

Congenital Mixed Type III Hiatal Hernia Revealed by Episodes of Cyanosis in a Neonate: A Case Report

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Abstract

Congenital hiatal hernia is a rare condition in neonates, particularly the mixed paraesophageal form (type III). Clinical presentation is often atypical and may be dominated by respiratory symptoms, which can delay diagnosis. We report the case of a 14-day-old premature neonate hospitalized for recurrent episodes of cyanosis. Thoracoabdominal computed tomography revealed a congenital mixed type III hiatal hernia with a right basal intrathoracic stomach associated with combined organoaxial and mesenteroaxial rotation, without signs of gastric ischemia. The hernia sac exerted a mass effect with anterior displacement of cardiovascular structures and was complicated by right lower lobe atelectasis and aspiration pneumonia. The presence of blood in the nasogastric aspirate raised suspicion of gastric volvulus and prompted early surgical management. The patient underwent surgical repair with hiatal closure and Nissen fundoplication. Postoperative recovery was favorable, with complete resolution of cyanotic episodes and sustained respiratory stability.

Keywords: congenital hiatal hernia; mixed hiatal hernia; neonate; cyanosis; gastric volvulus; Nissen fundoplication

Introduction

Congenital hiatal hernia (CHH) is a rare congenital anomaly resulting from defective formation of the esophageal hiatus and abnormal fixation of the stomach. It is uncommon in neonates and infants [1]. Mixed hiatal hernias (type III), characterized by herniation of both the gastroesophageal junction and the stomach into the thoracic cavity, are considered the most severe forms due to the high risk of gastric volvulus, strangulation, and cardiorespiratory distress [2,3].

In neonates, clinical manifestations are often nonspecific and misleading. Respiratory symptoms such as cyanosis, apnea, or recurrent pulmonary infections may predominate, sometimes in the absence of significant gastrointestinal signs [4–6]. Early diagnosis and prompt surgical management are essential to prevent potentially life-threatening complications.

Case Presentation

A 14-day-old male neonate was admitted to our pediatric surgery department for recurrent episodes of cyanosis. He was born prematurely at 36 weeks of gestation after a monitored pregnancy, delivered vaginally with an Apgar score of 8/10 and a birth weight of 2.5 kg.

Clinical examination revealed intermittent cyanosis, anteriorly positioned anus (figure 1), and convex feet (figure 2). Cardiovascular examination was normal. Transthoracic echocardiography excluded congenital heart disease. Abdominal ultrasound showed no abnormalities.



Figure 1: anteriorly positioned anus Figure 2 : convex fee

A chest x-ray was performed, revealing an intrathoracic air-fluid level, suggesting a hiatal hernia (figure 3).



Figure 3: a chest x-ray with a nasogastric tube in place

Thoracoabdominal computed tomography demonstrated a congenital mixed type III hiatal hernia with a right basal intrathoracic stomach, associated with combined organoaxial and mesenteroaxial gastric rotation, without evidence of gastric wall ischemia or pathological distension (figure 4). The hernia sac exerted a mass effect with anterior displacement of cardiovascular structures and was associated with right lower lobe atelectasis and aspiration pneumonia.

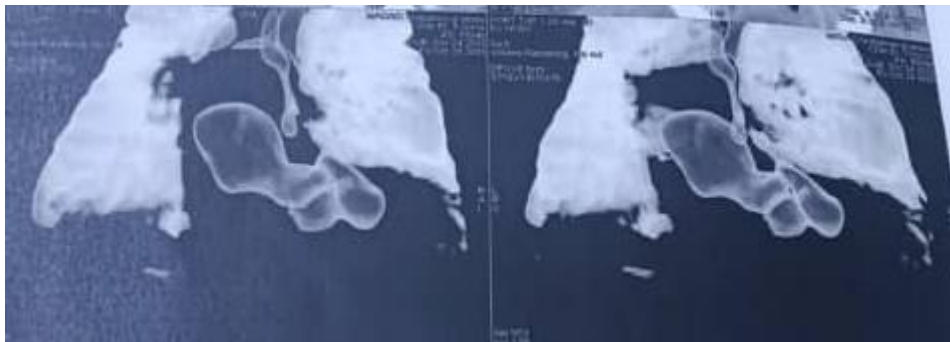


Figure 4: chest CT scan

Surgical Management and Outcomes:

After respiratory stabilization, surgical exploration was performed. Intraoperatively, the stomach was confirmed to be located within the thoracic cavity; however, no gastric volvulus or ischemia was observed (figure : 5 , 6), suggesting an intermittent or reducible torsion.



Figure 5 : stomach without signs of volvulus

Figure 6: hiatal hernia opening

Surgical treatment consisted of reduction of the stomach into the abdominal cavity, repair of the esophageal hiatus (figure: 7), and an anti-reflux Nissen fundoplication.



Figure 7: Repair of the esophageal hiatus

Postoperative recovery was uneventful. Clinical evolution was marked by complete resolution of cyanotic episodes, sustained respiratory stability, and good tolerance of enteral feeding.

Discussion

Congenital hiatal hernia is rare in neonates, with mixed type III and IV hernias representing the most severe forms [1,2]. Large paraesophageal and mixed hiatal hernias are associated with significant morbidity due to digestive and respiratory complications, particularly in infants [7].

In neonates, respiratory manifestations are frequently the primary mode of presentation. Cyanosis, apnea, recurrent pneumonia, and aspiration are explained by mediastinal compression caused by the presence of the stomach within the thoracic cavity, displacement of cardiopulmonary structures, and severe gastroesophageal reflux [4–7]. This explains why gastrointestinal symptoms may be minimal or absent, leading to delayed diagnosis.

In our case, the decision to proceed with surgical repair during the neonatal period was motivated by the symptomatic nature of the disease and the risk of potentially fatal complications. The patient presented with recurrent episodes of cyanosis and aspiration pneumonia, alarming manifestations of congenital mixed hiatal hernia [4,8].

In addition, the presence of blood in the gastric aspirate suggested possible mucosal ischemia and intermittent gastric volvulus, a recognized surgical emergency due to the risk of strangulation, perforation, and sudden deterioration [3,8]. Although no fixed volvulus was identified intraoperatively, the possibility of intermittent or spontaneously reducible torsion cannot be excluded and has been well described in cases of large hiatal hernias in children [7,8].

Given the unpredictable course of congenital paraesophageal and mixed hiatal hernias and the poor correlation between clinical severity and imaging findings, most authors recommend early surgical management once the diagnosis is established, even in neonates [2,7,9].

The surgical principles include reduction of the herniated abdominal contents, closure of the esophageal hiatus, and prevention of gastroesophageal reflux [7, 10–12]. Several authors recommend the systematic addition of an anti-reflux procedure due to the high frequency of severe reflux and its respiratory consequences.

Nissen fundoplication remains the most commonly used technique in pediatric patients and has demonstrated favorable short- and long-term outcomes. In our patient, surgical repair resulted in complete resolution of cyanotic episodes and stabilization of respiratory function, confirming the benefit of early intervention.

Conclusion

Congenital mixed type III hiatal hernia is a rare but potentially life-threatening condition in neonates. It may present with isolated respiratory symptoms, such as recurrent cyanosis, with minimal gastrointestinal signs. The presence of blood in gastric secretions should raise suspicion of gastric volvulus and justify urgent

surgical management. Early diagnosis and surgical repair, including hiatal closure and Nissen fundoplication, provide a favorable prognosis and prevent severe complications.

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