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Thoracoscopic diagnosis and repair of central congenital diaphragmatic hernia in a neonate: A case report of a rare entity

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ABSTRACT

Introduction: Central congenital diaphragmatic hernia that involves a central tendon defect is a very rare form of diaphragmatic hernias. Experience in the thoracoscopic repair of congenital diaphragmatic hernia has expanded in recent years, but this expansion was mainly for the most common type; the posterolateral. We report a case of central congenital diaphragmatic hernia, which is supposed to be the case number seventeen of those reported in literature. And we report the first use of thoracoscopic repair in such rare entity.

Case presentation: We present a case report of a Bengali neonate who had a prenatal diagnosis of left congenital diaphragmatic hernia, which was supposed to be the usual posterolateral type. As we usually approach such cases by thoracoscopy, the patient had an intraoperative diagnosis of central congenital diaphragmatic hernia. The defect was amenable to a successful thoracoscopic repair.

Conclusion: The rare neonatal central congenital diaphragmatic hernia could present as a left sided herniation that clinically resembles the usual posterolateral congenital diaphragmatic hernia. Thoracoscopic approach offers a tremendous tool for diagnosis as well as the management of this kind of presentation of a central congenital diaphragmatic hernia.

1. Introduction

Congenital diaphragmatic hernia (CDH) is a developmental discontinuity of the diaphragm that allows abdominal viscera to herniate into the chest. Central CDH that involves a central tendon defect is a very rare form of CDHs [1], with only few reported cases in the literature. Experience in thoracoscopic repair of CDH has expanded in recent years [2–5], but this was mainly in the posterolateral type of CDH. We present a case report of a neonate who had an intraoperative diagnosis of central CDH. We also report the first use of thoracoscopic repair of such rare entity.

2. Case report

A male Bengali newborn, who had a prenatal diagnosis of left diaphragmatic hernia (diagnosed at the 23rd week of gestation by fetal ultrasound screening and fetal MRI). He was delivered by cesarean section at 38 weeks of gestational age and had a birth weight of 3100 g. At birth, 1-min and 5-min Apgar score were 3 and 8, respectively. He was intubated and high frequency oscillating ventilation (HFOV) was used. Nasogastric tube was inserted and chest radiography has confirmed the diagnosis (Fig. 1). Echocardiography has shown small patent ductus arteriosus and patent foramen ovale, with bidirectional shunting and no pulmonary hypertension. There were no other associated malformations.

At day 2 of life, the newborn had a stable cardiorespiratory status, and thoracoscopic repair was performed. The patient was operated under general anesthesia while maintained on the HFOV machine. He was laid down on right lateral decubitus position. A 5 mm trocar for the optic lens was placed just below the edge of the scapula, and two further 3 mm operative trocars were placed, one in the 5th intercostal space (ICS) over the anterior axillary line, and the other in the 5th ICS midway between the optical trocar and spine. Once entered in the pleural cavity, the hernial sac and its contents were pushed down toward the abdominal cavity by the effect of intrathoracic CO_2 insufflation, direct gentile pushing by the surgeon, and by gravity due to the slight reverse Trendelenburg

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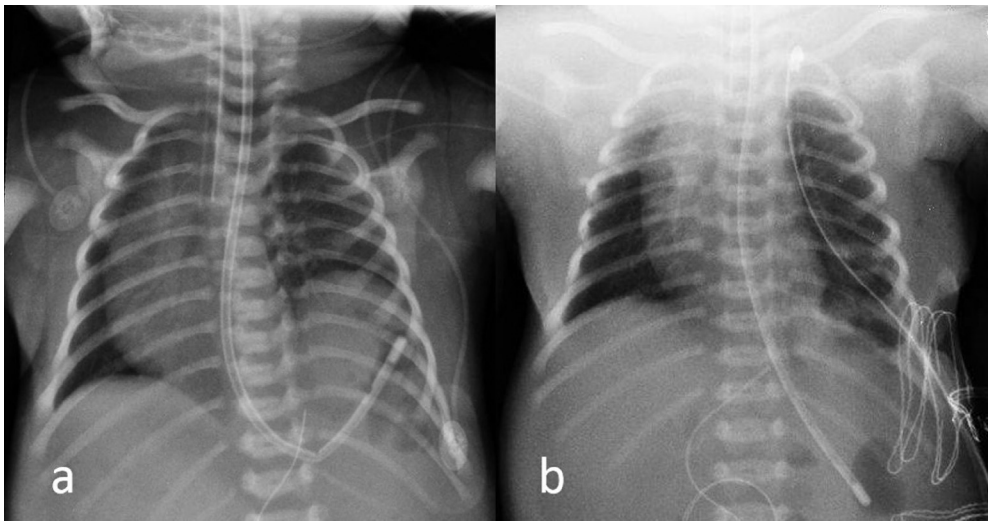

Fig. 1. a) Chest x-ray at birth showing the left-sided CDH with shifting of mediastinum toward the contralateral side. The nasogastric tube demonstrates the intra-thoracic position of the stomach. b) Chest x-ray obtained immediately postoperatively.

position. The diaphragmatic defect was not as usually found in most left-sided CDHs in the posterolateral position. Instead, it was bulging from beneath the pericardium and along the central portion of the diaphragm toward the left hemithorax. There was a thin hernial sac with good rim of surrounding diaphragmatic muscle. By definition, diaphragm eventration, either partial or complete, does not imply on such presentation. Therefore, an intraoperative diagnosis of central CDH due to a central tendon defect was assigned. The defect was repaired primarily with the sac being plicated using interrupted nonabsorbable stitches. The procedure was completed thoracoscopically (Fig. 2). Chest tube was left in place. The duration of operation was 145 min.

Post-operatively, the patient was on HFOV for 2 days, and then conventional ventilation was used. The patient was extubated 10 days later. Inotropic cardiac support was used for several days. The child had good cardiorespiratory improvement, and was discharged after two weeks. The child had a follow up of three years with a good general outcome and no complications.

3. Discussion

Congenital diaphragmatic hernia is yet a challenging malformation that necessitates highly specialized and experienced tertiary centers' role of management, especially for neonatal age group. Since the first laparoscopic CDH repair has been published in 1995 [6], minimally invasive approach has been widely adopted for management of CDH among tertiary centers worldwide.

Most CDHs (80–90%) are of the Bochdalek or posterolateral type, with much smaller numbers of anterior or Morgagni defects (2–5%) [1]. Perhaps, the most unusual and rarely reported is that of the central tendon defect [1]. There have been 16 cases of central CDHs reported in the literature [7–10]. Only one case of which had

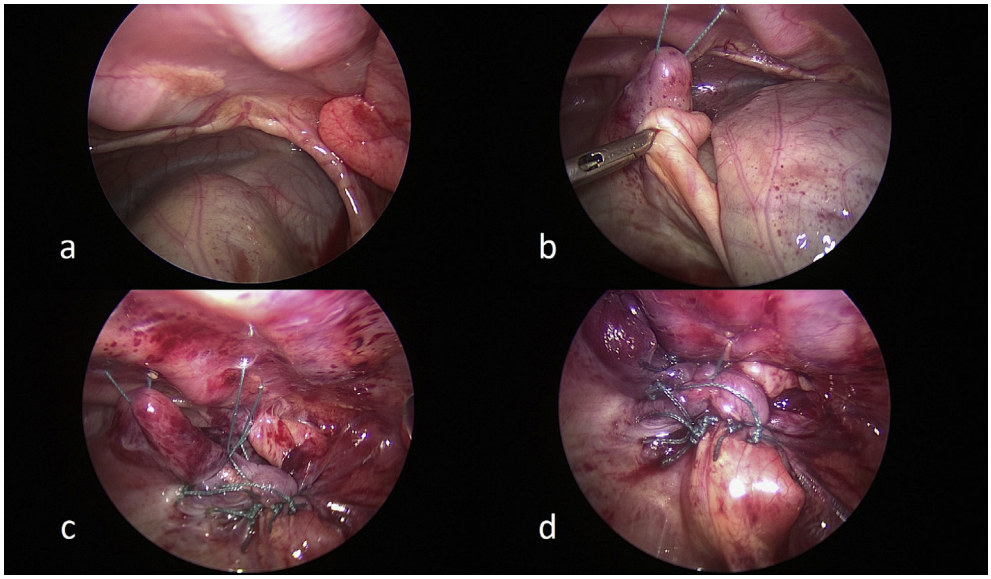

Fig. 2. Thoracoscopic approach for management of central CDH. a) The diaphragmatic defect is shown along the central tendon of the diaphragm. b) Approximation of the diaphragmatic edges using a suspension thread through the chest wall. c,d) Primary closure of the defect using nonabsorbable interrupted stitches.

a laparoscopic repair [8]. Our case presents the 17th "central CDH" case, and the first one managed in thoracoscopic approach.

Central diaphragmatic hernia often presents with massive pericardial effusion [7], especially when there is direct communication between the peritoneum and the pericardium with no hernial sac. In our case, there was a hernial sac and abdominal organs were herniated toward the left hemithorax. This explains the antenatal and preoperative suspicion of the more common left Bochdalek hernia, which we usually manage in thoracoscopic approach. To our knowledge, this is the first recorded thoracoscopic repair of such a defect in a neonate.

The outcome for patients with central tendon defect appears to be good. Of the 17 cases; those in the literature and ours, 13 (76%) survived [7–9], one was aborted antenatally, 2 died shortly after birth, and one died 2 days postoperatively due to sepsis [7,10]. All survivals, of the different surgical approaches, had successful repairs with no documented recurrences.

Minimally invasive techniques have the potential advantages of improved visualization, less need for narcotic postoperatively, and shorter length of intubation [11], in addition to the improved cosmesis, and faster recovery. This makes thoracoscopy to be a potential standard tool in the diagnosis and management of similar cases.

4. Conclusion

A central CDH should be considered in a neonate referred with a CDH of any other type or of any unusual presentation, like complete whitening of lung fields, or pericardium, or any other chest radiograph that is difficult to interpret. Thoracoscopy resembles a good diagnostic tool of such challenging cases that are usually difficult to be diagnosed preoperatively. In addition, thoracoscopic repair of central CDH, especially those with hernial sac, is a feasible and safe procedure, and results in good outcome.

Conflict of interest

Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

The authors declare that they have no competing interests.

RT carried out data collection, literature review and drafted the manuscript. TG participated in literature review, sequence alignment of the manuscript. MM contributed to draft the manuscript and preparation of figures. ML conceived the study, provided critical revision of the manuscript, approved the final version of the manuscript. All authors read and approved the final version of the manuscript.

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