Coexisting Cystic Lesions with Right-Sided Congenital Diaphragmatic Hernia in a Neonate

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Diaphragmatic defects involving the right side of the diaphragm are less common but represent a distinct entity. The clinical presentation, management, and prognosis differ significantly from a left-sided defect. We report a case of a term neonate who was diagnosed to have a congenital diaphragmatic hernia during the third trimester of pregnancy, operated at 36 h of life and subsequently found to have an associated mediastinal cyst in 1st week of life.

Case Report

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Keywords: Congenital diaphragmatic hernia, mediastinal cyst, teratoma, thoracotomy

INTRODUCTION

Congenital diaphragmatic hernia (CDH) involving the right side of the hemidiaphragm is less common and occurs only in around 10%–15% of cases. Nearly half of the cases have an associated congenital anomaly.[1,2] Bowel loops and liver represent the usual content of hernia. An enteric duplication cyst rarely presents as a content of herniated viscera. Teratomas rarely present as a symptomatic mediastinal mass in a neonate. The present case is a rare report showing the association of right-sided CDH with an uncommon and symptomatic mediastinal cyst in neonatal period.

CASE REPORT

A term (382/7 weeks) male neonate with a birth weight of 2480 g was delivered by lower segment cesarean section (indication being transverse lie with spontaneous leaking) to a 21-year primigravida mother, spontaneous conception without any history of consanguinity. The mother was on regular antenatal checkup at a peripheral health facility. There was no history of any radiation exposure or significant drug or substance abuse. Antenatal ultrasonography done at 18 weeks of gestation to detect congenital anomalies was reported as normal. Another ultrasonography done at 32 weeks of gestation detected congenital anomalies was reported as normal. Another ultrasonography done at 32 weeks of gestation was reported abnormal with a loop of dilated proximal bowel inside the thorax. The parents wished to continue expectant management at the peripheral center. After birth, the neonate did not require any resuscitation, but developed respiratory distress soon after and was referred to a higher center.

The neonate was admitted to the neonatal intensive care unit, was normothermic at the time of admission, and had respiratory distress, managed with low flow oxygen through nasal prongs. Chest radiograph was suggestive of cystic hyperlucent lesion with few visible bowel loops in the mediastinum. Computed tomography (CT) of the thorax was performed on 1st day of life and revealed a diaphragmatic defect near the opening of inferior vena cava in the right hemidiaphragm with few loops of intestine inside the chest [Figure 1]. Echocardiography and ultrasonography of the abdomen were done for any associated congenital anomalies that were found to be normal. There was mild pulmonary hypertension (pulmonary artery peak systolic pressure 38 mmHg, normal left ventricular and right ventricular functions) as evidenced from echocardiography, which was managed conservatively. The neonate remained hemodynamically stable with a normal arterial blood gas (pH 7.38, PCO2 41, PO2 96, HCO3 21.6). Surgical repair of the defect was attempted at 36 h of life through the transabdominal route, which revealed a defect of 1.5 cm × 2 cm in the right hemidiaphragm, with a

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loop of jejunum entering into the thorax through the defect and a duplication cyst on the mesenteric wall of jejunum with malrotation. During surgery, loops of the intestine could be pulled to the abdomen, cyst of size 3 cm × 3.5 cm was excised, end-to-end jejunal anastomosis was done, malrotation was corrected by releasing bands, and diaphragmatic defect was closed by nonabsorbable sutures. Diagnosis of enteric (Jejunal) duplication cyst was confirmed after histopathological examination.

In the postoperative period, the baby remained on invasive ventilatory support for 24 h and later was extubated and managed with low flow oxygen. Chest radiograph during postoperative period showed normal lung expansion with nasogastric tube inside the stomach along with the presence of the cystic structure. Arterial blood gas was normal (pH 7.44, PCO2 32, PO2 122, HCO3 19.4). On the 6th postoperative day, the neonate developed high-grade fever with respiratory distress. Laboratory evaluation revealed a normal hemoglobin (16.8 g/dl), total leukocyte count 6400/mm³, absolute neutrophil count 3700/mm³, micro erythrocyte sedimentation rate 8 mm in 1st h, immature-to-total neutrophil ratio 0.15, and C-reactive protein 5.80 mg/dl, with cerebrospinal fluid (CSF) pleocytosis (120 cells/mm³, glucose – 95 mg/dl, protein – 63.3 mg/dl) suggestive of meningitis. Antibiotics were upgraded after sending blood and CSF cultures. Klebsiella species was isolated from blood culture after 48 h and was sensitive to colistin.

On the 7th postoperative day, respiratory distress worsened, requiring noninvasive respiratory support. Chest radiograph revealed a nonhomogeneous cystic structure inside the thorax. Contrast-enhanced CT of the thorax confirmed the presence of a cystic structure inside the thorax [Figure 2]. The neonate was reoperated on day 11 of life. Right thoracotomy was done and a cystic mass of size 7 cm × 5 cm was identified in the posterior mediastinum [Figure 3]. Purulent materials aspirated from the cyst and sent for culture. The cyst was excised and sent for biopsy. Microscopic examination revealed cystic structures lined by squamous epithelium and at places ciliated columnar epithelium, cartilaginous structures, and mucus-secreting glands suggestive of a cystic teratoma.

The infant remained on invasive ventilation for 24 h after surgery and subsequently extubated to noninvasive support. The neonate continued to require noninvasive respiratory support for 7 days and later was managed with low flow oxygen through nasal prong for subsequent 4 days. *Staphylococcus aureus* was isolated from the pus culture specimen, which was treated with intravenous antibiotics.

The infant was discharged from the hospital on day 28 of life and is on regular follow-up for the last 14 months. The infant has been doing well, but there have been a few short hospitalizations for wheezing episodes in the last 6 months attributed to viral infection.

**DISCUSSION**

Right-sided CDH represents a distinct entity. Antenatal diagnosis, clinical presentation, and prognosis have been described to be significantly different from left-sided defects. About 40%–60% of patients have other associated anomalies such as cardiac, urogenital, gastrointestinal, chromosomal, or musculoskeletal anomalies.[3] Right-sided CDH frequently includes liver as a part of herniated viscera. A similar echotexture of the liver and lung may pose diagnostic difficulties with antenatal ultrasonography.[6] Magnetic resonance imaging plays an important role in the preoperative diagnosis of right-sided CDH.
imaging (MRI) has been found to be useful in detecting fetal anomalies and can be a valuable adjunct to evaluate the position of the liver and estimating lung volume.\(^{[5,6]}\)

Over 60% of CDH cases are initially suspected on a routine 18–22 week sonographic fetal anatomic survey. Others may be incidentally identified at a subsequent examination; presentation at an older gestational age may be due to a lack of herniated abdominal contents when the defect is small and has not allowed passage of abdominal contents into the fetal thorax, or because of technical or interpretive issues on an earlier examination.\(^{[2]}\) Sensitivity of ultrasonography is higher when there are associated abnormalities, when the defect is large, with advancing gestational age, and when experienced fetal ultrasonographers are performing the examination.\(^{[7]}\) European reference network suggests to include a detailed fetal anatomic survey, fetal chromosomal microarray, ultrafast fetal MRI to determine the degree of liver herniation and estimate lung volume, and echocardiography during prenatal assessment of patients with suspected CDH.\(^{[8]}\)

In many cases, detailed evaluation may not be feasible, particularly in cases presenting at a later gestational age. Hence, a good preoperative screening is needed in such cases.

Teratomas are congenital tumors that contain derivatives of all three germ layers and arise from pluripotent embryonal cells. Recent classifications also include monodermal types.\(^{[9]}\) The lesions can be heterogeneously solid and cystic. Immature teratomas are less common than their mature counterparts but tend to grow faster and to a larger size. They commonly occur in sacrococcygeal region. Mediastinum is the second most common extragonadal site of occurrence of teratomas, predominantly arising in anterior mediastinum and occasionally in posterior mediastinum.\(^{[10]}\) Most anterior mediastinal teratomas are diagnosed on prenatal scan. With two-dimensional ultrasonography, the diagnosis of mediastinal teratoma is comparatively easy if a multilobulated mass has both cystic and solid components with calcification and acoustic shadows. However, in the absence of calcification, as in the present case, a correct diagnosis from the prenatal ultrasonography can be difficult.\(^{[11]}\) CT scan is of great value for the diagnosis of mediastinal teratoma.\(^{[11]}\) Some teratomas may remain asymptomatic and are often discovered incidentally on chest radiographs. Large lesions may cause fetal hydrops or airway compromise and may require intubation and care in intensive care unit.\(^{[12]}\)

Although the case was managed successfully in postnatal life, few points in the management need to be highlighted. First, the diaphragmatic defect was not detected during fetal anomaly scan and though detected at a later scan, a thorough evaluation of severity and associated anomalies was not done. This may be related to a lack of experience and facility at a peripheral health setup. The Canadian CDH Collaborative guideline suggests the need of ultrasonographic assessment of observed-to-expected lung–head ratio between 22 and 32 weeks of gestational age to predict the severity of pulmonary hypoplasia in isolated CDH, and fetal MRI may be useful for assessment of lung volume if a facility is available.\(^{[13]}\) Second, postnatal course of CDH is frequently complicated by the onset of pulmonary hypertension. In the present case, the patient developed mild pulmonary hypertension as evidenced from echocardiography, and could be managed conservatively. The neonate remained hemodynamically stable with a normal arterial blood gas. Considering relative clinical stability of the baby, a decision not to intubate was taken. As many as 45% cases of CDH, have an associated intestinal rotational abnormality. Routinely, correction of malrotation in cases of CDH is not done. Although, routinely not needed, many surgeons prefer to correct malrotation. In a recent retrospective study, nearly 88% of malrotation were corrected during initial repair, and a complete Ladd’s procedure was performed in 34% of cases.\(^{[14]}\) Due to the presence of adhesion bands in small intestine, a decision to correct malrotation was taken in the present case. Third, the presence of mediastinal cyst was missed in CT scan done after birth. The presence of two separate cystic structures of similar echogenicity might have been the reason for false interpretation. This highlights the importance of thorough evaluation during postnatal period to look for other associated anomalies using MRI. The severity of pulmonary hypoplasia and/or pulmonary hypertension helps to decide the timing of surgical correction. As the neonate remained hemodynamically stable and there was no significant pulmonary hypertension, surgical repair...
was attempted at 36 h of life. Surgical repair of CDH can be accomplished through a thoracic or abdominal approach. In the present case, the defect was approached through a subcostal incision. During primary repair, the defect was found to be small (1.5 cm) and the loops of intestine could be pulled back to the abdominal cavity and the diaphragmatic defect was closed. Although a cystic structure inside the thorax was observed in the initial CT scan, it was attributed to the jejunal cyst observed during surgery. Presence of two cystic structures could not be realized during initial surgical correction in this case, and the mediastinal cyst was missed during initial surgical repair. The fact that the defect was smaller and the surgeons could pull the intestinal contents back to the abdominal cavity might have led to an assumption of successful primary repair.

**CONCLUSION**

Detailed evaluation and assessment of severity should preferably be done at a tertiary care center in antenatally suspected cases of CDH. A high index of suspicion for other malformations, thorough postnatal clinical and radiological evaluation are necessary even in cases of isolated CDH detected during antenatal period.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

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