



Right congenital pleuro-peritoneal hiatus hernia

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Devi SANKAR K ⁺
 Sharmila BHANU P
 Susan PJ
 Gajendra K

Department of Anatomy, Narayana Medical College, Chinthareddy Palem, Nellore, Andhra Pradesh, INDIA.



⁺ Devi Sankar K, MSc
 Assistant Professor
 Department of Anatomy
 Narayana Medical College
 Chinthareddy Palem, Nellore
 Andhra Pradesh, 524 002, INDIA.
 ☎ +91 949 0948006
 ✉ lesanshar@gmail.com

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ABSTRACT

Congenital diaphragmatic hernias are of various types which are due to the defect in the diaphragm and can be encountered in any period of life. Left mediastinal shift with right congenital diaphragmatic hernia is rare and life threatening malformation. We describe a case of right congenital diaphragmatic hernia of a newborn male infant, which died shortly after birth. The lobes of the liver were enlarged and occupied whole of the abdominal cavity. The stomach and intestinal loops were herniated into the right pleural cavity and partly into the left pleural cavity. Severe hypoplastic right lung, trilobed left lung and dextrocardia also were observed. © IJAV. 2009; 2: 153–155.

Key words [diaphragmatic hernia] [unilateral congenital diaphragmatic hernia] [mediastinal shift] [dextrocardia] [pulmonary hypoplasia]

Introduction

Congenital diaphragmatic hernia (CDH) is the herniation of abdominal contents into the thoracic cavity due to the defect in the diaphragm. CDH occurs 1 in every 2000–4000 live births and accounts for 8% of all major congenital anomalies [1]. CDH is a life-threatening malformation believed to be the consequence of failure of closure of the pleuroperitoneal canal which is usually unilateral most frequently seen on left side. Very rarely diaphragmatic hernia occurs bilaterally [2]. We report a case of unilateral congenital right diaphragmatic hernia (CRDH) with left mediastinal shift. The fetus with CRDH was in no danger until birth but died shortly after birth due to respiratory distress [3].

In human, CRDH are approximately five times less common than those on the left. Defects of the right diaphragm, although surgically and anatomically similar to those on the left, are more inconspicuous in presentation. Because the symptoms of the former are less severe, however, they tend to be diagnosed later. Some are asymptomatic and go unrecognized through life [4].

Case Report

A 36-year-old pregnant woman from a nearby village was admitted in maternity ward of Narayana General Hospital, Nellore. The prenatal ultrasonographic findings at 36th week of gestation showed CRDH. A single male infant weighing 2,572 grams was delivered vaginally at

38 weeks of gestation after spontaneous labor. The infant developed severe respiratory distress immediately after birth and died 2 hour of age despite intensive resuscitation. With the parent's written consent, the dead infant was sent to anatomy department museum, Narayana Medical College, Nellore. The infant was injected and immersed in routine formalin and allowed for fixation. The infant's chest and abdominal wall was incised and opened carefully. Chest cavity showed mediastinal shift towards left side.

A large defect was found in the right side of the diaphragm, confirming the CRDH. All parts of the gastrointestinal system including a part of the right lobe of liver were herniated first into the right side and then left pleural cavities behind the pericardium. Only a small portion of the gastrointestinal system (most of the liver, right sided descending and sigmoid colon) and urinary organs were seen in the abdominal cavity. There was a complete absence of spleen. The right pleural cavity showed severe hypoplasia of right lung. In addition, the thoracic cavity showed trilobed left lung and dextrocardia (Figures 1, 2).

Discussion

Congenital diaphragmatic hernias seen in the fetal and newborn are due to large defect or complete absence of the diaphragm and have many problems in diagnosis and treatment [5]. The defects are usually unilateral and 75% of cases were found to be left CDH. Bilateral hernias are rare and found in only 2% of cases [6]. Although the CDH

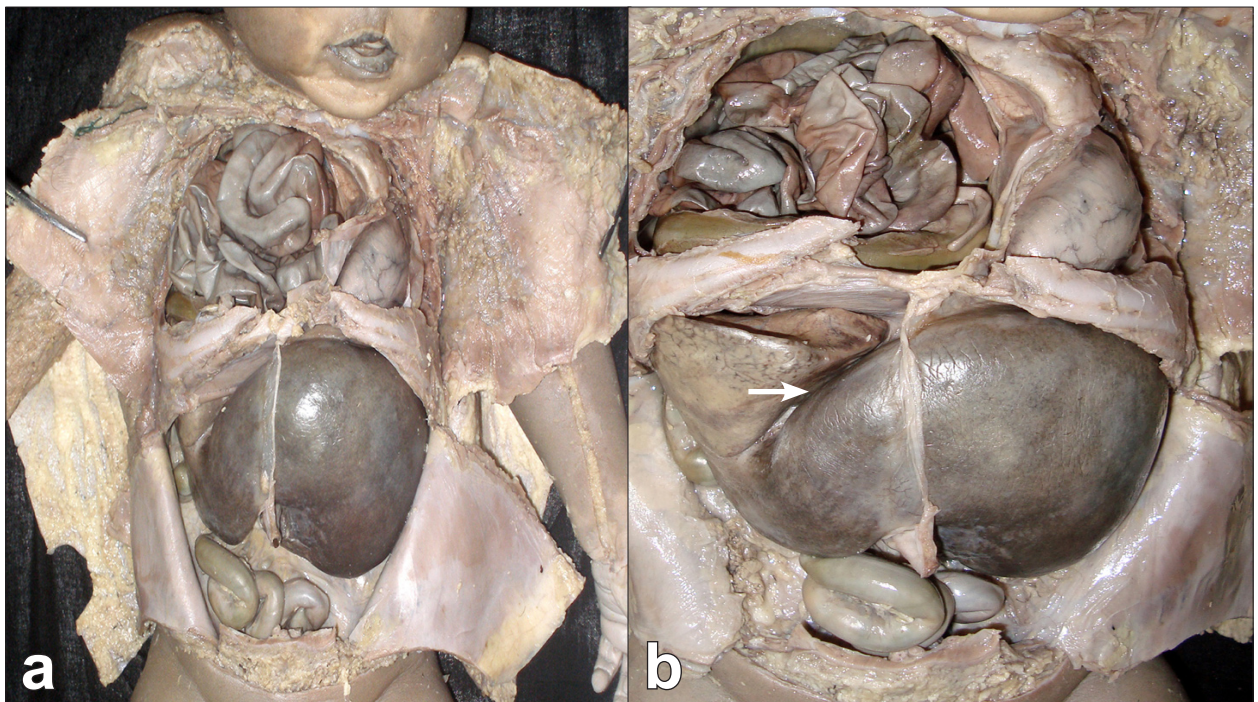


Figure 1. a) Dissection of thoracic and abdominal cavity showing the right congenital diaphragmatic hernia with mediastinal shift. Herniation of part of the liver into the right thoracic cavity through the hiatus with right sided descending and sigmoid colon can be seen in abdomen; b) Herniated part of the liver is pulled out of the hiatus to show the *groove* (arrow) formed by strangulation of the diaphragm.

usually presents during neonatal period, 13% of cases may be found in the later periods, the approximate age being 16-18 years [7].

The pleuroperitoneal canals close during 10 week of intrauterine life which allow fetal diaphragm to separate the thoracic and abdominal cavities. Physiologically herniated fetal gut has to return into the abdominal cavity by 10-12 weeks which when the diaphragm is not closed, moves into the chest cavity resulting in CDH frequently seen on the left side. [8].

This type of hernias occur during the development due to the lack of fusion of pleuroperitoneal membrane with septum transversum which results in the congenital communication between the peritoneal and thoracic cavities, often seen on the left side [9]. The opening is usually triangular or elliptical in shape which may be within the diaphragmatic muscle or with a complete absence of posterolateral part of the muscle. The abdominal viscera usually involved in this type of hernia are caecum, appendix, ascending colon, transverse colon and all parts of small intestine and sometimes spleen also may be involved. The stomach is usually seen in the abdomen [10,11]. The herniated viscera compresses the developing lung tissue and lead to pulmonary hypoplasia on contralateral sides because of the mediastinal shift which reduces the number of bronchial generations results in hypoplastic lungs [7,12].

In our case, a large elliptical opening was found in the right side of the diaphragm which is a pleuroperitoneal hiatus. Except the liver, descending and sigmoid colon and the urinary organs, all the abdominal viscera are herniated into the thoracic cavity. In pleuroperitoneal hiatus hernia usually

the stomach is retained in the abdominal cavity [11]; but in our case, whole of the stomach was herniated. In addition, part of the right lobe of the liver also was herniated with strangulation formed by the band of diaphragmatic muscle forming a groove on its anterior surface. The left lobe was markedly enlarged. Another striking feature in our case is that there was a complete absence of spleen. The thoracic cavity showed severe hypoplasia of right lung, trilobed left lung and dextrocardiac heart with its apex directed towards the right side.

Pulmonary hypoplasia and hypertension may be the reasons of high mortality rate at birth. The dysfunction of surfactant system combined with cardiac malformations may further complicate the conditions of congenital diaphragmatic hernia [1,13]. The hypoplasia of the right lung may be the reason for the respiratory distress which would have lead to the death of the baby in our case. The agenesis of spleen observed in our case may be associated with congenital malformations of heart and sometimes instead of spleen the other organs like kidneys may also be absent [14].

The occurrence of CRDH associated with other anomalies like gastrointestinal, cardiac and abdominal wall defects which can be diagnosed prenatally, allows the evaluation of fetal life after the birth. The definitive diagnosis of this type of hernias can be demonstrated by the degree of displacement of abdominal organs into the thoracic cavities, by ultrasonographic examinations. Knowledge of this right diaphragmatic hernia during the prenatal and antenatal periods may assure for its management and surgical corrections which can help to reduce the mortality rate.

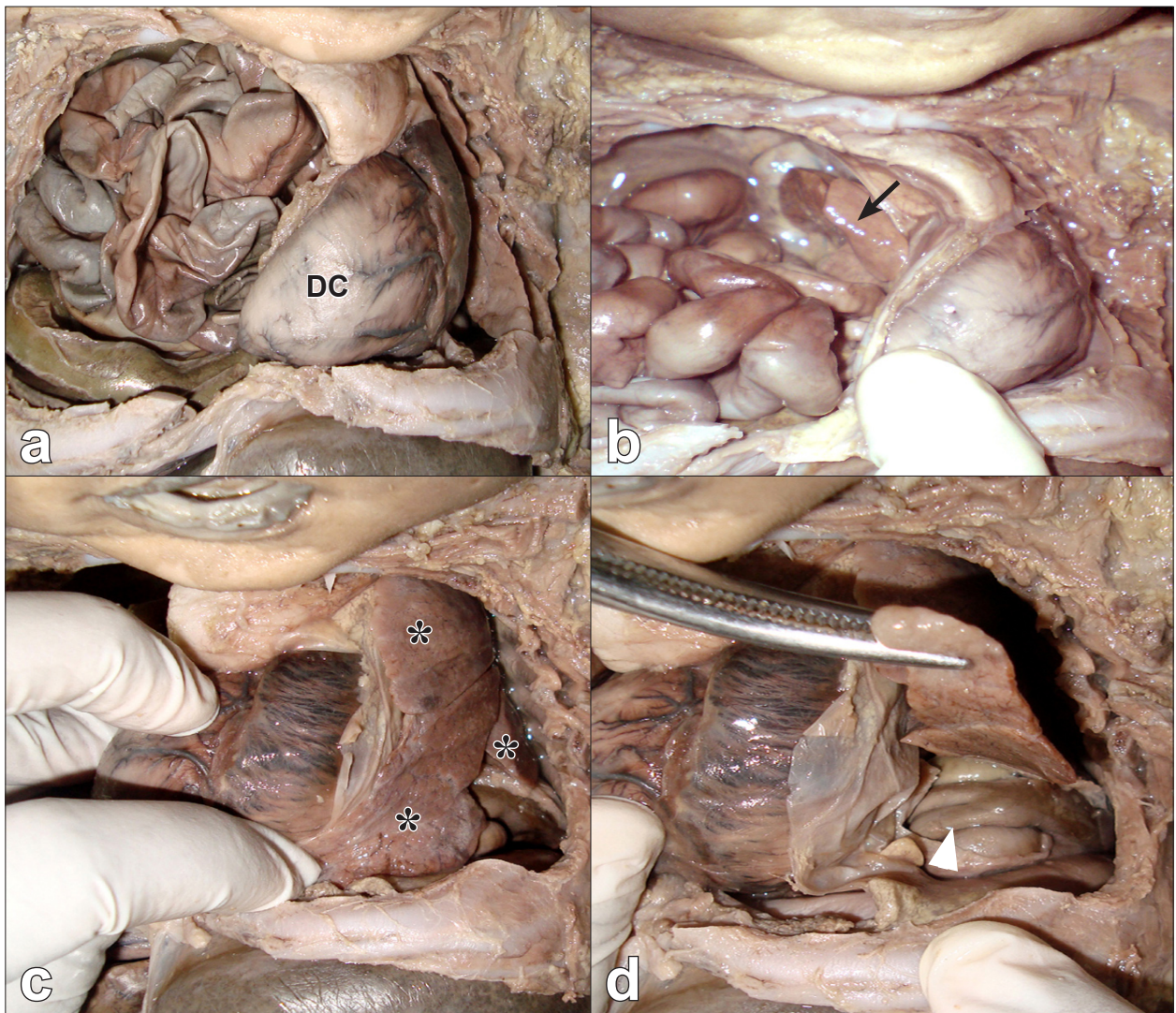


Figure 2. a) Dissection of thoracic cavity showing the left mediastinal shift and *dextrocardia* (DC); b) *hypoplastic right lung* (arrow); c) *trilobed left lung* (asterisk); d) left lung is lifted to show the herniation of the bowel loops from the right to left thoracic cavity behind the *pericardium* (arrowhead).

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