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Case Report

Complex Surgical Management of a Symptomatic Morgagni-Larrey Hernia in an Adult with Down Syndrome: A Rare Case

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Abstract

The presentation of a symptomatic Morgagni-Larrey hernia in adults is an exceptional clinical occurrence, particularly among individuals with Down syndrome, where congenital diaphragmatic hernias are commonly addressed during infancy. This report presents a rare case of a 25-year-old male with Down syndrome who experienced respiratory distress due to a Morgagni-Larrey hernia. The case is distinguished by the absence of the anteromedial edges of the hernia, which posed a significant surgical challenge. To manage this unique presentation, a mesh plug technique was adopted, and tailored to accommodate the intricate anatomical considerations. This case underscores the critical need for surgical innovation in the treatment of complex hernia presentations and highlights the rarity of such cases in the adult Down syndrome population, emphasizing the importance of tailored surgical intervention in these patients.

Keywords: *Morgagni; Morgagni-Larrey; Complex Hernia; Down Syndrome; Complex Hernia*

Introduction

Congenital diaphragmatic hernia (CDH) signifies a spectrum of pathological anomalies characterized by an abnormal opening in the diaphragm, permitting the intrusion of abdominal viscera into the thoracic compartment. This condition is predominantly identified in neonatal period, exhibiting a survival rate of approximately 67% [1]. The occurrence of CDH stands between 0.8 to 5 instances per 10,000 live births, influenced by a confluence of genetic, environmental, and nutritional factors [2]. Within the CDH classification, several variants are recognized, including Bochdalek hernia, Morgagni hernia, diaphragm eventration, and central tendon defects.

Among these, the Morgagni hernia is particularly rare, accounting for a mere 2% to 5% of all congenital diaphragmatic hernia cases [3]. This variant is distinguished by herniation through the foramina of Morgagni, anatomically situated postero-inferior to the xiphoid process. Predominantly right-sided due to additional support from pericardial attachments on the left diaphragm, Morgagni hernias present on the left in about 5% of cases and bilaterally in 4% [6]. When occurring on the left, the condition is specifically termed a Morgagni-Larrey hernia.

The phenotypic expressions associated with Morgagni hernias often include cardiac anomalies, ranging from 25% to 60%, and a notable 15% to 71% concurrence with trisomy 21 [4]. The propensity for these hernias in individuals with Down syndrome is conjectured to stem

from impaired dorsoventral migration of rhabdomyoblasts, a process exacerbated by the increased cellular adhesion characteristic of trisomy 21. This pathophysiological insight provides a rationale for the observed heightened recurrence rates of herniation post-repair in the Down syndrome population [5].

Case Presentation

A 21-year-old male with Down syndrome, presenting with a complex medical profile inclusive of bronchial asthma, type I diabetes mellitus, epilepsy, and hypothyroidism, was referred to our institution with concurrent pneumonia and congenital diaphragmatic hernia. Initially managed at a secondary facility, the patient required intubation and admission to the Intensive Care Unit (ICU) due to respiratory failure compounded by uncontrolled seizures, despite the administration of bilevel positive airway pressure (BiPAP).

Upon transfer, the patient was sedated, mechanically ventilated, and stabilizing under the following settings: FiO_2 at 40%, PEEP set to 5 cm H_2O, with satisfactory oxygen saturation (SpO_2 at 97%). Vital signs were recorded with a heart rate (HR) of 76 bpm, blood pressure (BP) of 112/88 mmHg, and temperature (T) maintaining at 37°C. Neurological assessment via the Glasgow Coma Scale (GCS) rendered a score of 10 out of 15. Pulmonary assessment evidenced bilateral air entry without disparity, and abdominal palpation indicated a soft, non-distended profile.

Diagnostic radiography of the chest (Figure 1) depicted bilateral infiltrates with notable opacification and consolidation localizing to the right upper and middle lobes. A mediastinal shift to the right was apparent, accompanied by visualization of bowel loops within the left thoracic space.

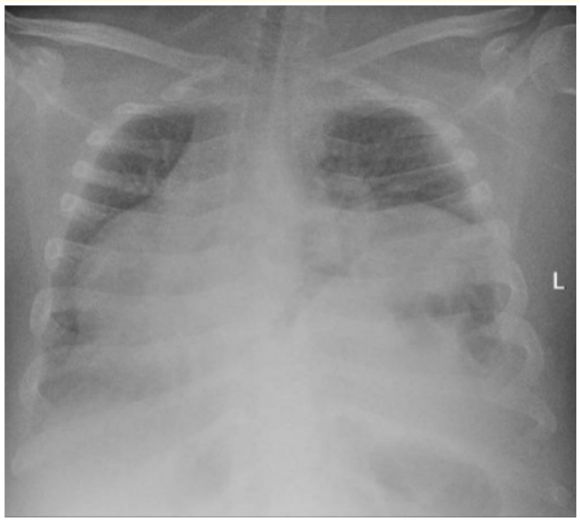

Figure 1: *Chest x-ray (anteroposterior) showing mediastinal shift to the right side with view of bowel loops in the left pleural cavity.*

Subsequent computed tomography (CT) of the thorax with IV contrast delineated a significant left-sided diaphragmatic hernia (Figure 2 and 3), measuring 9 cm and encapsulating mesenteric fat alongside the lower pulmonary lobes. Additionally, the left lung demonstrated diffuse ground-glass opacities, and atelectasis was bilaterally evident with accompanying small pleural effusions.

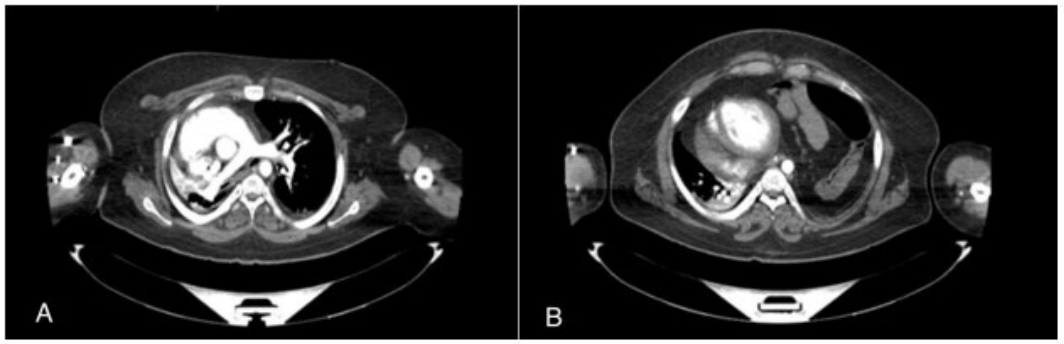

Figure 2: *Computed tomographic scan (transverse section) showing A) heart completely shifted to the right side and B) bowel loops herniating through the left side of the diaphragm.*

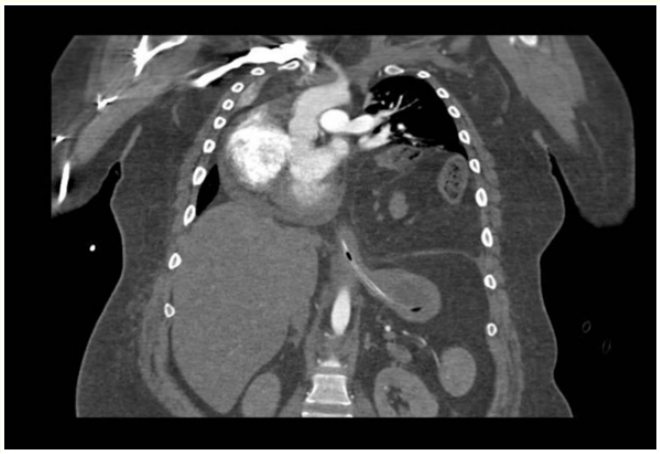

Figure 3: *Computed tomographic scan (sagittal section) showing the diaphragmatic defect with herniated bowel loops in the pleural cavity.*

The patient's ICU tenure spanned 5 days, during which a regimen of vancomycin and meropenem was administered to manage pneumonia. Post-stabilization and extubation, a left thoracotomy was scheduled under general anesthesia. In preparation, multi-disciplinary consultations were conducted to achieve optimal preoperative homeostasis, including glycemic regulation and seizure prophylaxis.

Intraoperatively, via a left lateral thoracotomy approach, a pronounced anteromedial diaphragmatic defect was encountered, hosting intrathoracic large bowel and omentum (Figure 4). Following omentectomy and reduction of the herniated bowel trans-fixation was carried out for the hernial sac. The endeavor to approximate the hernia margins was met with difficulty, given the obscured anteromedial edge behind the xiphoid and the defect's extensive dimensions. Anterior to the medial side of the hernia was the xiphoid process and medial to it was the pericardial tissues, a mesh plug technique was utilized just above the liver and mesh was fixed anteriorly to sternal periosteum and medially to pericardial tissue and posteriorly to the remaining parts of the diaphragmatic muscle and laterally to the left hemidiaphragm ensuring full closure of the defect.

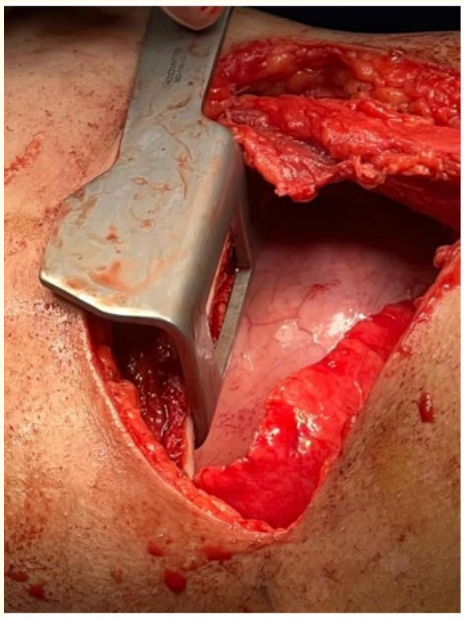

Figure 4: *Open thoracotomy with bowel present within the pleural cavity.*

Postoperative care entailed continued ICU monitoring with subsequent removal of the anterior chest tube on postoperative day two and extubation on day three. The patient was transitioned to ward care in a stable state, with the posterior chest tube being removed by the fifth day, marking satisfactory pulmonary re-expansion. The patient's recovery was uneventful, and was soon discharged home.

The patient was followed up in the clinic 1 month to 1 year after discharge, showing improved respiratory function, complete bilateral lung expansion and no recurrence.

Discussion

The Diaphragm is a muscular structure developed from the embryonic remnants, the septum transversum, the pleuroperitoneal folds, and the somites [7]. Morgangi hernia occurs when the abdominal contents herniate through an anterior defect that is created when the anterior pleuroperitoneal membrane fails to fuse with the sternum and costal cartilages, resulting in an anatomical defect in the costosternal trigone called the foramina of Morgagni [8]. Of all CDH, Morgagni hernia is the rarest presenting mostly in childhood [9]. With some reports documenting its presentation in adulthood [10,11].

Morgagni-Larrey hernia, a rare clinical entity, was first described by an Italian anatomist Giovanni Battista Morgagni. Larrey never described a hernia but rather described that the sternocostal triangle (Larrey's space) can be used as a surgical approach to drain cardiac tamponade. Therefore, it has been suggested that any anterior diaphragmatic hernia, whether right or left, should be named as the hernia of Morgagni, or Morgagni-Larrey hernia [12].

Most of the cases of Morgagni hernia are diagnosed incidentally on a chest radiograph and they are mostly right-sided, whereas left sided hernias such as with our case, are rare. In the former type, the herniated mass comprises of omentum followed by the colon and small intestine, while in the later types, the most herniated type is the stomach [13], in addition to this, the Morgagni-Larrey hernia sac mostly contains the transverse colon with the omental fat.

If symptomatic, Morgagni hernia can mimic respiratory or cardiovascular diseases, and in cases where the viscera get strangulated or incarcerated, abdominal pain prevails. CECT scan is the investigation of choice for such cases, as it can provide details of the anatomy of hernia along with its contents and the presence of any complications. Diagnosis is established once a retrosternal mass of fat density or a combination of omentum and a hollow viscus organ is present [14]. In cases of diagnostic difficulty such as the need to differentiate CDH from other mediastinal masses, magnetic resonance imaging (MRI) has been reported to be used [15].

Surgical repair of the defect is advised even in asymptomatic cases to prevent complications such as strangulation. Bowel obstruction, volvulus, and/or necrosis, which can occur in up to 10% of cases [16], [17]. The approach for hernia repair can either be via abdominal approach (laparotomy or laparoscopically) or transthoracic approach [18]. There is no common consensus on which approach is better.

The hernia defect can be repaired using Mesh or simply by non-absorbable sutures. Decision whether to use or not use Mesh depends on the size of hernia and the possibility of achieving a tension-free repair without a prosthesis. In a case series of 36 patients, it has been reported that there can be successful repair of the defect without using Mesh and have no recurrence [19]. Risk factors for recurrence include the closure of the defect under tension without the use of a patch, leaving the sac in place without resection, use of absorbable suture for repair, and a patient history of Down syndrome [17].

Conclusion

This case illustrates a rare instance of a symptomatic Morgagni-Larrey hernia in an adult patient with Down syndrome, an atypical presentation given the predilection for addressing congenital diaphragmatic hernias in pediatric patients. The complexity of the case was accentuated by the absence of the anteromedial edges of the diaphragmatic defect and its proximity to vital structures such as the xiphoid process and pericardium. The successful management of this case, despite the surgical challenges, underscores the critical importance of individualized operative strategies and interprofessional collaboration in managing rare and anatomically complex conditions.

Moreover, the case highlights the necessity for vigilance in the postoperative period, particularly in patients with Down syndrome who may exhibit a heightened risk for recurrence. The employment of a mesh plug technique in the context of absent diaphragmatic edges presents a viable option for ensuring robust repair and minimizing the potential for recurrence. This case contributes to the limited body of literature on Morgagni-Larrey hernias in adults with Down syndrome and serves as an insightful reference for the surgical management of such intricate cases.

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