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Preterm Identical Twins with Hiatal Hernias

Keywords: Hiatal hernia; Neonatology; Familial; Pediatric surgery; Preterm infants

Abstract

Hiatal hernias are usually sporadic and uncommon, requiring a high index of suspicion. Preterm monochorionic-monoamniotic twins with recurrent emesis had inconclusive radiographs with contrast study consistent with hiatal hernia. Genetic testing was performed due to concerns for familial inheritance. Early diagnosis and treatment of hiatal hernias are essential to prevent complications.

Open Access Case Report

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Introduction

Hiatal hernia (HH) is a type of diaphragmatic hernia in which abdominal organs protrude through the esophageal hiatus of the diaphragm into the posterior mediastinum [1]. There are four types of hiatal hernias, depending on the location of the gastroesophageal (GE) junction in relation to the diaphragm [1]. Type I, the most frequent, constituting 85-95% of hiatal hernias, is an axial hernia, where the gastric cardia slides into the chest cavity. Type II is a paraoesophageal hernia characterized by normal positioning of the GE junction while the hernia sac contains the gastric fornix. Type III is a combination of Types I and II, in which more than 50% of the stomach is herniated into the mediastinum. Type IV is characterized by the herniation of the stomach along with other abdominal organs into the mediastinum.

The incidence of hiatal hernias increases with age [2]; Hence, they are more common in the adult population and are relatively uncommon among the pediatric population [3,4]. HH are usually sporadic cases, but there have been reported cases of familial presentations [3-6].

Early diagnosis and surgical treatment of HH is essential, as there is risk for severe reflux, strangulation, Barrett's esophagus, chronic gastritis, lower esophageal stenosis, esophageal ruptures, recurrent aspiration causing respiratory tract infections and volvulus [1,7].

We present a case of monochorionic monoamniotic twins diagnosed with HH after weeks of unrelenting gastroesophageal reflux and coffee ground colored emesis.

Case Report

Monochorionic-monoamniotic female twins were born at 31 weeks gestation via cesareansection secondary to maternal preeclampsia complicated by cord entanglement. Both twins required continuous positive airway pressure due to respiratory distress syndrome. A few weeks after birth, both twins were noted to have inguinal hernias and one twin had an umbilical hernia. At around 4 weeks of age, both twins had persistent coffee -ground colored, large volume emesis resulting in multiple feeding adjustments. Radiographs were only significant for occasional mildly dilated bowel loops, until a lucencywas noted on anteroposterior and lateral films (Figure 1), prompting the diagnosis of HH. Concomitantly, there was recent difficulty in advancing Twin B's nasogastric tube into the stomach (Figure 1). Twin B underwent upper GI series with contrast, which revealed a sliding HH (Figure 2). Abdominal radiographs of the other twin also showed the same lucency, and upper GI contrast study also confirmed HH.

The family reported several family members with hernia requiring surgical repair. The father had an umbilical hernia repaired as an infant and a maternal uncle with an unknown hernia underwent repaired at age 6. Furthermore, the twins' mother had history of reflux as a child. To rule out connective tissue disorder, a comprehensive [Invitae] genetic panel of 92 genes was done on both twins and was found to be normal.

Twin A underwent surgery for repair of the paraoesophageal hernia, Nissen fundoplication, repair of umbilical hernia and bilateral inguinal hernias and G tube placement due to poor oral motor function. However, during surgery, a posterior esophageal hiatal defect was found, as well as an omphalomesenteric duct remnant with small intestine passing through it, concerning for an internal

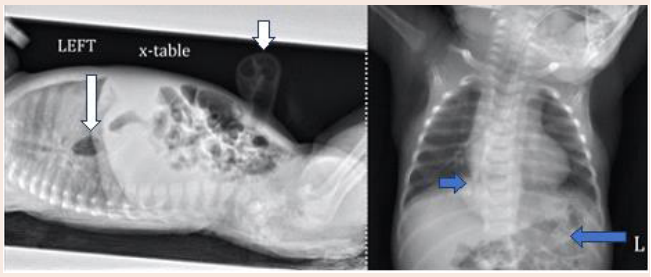

Figure 1: Cross table radiograph (left) showing large umbilical hernia (small white arrow) and hiatal hernial above the diaphragm (long white arrow). AP radiograph (right) shows two nasogastric tubes with one entering the herniated part of the stomach (small blue arrow) and the other in the correct placement (long blue arrow).

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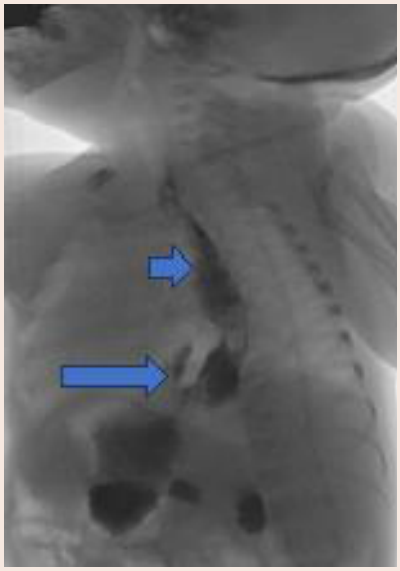

Figure 2: Upper GI contrast series showing large sliding hiatal hernia (long arrow) and severe reflux (short arrow).

hernia. The omphalomesenteric duct was excised with reduction of the internal hernia, and the posterior esophageal hiatal hernia was reapproximated. Twin B underwent paraoesophageal hernia repair and Nissen fundoplication, as well as bilateral inguinal hernia repair. After surgery, symptoms improved significantly.

Discussion

We presented monochorionic monoamniotic twins with persistent coffee-ground colored emesis, with suspicious radiographs, and later confirmed to have HH by upper GI contrast studies. Due to multiple hernias and strong family history of hernias, genetic testing was done to rule out connective tissue disorder.

A congenital HH is an uncommon diagnosis in neonates [4,8,9]. The pathogenesis of congenital HH is poorly understood, but it is thought to be due to a developmental abnormality or genetic predisposition [8]. Connective tissue disorder panel was normal, but this was specific testing for only a subset of genes which does not rule out genetic etiology. The literature is consistent with a familial predilection for congenital hiatal hernias [4,10]. Congenital HH have been reported in patients with identified genetic mutations, such as Cornelia de Lange syndrome and duplication in 9q22.31q22.32 [8,11].

The diagnosis of a HH requires a high index of suspicion [9]. Although HH can be diagnosed using ultrasound [9], contrast swallow study is the most conclusive diagnostic modality [12]. It is important to diagnose and surgically treat HH to prevent complications [1,7].

Conclusion

HH are rare in neonates and require a high index of suspicion in the context of persistent emesis. While radiographs and ultrasound may be helpful, a radio contrast upper GI study is the gold standard. Early surgical repair with fundoplication will prevent Long-term complications. Our experience shows that although rare, HH should be considered when a neonate develops severe reflux, especially if other hernias are present.

Acknowledgement

All authors declare no conflicts of interest.

The parents of the children in question are aware of this case report and have given their consent.

References

1. Babuci S, Gorbatiuc O, Jalbă A, Eremia V, Negru I, et al. (2020) Type III Congenital Paraoesophageal Hiatal Hernia- A Rarity in Pediatric Surgery. A Clinical Case and Literature Review. JURNALUL PEDIATRULUI 23: 41-47.
2. Pawluszewicz P, Wojciak P, Diemieszczyk I, Golaszewski P, Wozniewska P, et al. (2018) Przepukliny przeponowe - epidemiologia, patogeneza, diagnostyka. Postępy Nauk Med 31: 274-278.
3. Kakaje A, Al Khoury F, Alfarroukh A, Khouri L (2019) Familial sliding hiatus hernia in four siblings with uncommon features: a case series report. J Surg Case Reports [Internet] 2019: 1-3.
4. Baglaj SM, Noblett HR (1999) Paraoesophageal hernia in children: familial occurrence and review of the literature. Pediatr Surg Int 15: 85-87.
5. Goodman RM, Wooley CF, Ruppert RD, Freimanis AK (1999) A possible genetic role in esophageal hiatus hernia. J Hered 60: 71-74.
6. Carré IJ, Johnston BT, Thomas PS, Morrison PJ (1999) Familial hiatal hernia in a large five generation family confirming true autosomal dominant inheritance. Gut 45: 649-652.
7. Garvey EM, Ostlie DJ (2017) Hiatal and paraesophageal hernia repair in pediatric patients. Semin Pediatr Surg 26: 61-66.
8. Chang CA, Di Donato N, Hackmann K, Argiropoulos B, Ferreira P, et al. (2020) Congenital hiatal hernia segregating with a duplication in 9q22.31q22.32 in two families. Am J Med Genet A 182: 3040-3047.
9. Sharba L, Al-Hakkak M, Al-Faham F, Abdul Hussein J (2020) Surgical management of congenital hiatal hernia: A retrospective cohort study, single-center experience. Int J Pharm Res 12: 3740-3748.
10. Sidd JJ, Gilliam JI, Bushueff BP (2014) Sliding Hiatus Hernias in Identical Twins. Br J Radiol [Internet]. 39: 703-704.
11. McDowell BC, Horst KK, Klinkner DB (2022) Congenital paraesophageal hernia with gastric outlet obstruction in a neonate with Cornelia de Lange Syndrome. Radiol Case Reports. 17: 1478-1482.
12. Joghee S (2020) Hiatal Hernia Presenting as Difficulty in Inserting Feeding Tube in a Neonate Born Preterm. J Pediatr 218: 256.