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Case Report

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Incidental extralobar pulmonary sequestration during thoracoscopic repair of congenital diaphragmatic hernia: A rare association

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Abstract

Congenital diaphragmatic hernia (CDH) is a rare malformation with high morbidity and mortality, and associated anomalies may be detected prenatally or encountered during repair. The coexistence of CDH and bronchopulmonary sequestration (BPS) is rare, and prenatal imaging often fails to detect both anomalies simultaneously. Herein, we reported a male neonate delivered at 37+4 weeks of gestation who underwent thoracoscopic repair for left posterolateral CDH, during which an extralobar pulmonary sequestration was incidentally identified and excised. The coexistence of BPS and CDH presents a unique clinical scenario with potential implications for diagnosis and surgical management. This case emphasizes the importance of detailed prenatal assessment and vigilant intraoperative inspection in CDH patients. Associated anomalies should always be considered, and thoracoscopy provides both diagnostic and therapeutic benefits in this setting.

Keywords: Congenital diaphragmatic hernia, bronchopulmonary sequestration, thoracoscopic repair.

Congenital diaphragmatic hernia (CDH) is a rare but life-threatening malformation, affecting approximately 1 in 2,500 live births, and remains associated with high morbidity and mortality.[1] It results from a diaphragmatic defect that permits herniation of abdominal viscera into the thoracic cavity, often leading to pulmonary hypoplasia and cardiorespiratory compromise.

Bronchopulmonary sequestration (BPS) is another uncommon congenital anomaly, characterized by nonfunctioning lung tissue with systemic arterial supply and no communication with the tracheobronchial tree.[2] Bronchopulmonary sequestration is classified as intralobar pulmonary sequestration and extralobar pulmonary sequestration and is more often associated with other anomalies, including CDH. The coexistence of CDH and BPS is rare and may be recognized antenatally or diagnosed incidentally during surgery.[3-6]

Herein, we reported a neonate with left-sided Bochdalek CDH who underwent thoracoscopic repair, during which an incidental EPS was identified and excised.

CASE REPORT

A male neonate weighing 3655 g was delivered at 37+4 weeks of gestation to a 38-year-old gravida 6, para 5, abortus 1 mother by cesarean section due to acute fetal distress. The APGAR scores were 8 and 9 at 1 and 5 min, respectively. The infant cried immediately after birth, and delayed cord clamping was performed for 60 sec. Initial stabilization achieved target oxygen saturations without the need for advanced resuscitation.

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No fetal anomaly was detected during antenatal evaluation. A written informed consent was obtained from the parent of the patient.

Following the onset of postnatal respiratory distress, a chest radiograph demonstrated bowel loops in the left hemithorax (Figure 1), consistent with congenital diaphragmatic hernia, upon which the patient was consulted to our department. The patient was intubated and transferred to the neonatal intensive care unit. Echocardiography revealed structurally normal cardiac anatomy with patent foramen ovale (left-to-right shunt), mild tricuspid regurgitation (maximal gradient 25 mmHg), and no significant pulmonary hypertension. Transfontanel and abdominal ultrasonography were normal.

After clinical stabilization, thoracoscopic repair was decided on the second postnatal day. The patient was placed in a right lateral decubitus and mild prone position. A 5-mm optical port was introduced through the fourth intercostal space in the left mid-axillary line, and two additional working ports, one 5 mm and the other 3 mm, were placed in the fifth intercostal space on the anterior and posterior axillary lines, respectively. The surgeon and the camera assistant stood at the anterior side of the patient, the scrub nurse was positioned toward the anterior caudal side, and the anesthesiologist at the head side; the monitor was placed at the posterior caudal side (Figure 2). A pneumothorax was created by carbon dioxide insufflation at a pressure of 5 mmHg. Intraoperatively, a 3×4 cm posterolateral diaphragmatic defect covered by a hernia sac was identified, through which the intestinal loops and stomach had herniated into the thoracic cavity. During exploration, an extralobar pulmonary sequestration arising from the lower thoracic cavity was identified, receiving its arterial blood supply from the thoracic aorta. The hernia sac was preserved, the herniated viscera were reduced, and the defect was closed with interrupted 3-0 Prolene sutures reinforced with a polypropylene mesh overlay. The anomalous systemic feeding artery was ligated with clips, and the sequestrated segment was excised (Figure 3). A chest tube was placed before closure. The total operative time was 120 min. There was no intraoperative or postoperative bleeding, and no blood transfusion was required.

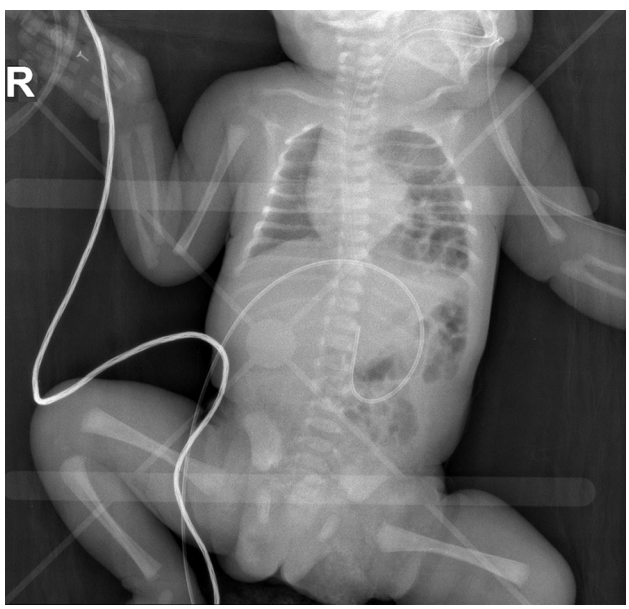


Figure 1. Postnatal babygram showing bowel loops displaced into the left hemithorax.

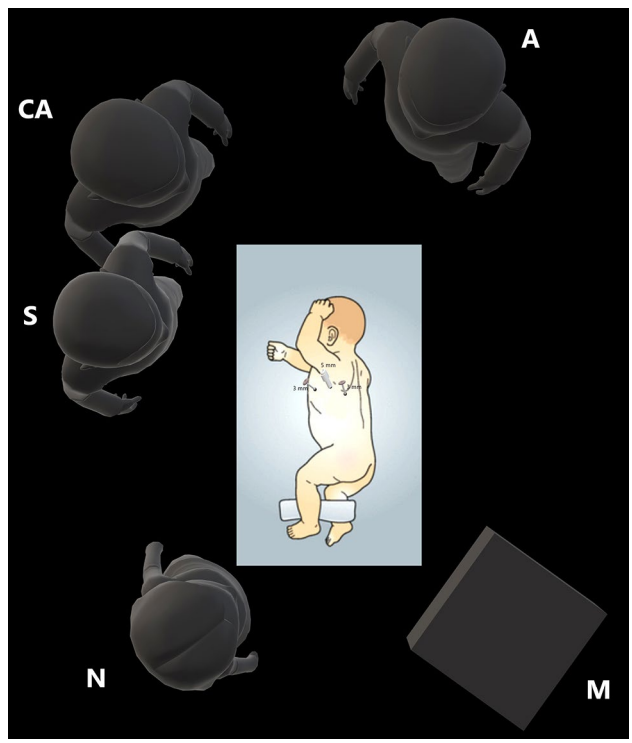


Figure 2. Operative setup and port placement. The patient was positioned in a right lateral decubitus position with a slight prone tilt.

S: Surgeon; CA: Camera assistant; N: Scrub nurse; A: Anesthesiologist and monitor (M) orientation are shown schematically.

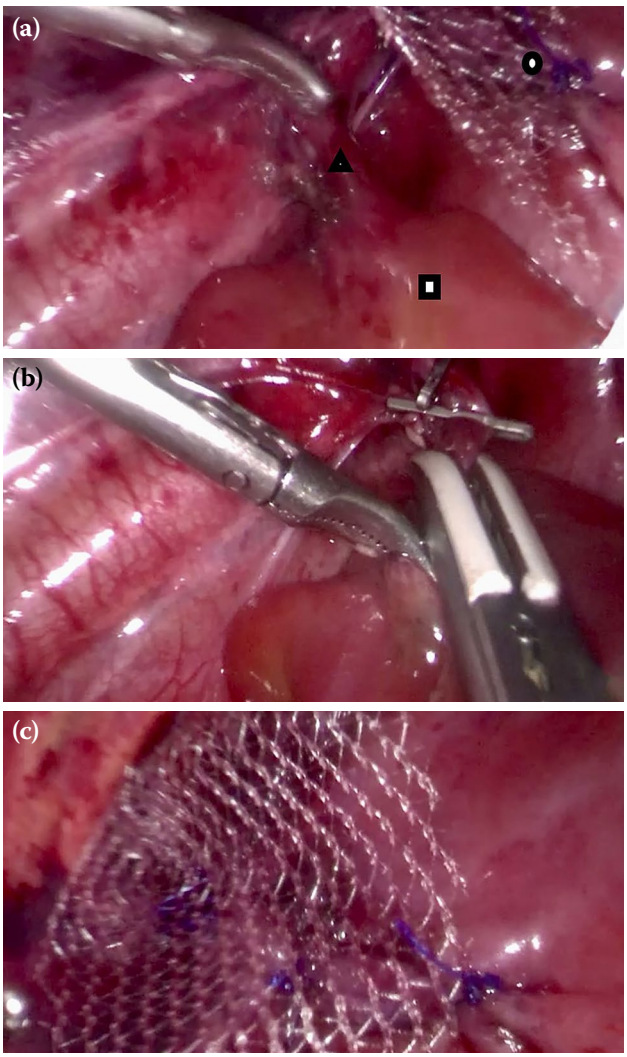


Figure 3. Intraoperative views. **(a)** Thoracoscopic view during diaphragmatic repair (circle) demonstrating a bronchopulmonary sequestration (square) with its systemic arterial supply originating from the aorta (triangle). **(b)** The aberrant arteries arising from the aorta are clipped and divided. **(c)** The diaphragmatic defect is closed and reinforced with a polypropylene mesh.

Postoperatively, the infant required prolonged ventilatory support and was extubated on the ninth day, and the thoracic drain was removed on the same day. Noninvasive support was gradually weaned, and the patient was breathing room air by the 15th day. The postoperative course was uneventful, and the patient was discharged in good condition. Follow-up at two months demonstrated normal growth and respiratory status; a chest radiograph showed good diaphragmatic border (Figure 4).

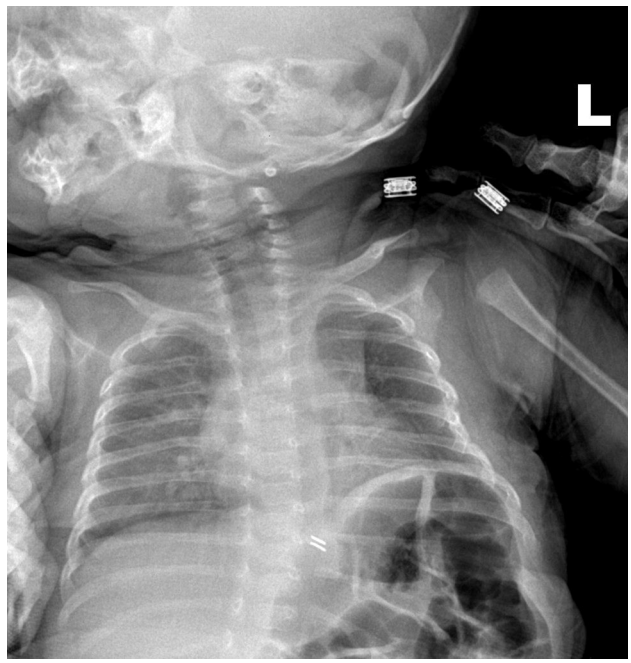


Figure 4. Postoperative chest radiograph at three months demonstrating a well-contoured and intact left diaphragm.

DISCUSSION

The coexistence of CDH and BPS is extremely rare, and large-scale data from the CDH Study Group indicate a prevalence of approximately 3.4% for CDH with concurrent BPS, aligning with smaller series that report rates ranging from 1.2 to 30%.[3,4] Conversely, CDH has been reported as the most frequent congenital anomaly associated with BPS, occurring in up to 40% of cases, suggesting a developmental interplay between the two conditions.[7] Its embryologic basis can be linked to overlapping critical periods of thoracoabdominal development. Human diaphragm formation begins around the fourth week of gestation, and closure is typically completed by the eighth week, while aberrant pulmonary tissue may detach from the tracheobronchial tree during the same early gestational period. This temporal overlap suggests that disturbances in early embryogenesis may predispose to the simultaneous development of both anomalies, with diaphragmatic defects potentially facilitating abnormal positioning or sequestration of pulmonary tissue.[2] Embryological theories propose that sequestration develops at

four to five weeks of gestation and may interfere with diaphragm closure at around 10 weeks. The mass effect of sequestration may act as a physical barrier, preventing normal fusion of the pleuroperitoneal canals, thereby facilitating CDH formation. Notably, two contrasting outcomes have been described: in certain fetuses, sequestration appears to exert a protective effect by restricting herniation and facilitating lung development, while in others it exacerbates the diaphragmatic defect, thereby intensifying pulmonary hypoplasia and contributing to unfavorable outcomes.[8-10] In our patient, the postnatal and perioperative course was uneventful, with no evidence of pulmonary hypertension. Therefore, despite the potential for adverse interactions between CDH and BPS, this coexistence did not appear to negatively impact the clinical outcome in this case; in fact, the course was favorable.

With the advances in prenatal ultrasound and fetal magnetic resonance imaging, simultaneous CDH and BPS often diagnose antenatal detection.[6] However, sequestration may serve as an "anatomical barrier," preventing visceral herniation and masking the diaphragmatic defect.[10,11] Thus, when abdominal contents herniate early, they may obscure sequestration itself. Therefore, the presence of both lesions complicates prenatal imaging and counseling. Postnatally, computed tomography or magnetic resonance imaging can also assist in characterizing sequestration, but some cases of BPS may only be recognized intraoperatively, as in our patient.

The management of BPS associated with CDH is surgical excision.[12] Although observation has been proposed for asymptomatic lesions, surgical resection of BPS has been advocated to prevent potential complications, including infection, hemorrhage, and the rare risk of malignant transformation.[12,13] In cases of concomitant CDH and BPS, simultaneous correction by a thoracoscopic approach is feasible even in neonates.[14] Although laparotomy allows successful repair of CDH, we consider thoracoscopy advantageous, as it provides superior visualization, facilitates secure ligation of aberrant systemic vessels, and enables simultaneous CDH repair and BPS resection. In contrast, transabdominal approaches may carry the risk of missing small sequestrations hidden within the thoracic cavity. In our case, the sequestration was identified solely due to thoracoscopic exposure and excised safely; this lesion might not have been detected through the abdominal approach.

In our case, a hernia sac was present. It has been emphasized that differentiating a diaphragmatic hernia with a sac from a localized eventration remains challenging. While sac excision has been associated with lower recurrence rates, the presence of a sac during thoracoscopy may facilitate reduction and reduce the risk of visceral trauma.[15] In our patient, it is conceivable that a weakened diaphragm herniated close to term, explaining the absence of antenatal diagnosis. On thoracoscopy, the herniated internal organs were easily reduced, and the repair was completed, leaving the sac. Notably, previous reports of CDH associated with pulmonary sequestration have also described the presence of a hernia sac, suggesting that this finding may not be coincidental but rather reflect a shared developmental mechanism.[12-14]

The prognostic significance of BPS in CDH remains debated. Some authors suggest sequestration may confer a protective effect by limiting visceral herniation, thereby improving lung growth.[3,10,11] Diesen and Megison[5] have reported no survival benefit or even poorer outcomes in patients with CDH and BPS, potentially because of larger diaphragmatic defects and a greater need for patch repair. Furthermore, BPS may remain undetected when an abdominal surgical approach is used. Large-scale analyses, such as that by Coughlin et al.,[3] suggest higher extracorporeal membrane oxygenation use and mortality in CDH and BPS cases, while smaller series report potential protective effects. Overall, we believe that the prognosis continues to be driven primarily by CDH severity and pulmonary hypertension, with BPS serving as a potential but inconsistent modifier.

In conclusion, the coexistence of CDH and BPS is rare and may be diagnosed during the antenatal/postnatal period or surgery. If detected, simultaneous CDH repair and BPS resection can be performed safely. Our case highlights the diagnostic and therapeutic advantage of the thoracoscopic approach, which not only allowed for definitive

CDH repair but also facilitated the incidental identification and safe excision of an associated pulmonary sequestration.

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AI Disclosure: The authors declare that artificial intelligence (AI) tools were not used, or were used solely for language editing, and had no role in data analysis, interpretation, or the formulation of conclusions. All scientific content, data interpretation, and conclusions are the sole responsibility of the authors. The authors further confirm that AI tools were not used to generate, fabricate, or 'hallucinate' references, and that all references have been carefully verified for accuracy.

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