to a cholestatic jaundice. In our knowledge there are just four cases reported before in the litterature and all of them were in the right sided.

CASE HISTORY

A 2.5 year old child, with little previous past history such as a mother diabetic end an un ombilicus hernia surgy five months ago. The patient is presented to hospital with 3 weeks history of obstructive jaundice and abdominal pain also vomiting episods. The patient was admitted under the medical team and the clinical exam has founded an obstructive jaundice as more as an off center umbilicus with an abdominal distention and livermegaly (Figure-1). Workup including liver function tests was remarkable for total bilirubin of 39 mg/dL, direct bilirubin 26,6 mg/dL, alkaline phosphatase 1679 IU/L, GGT 2110 IU/L, LDH 388 IU/L, ASAT 164 U/L, ALAT 132 U/L. An Ultrasonography of the abdomen performed demonstrated the principale bile duct dilatation and homogeneous livermegaly. We completed the assessment by an abdominal TDM so describing a diaphragmatic hernia antéro intern left sided with visceral contents (hail and colonist) responsible of a dilatation of main biliary duct (Figure-2). The patient has been operated with reduction of defects, repairation of diaphragmatic defect also the arrangement of hail as common completed mesenter ,appendicectomy and liver biopsy; the anatomopathology study of the piece is in favour of hepatic fibrosis mutilate and nodular cirrhotic A3F4. The obstructive jaundice clinical and biological has ben spectaculary resolved within a week of Post surgery.

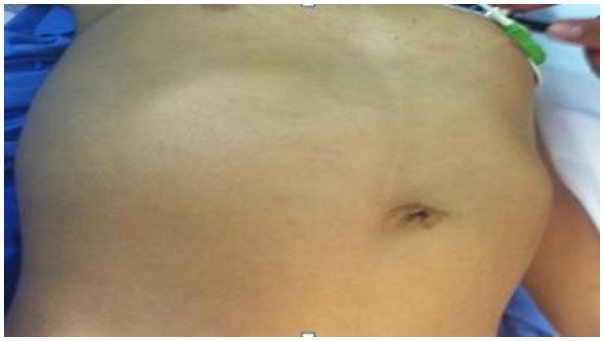

Fig-1: Image showing abdominal distension with an omblic desaxed on the left

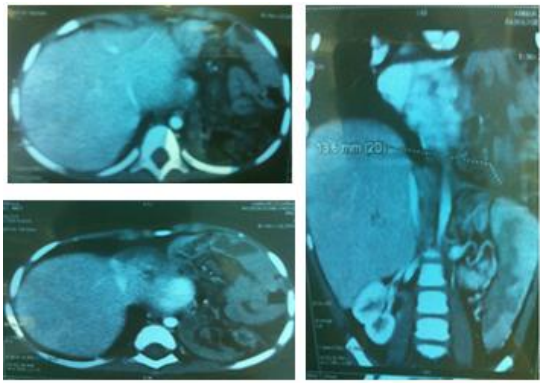

Fig-2: Abdominal CT showing a left anterior internal diaphragmatic hernia with visceral content (hail and colon) with dilation of the main bile duct

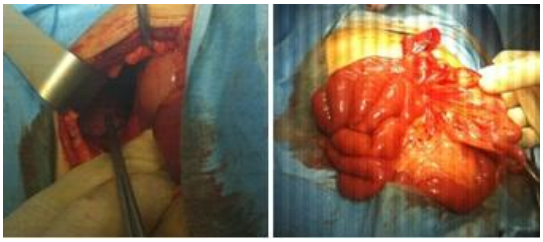

Fig-3: A left antero diaphragmatic hernia with visceral content (hail and colon)

DISCUSSION

Congenital diaphragmatic hernia (CDH) is very rare with an incidence of between 1 in 2,500 to 12,000 live births [1]. The most common type of CDH is a Bochdalek hernia also known as a postero-lateral diaphragmatic hernia occurring in the vast majority present on the left side of diaphragm, other types include Morgagni hernia, diaphragm eventration and central tendon defects of the diaphragm [2, 4]. Prenatal diagnosis is typically made on ultra sound examination [5]. In the vast majority of cases, this condition presents with severe respiratory distress in the immediate neonatal period. A minority of patients, however, present after the first 24–72 h of life; in these instances the symptoms tend to be chronic with gastrointestinal and pulmonary manifestations [6]. As a result of the compression we can have bowel obstruction, gastric reflux and pancreatitis also pulmonar hypoplasia of the ipsilateral side of the hernia, which should be treated immediately by endothracheal intubation [3].

Obstructive jaundice secondary to CDH is a rare presenting symptom that has been exceptionally reported in the literature for diaphragmatic hernias. Our article include the case of a child girl with the association of congenital left-sided Bochdalek hernia caused by the stretch of the bilary duct. In literature we have found just fewer than 100 cases of patients (superor than 19 years old) surfing from respiratory symptom due to diaphragmatic hernia without any obstructive jaundice. Although, four cases of neonates with a Bochdalek hernia have been presented with obstructive jaundice. Obstruction of the extrahepatic biliary system by the herniated content has ben termed ‘‘choledochal semivolvulus’’ by Caldeiro *et al*., [7]. The first cases was reported in 1988 on the archives of the pediatrie , one year after it was published on the surgery pediatric the same cases. On 2001 GARCIA MUNOZ reported the case of a new born wth early sepsis and obstructive jaundice associated with right sided diaphragmatic hernia. In 2009 A. BEKMEZIAN illustrate the cases of a six months baby who suffer from an obtructive jaundice due to a diaphragmatic hernia; Later in 2011 ROY M tolk about morganic hernia presenting as obstructive jaundice.

CONCLUSION

Diaphragmatic hernia is exceptionally rare in children as well as in adults it can be manifested by respiratory signs but must we have in the head in front of cholestatic jaundice associated with a fluctuating digestive signs as described in our observation.

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