CASE REPORT

Paraesophageal hiatus hernia in an 8-month-old infant with organoaxial volvulus of the stomach

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SUMMARY

Paraesophageal hiatus hernia is an extremely rare variety of hiatus hernia and it is rarely seen in the paediatric age group. We present an instance of this hernia in which an 8-month-old female infant was admitted with a history of ongoing vomiting and irritability since past 7 days. The subsequent imaging study showed paraesophageal hiatus hernia with twisting of the stomach along its longitudinal axis (organoaxial volvulus). Subsequent surgical repair led to the resolution of symptoms. This kind of volvulus is rare in hiatus hernia and once detected it requires prompt surgical management. Given the asymptomatic presentation of hiatus hernia clinicians need to realise that this can be associated with volvulus of stomach which, if left unrepaired, can lead to severe complications. Thus, prompt consideration is necessary with timely surgical referral to alleviate this rare but important variety of hiatus hernia.

BACKGROUND

Hiatus hernia is an uncommon presentation in the paediatric age group as most of the cases are due to acquired age-related laxity of structures supporting the gastro-oesophageal junction (GEJ). Paraesophageal hernia only represents 5% of this group. Initial symptoms are often mistaken with gastro-oesophageal reflux disease. Presentation with volvulus of the stomach in an infant has rarely been reported in medical literature.

CASE PRESENTATION

An 8-month-old female infant attended the paediatric emergency department (ED) with a history of ongoing vomiting and irritability since past 7 days. Her vomitus was noted to be coffee ground in colour at one occasion before coming to ED. She also had mild cough along with reduced oral intake in the past few days. She was born by normal vaginal delivery at full term with a birth weight of 3300 g. She never attended the ED before and was exclusively breastfed until 6 months of age. She had started tolerating solid feeds recently. There had been no sick contacts in the family recently and her vaccinations were up to date. She had no known drug allergy and her development was appropriate for the age.

On examination she was afebrile, irritable and mildly lethargic. Vital parameters were within normal limit and systemic examination was normal. Her growth centiles were appropriate for her age. She was admitted for observation and intravenous fluid support. Her full blood counts, urea,

Figure 1 Chest X-ray (PA) showing large gas shadow behind the heart.

creatinine, electrolytes and coagulation parameters were within normal limits (see investigation for values). Subsequent worsening of her vomiting episodes in the ward prompted us to perform a chest X-ray which revealed a huge gas shadow behind the heart along with a small area of right basal atelectasis (figure 1). Suspicion was raised of diaphragmatic hernia. Barium meal follow through showed intrathoracic hold up of barium with tertiary contractions above the stomach. The relative position of gastric inlet and outlet suggested a degree of rotation of the stomach along its longitudinal axis giving rise to rare variety of organoaxial volvulus of the stomach due to paraesophageal hiatus hernia (figure 2). Rest of the barium passage through the gut were normal.

She underwent laparoscopic hiatal hernia repair after initial stabilisation. Postoperatively she developed transient gastroparesis, which resolved with short-term oral erythromycin supplement as a prokinetic agent.

INVESTIGATIONS

Blood tests revealed a white cell count of $8.7 \times 10^9/L$ (neutrophils $4 \times 10^9/L$, lymphocytes $3.5 \times 10^9/L$); haemoglobin 13.5 g/dL; platelets $252 \times 10^9/L$ (140–450); urea 5.34×10^9 mmol/L (2.60–7.30); creatinine 36 mmol/L (17–44); sodium 138 mmol/L (136–145); potassium 4.7 mmol/L (3.5–5); chloride

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Figure 2 Barium meal follow through showing herniated stomach in the thoracic cavity, which seems to rotate along its longitudinal axis.

100 mmol/L (98–107); prothrombin time 11 s (9.5–12.7); activated partial thromboplastin time 30.5 s (25.7–38.8) and an International Normalised Ratio of 1.0 (0.9–1.2).

DIFFERENTIAL DIAGNOSIS

- ▶ Gastro-oesophageal reflux disease
- ▶ Cow milk's protein intolerance
- ▶ Viral gastritis

OUTCOME AND FOLLOW-UP

The patient is doing well at 4 months follow-up.

DISCUSSION

Hiatus hernia is an uncommon presentation in the paediatric age group as most types are acquired except some congenital ones. Hiatus hernia is either sliding type (95%) or paraesophageal type which comprises only 5% of the total group. In sliding hernia GEJ moves upward to an intrathoracic position whereas in paraesophageal hernias part of the stomach moves into the thorax by the side of GEJ and the position of GEJ remains fixed containing the herniated stomach sac into the posterior mediastinum.¹ This can occur due to acquired laxity of the supporting structural elements around GEJ as age advances.² In rare instances it has been noted in conjunction with congenital defects of the ligaments supporting GEJ leading to its presentation in early age groups as in our case. Attenuated and/or deficient fixation of supporting gastric ligaments around GEJ can lead to abnormal mobility of the stomach with potential for gastric volvulus if hiatal hernia is present.³

Most of these hernias remain asymptomatic. Usual presentations are with symptoms of gastro-oesophageal reflux disease such as vomiting after feeds, heart burn and retrosternal chest pain. Our patient presented with irritability and it could be the effect of retrosternal burning sensation.

Chest X-ray can be normal. Thoracoabdominal CT scan is very helpful in diagnosing paraesophageal hernia as it has the potential to show all the anatomical details around the diaphragm.² Most often suspicion is not raised of paraesophageal hernia and CT scan is not ordered initially. Upper gastrointestinal series such as barium swallow and follow through is essential in confirming paraesophageal hiatus hernia. Barium swallow can also depict the position of herniated fundus of the stomach in relation to its either longitudinal or transverse axis, as

detection of volvulus of stomach requires emergent surgical correction.^{4–6} Herniated fundus of stomach can twist around its own longitudinal axis or along the axis of mesentery giving rise to organoaxial or mesentericoaxial volvulus of the stomach, respectively. Both of these are very rare presentations. Infants usually present with irritability due to retrosternal discomfort and is often not thought of, as irritability is assigned to other causes mainly meningitis. Insight of this important causation is very important in clinicians as delay in diagnosis may put herniated fundus of stomach at the risk of progressive enlargement leading to higher complication rates.¹

Treatment is either laparoscopic repair or open surgical repair of paraesophageal hiatus hernia. Sliding hiatus hernia can be left as such until complications occurs as mortality from elective repair (1.4%) is not much higher than chances of developing acute symptoms requiring emergent surgery (1.1%).⁶ However paraesophageal hiatus hernia always mandates surgical repair in the paediatric age group. Recently, improved techniques of laparoscopic repair have decreased complications and recurrence rates.⁷

Keeping in view the rarity of paraesophageal hernia in the paediatric age group and its subtle presentation that can be easily misdiagnosed as other non-surgical conditions, it is important that clinicians bear in their mind, the possibility of this important surgical entity as some of it may need emergent surgical repair. There are very few case reports highlighting these important facts in medical literature.

Learning points

- ▶ Paraesophageal hiatus hernia is an old age disease and is extremely rare in paediatric patients.
- ▶ Initial symptoms are misdiagnosed as gastro-oesophageal reflux disease.
- ▶ If left untreated, herniated gastric sac can undergo twisting producing volvulus that requires urgent surgical repair with high complication rates.
- ▶ Prompt consideration of this entity may help diagnosing this rare condition in infants who does not respond well to adequate medical management of gastro-oesophageal reflux disease.

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Competing interests None.

Patient consent Obtained.

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