Hypertrophic pyloric stenosis, an unusual presentation and rare association: case report and literature review

Majd A. Hadad and Mohammad S. Mallick

Hypertrophic pyloric stenosis (HPS) is a commonly encountered surgical condition that occurs in infancy, but not in neonates. This report describes a 10-day-old male patient with atypical presentation of HPS, with an incidental diagnosis of eventration of the left diaphragm. The symptoms were present since birth with nonprojectile, nonbilious vomiting and gastric distension. A contrast study showed a gastric outlet obstruction with the possibility of gastric volvulus. Emergency surgery established the diagnosis. To the best of our knowledge, the association of HPS with diaphragmatic defect and gastric volvulus is rare, and few cases have been reported in the literature. *Ann Pediatr Surg* **10:133–135 © 2014 Annals of Pediatric Surgery.**

Annals of Pediatric Surgery 2014, **10**:133–135

Keywords: gastric volvulus, hypertrophic pyloric stenosis, vomiting

King Fahad Medical City, Riyadh, Saudi Arabia

Correspondence to Majd A. Hadad, MS, MRCS, Surgery Department, Division of Pediatric Surgery, King Fahad Medical City, PO Box 365480, Riyadh 11393, Saudi Arabia
Tel: +966 557 915 996; fax: +966 288 9999 17358;
e-mails: hadad3majd@yahoo.com, mhadad@kfmc.med.sa

Received 14 March 2012 **accepted** 26 May 2014

Introduction

Hypertrophic pyloric stenosis (HPS) occurs at a rate of one to four per 1000 live births in White infants but is seen less in non-White children. Male infants are affected more often with a 4 : 1 male to female ratio. Risk factors for HPS include family history, sex, younger maternal age, being a first-born infant, and maternal feeding patterns [1]. It is caused by hypertrophy of the circular muscular layer of the pylorus, resulting in elongation and thickening of the pylorus and may progress to complete obstruction of the gastric outlet. Pyloric stenosis usually appears at 2–8 weeks after birth and the mean age at diagnosis is 3 weeks [2]. Huang *et al*. [3] found that only 8.41% of patients were diagnosed before 3 weeks of age in a study on 214 infants with HPS, confirming that neonatal HPS is rare but does exist. The group diagnosed before 3 weeks of age did not demonstrate any statistical difference in terms of the male sex predilection, positive family history, and frequency of coexisting malformation, and showed short timeframe of diagnosis with longer duration of hospital stay.

HPS has been found to be associated with certain malformations such as malrotation [4], chronic gastric volvulus [5,6], diaphragmatic eventration [5], diaphragmatic hernia [7], genitourinary tract abnormalities, and esophageal atresia [4,7].

Case report

A 10-day-old infant presented with a history of nonprojectile, nonbilious milky vomiting since birth. He was born full term with normal Apgar score and weight of 3.5 kg.

The antenatal ultrasound examination was normal. Patient examination revealed mild dehydration, soft lax abdomen, no palpable mass, and normal male genitalia.

Laboratory investigation showed metabolic alkalosis (pH 7.50, PCO_2 42.6 mmHg, HCO_3 32.7 mmol/l) and electrolytes (sodium 139 mmol/l, potassium 5 mmol/l, chlorine 96 mmol/l). The plain chest and abdominal radiography showed a mediastinal shift to the right side, elevation of the left hemidiaphragm, stomach distention, and the nasogastric tube coiled up below the left diaphragm (Fig. 1). Upper gastrointestinal (UGI) contrast study showed the stomach distended, rotated, and laying below the elevated left diaphragm with complete gastric outlet obstruction (Fig. 2). A diagnosis of gastric volvulus with eventration of the left diaphragm was made. The patient was taken to the operative room and laparotomy was performed.

The intraoperative findings were distended, thick walled stomach, no gastric volvulus, mild eventration of the left diaphragm, and pyloric olive (Fig. 3). Pyloromyotomy was performed.

An intraoperative fluoroscopy with installation of Omnipaque contrast (GE Healthcare, USA) through the nasogastric tube showed the passage of the contrast from the stomach to the duodenum and upper jejunum, and the possibility of other associated anomalies such as intestinal malrotation was ruled out.

The eventration of the left diaphragm was left alone for conservative management.

The postoperative period was uneventful. The patient was discharged on third postoperative day. After 6 months of follow-up, patient was doing well with no vomiting and gain of weight.

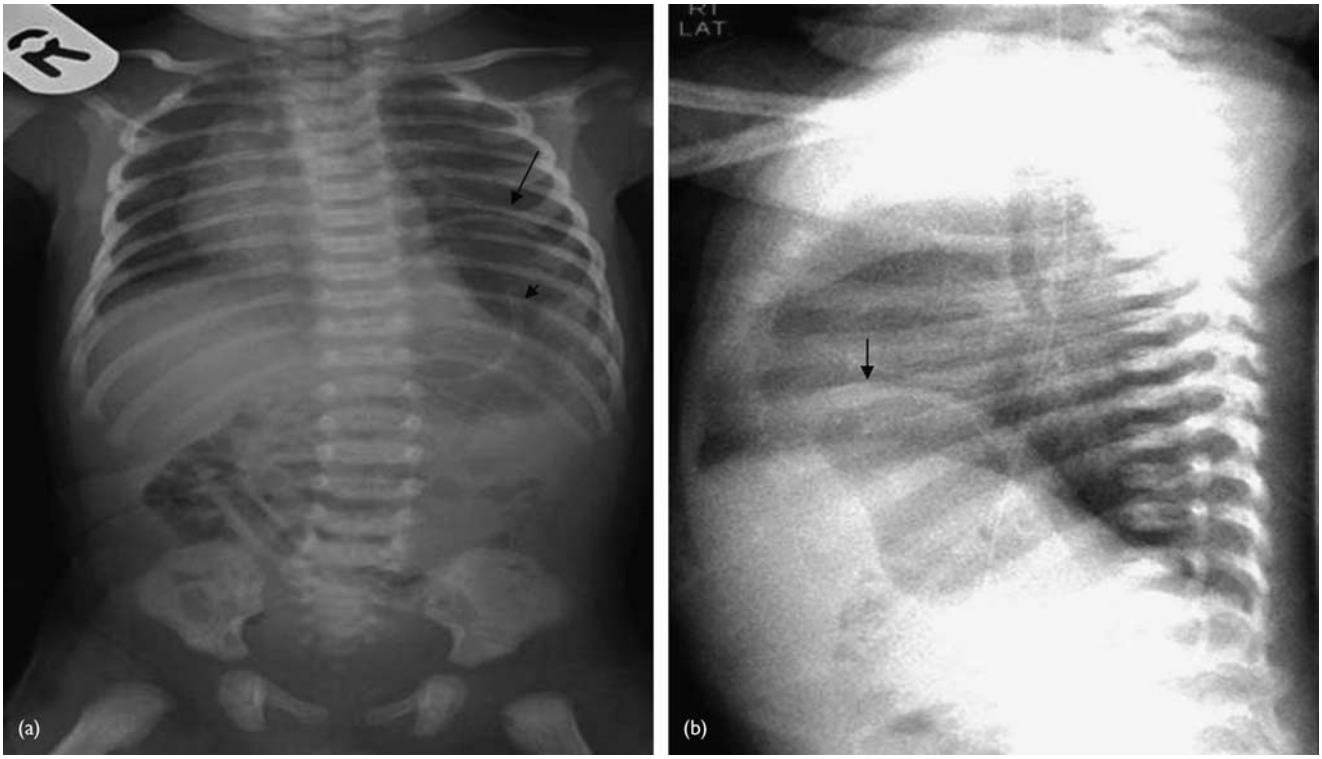
Discussion

HPS usually appears at 2–8 weeks of life as nonbilious projectile emesis. The mean age at diagnosis is 3 weeks of age. The emesis is progressive and can lead to gastritis [2].

The initial emesis may appear to be reflex and, with progress of the disease, becoming forceful [1].

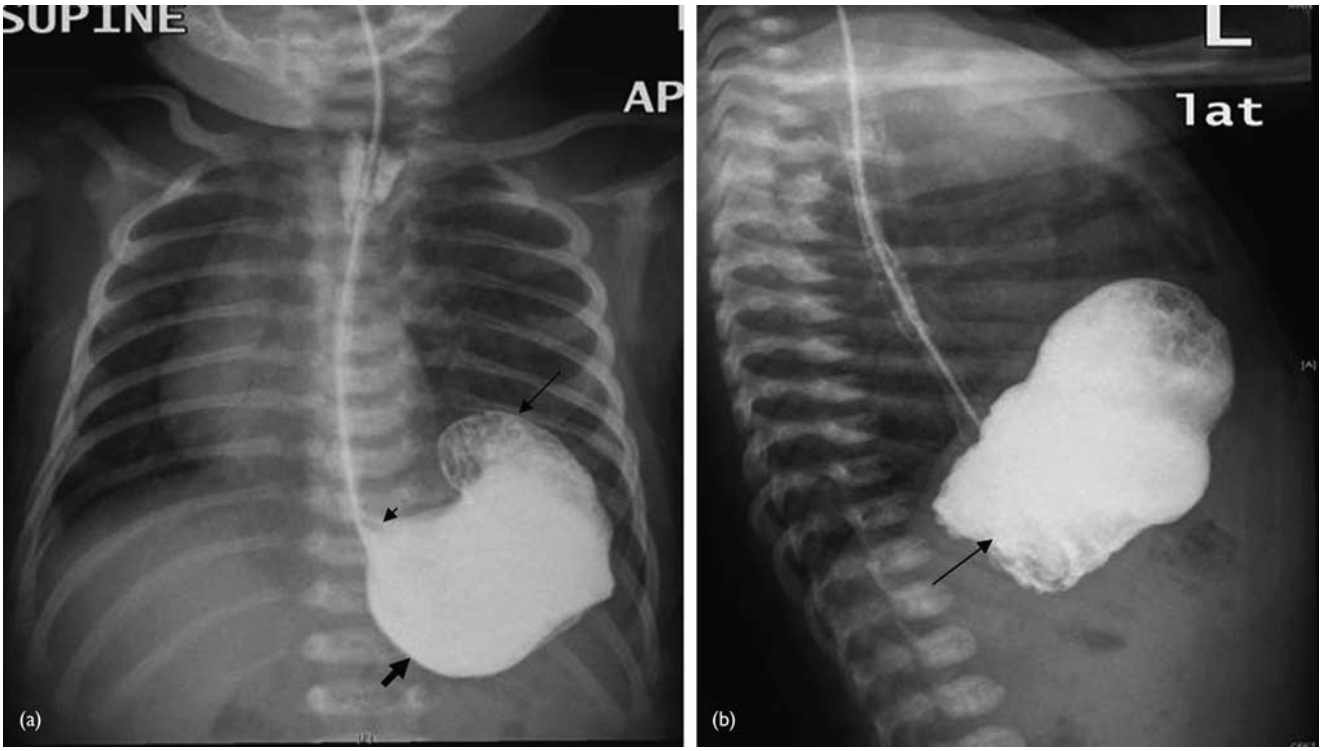
1687-4137 © 2014 Annals of Pediatric Surgery DOI: 10.1097/01.XPS.0000452063.38321.b1

Fig. 1



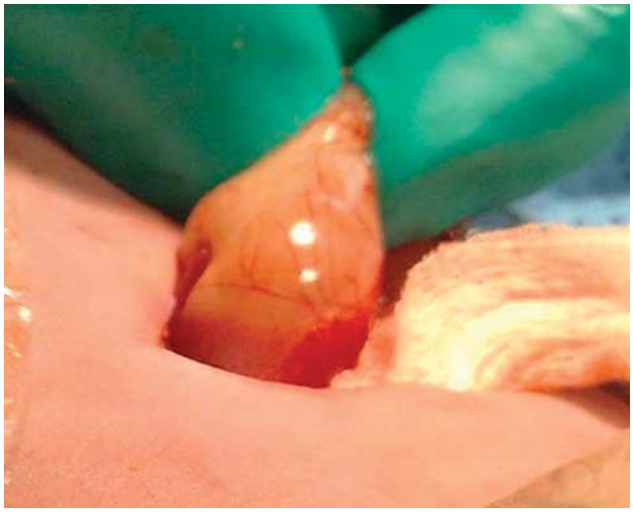
(a) Plain chest and abdomen radiography in the supine position shows left diaphragm elevation (arrow). Reaching the seventh rib with distended stomach, the nasogastric tube (arrow head) coiled and seen superimposed, over the lower left lung, the mediastinum shifted to the right side and normal gas distribution in bowel. (b) Plain chest in the left lateral position shows that the left diaphragm is intact and elevated more than the right side (arrow).

Fig. 2



(a and b) Upper gastrointestinal study series shows gastroesophageal junction (arrow head) is in a normal position, the fundus (wide arrow) is positioned below the pylorus, the pylorus is in the left upper abdomen with complete cutoff gastric outlet (thin arrow).

Fig. 3



Pyloric olive.

In our case, the vomiting was nonprojectile that can be explained by early presentation.

The bilious emesis does not rule out HPS. It reported to be present in 1.4% of the affected infants and hematemesis because of gastritis present in 5% of the infants with HPS [8].

Diagnosis is made typically by history and physical examination augmented by radiographic imaging such as ultrasound or UGI contrast study [1,2].

The previously described 'classical' features of HPS such as hypochloremic, hypokalemia, metabolic alkalosis, palpable pyloric mass, and emaciation are now reported less frequently.

The reason for this change is likely due to HPS being diagnosed earlier because of the use of ultrasound [8,9]. The association of HPS with chronic gastric volvulus and diaphragmatic eventration has been reported [5,6]. In addition, the association of diaphragmatic hernia with gastric volvulus has been reported [10,11]. Kotobi *et al.* found congenital diaphragmatic hernia in 65% of the children with acute gastric volvulus, and 84% of those aged less than 1 month [10].

Generally, gastric distention, regardless of the cause, can cause rotation of the stomach in neonates and infants with lax or immature ligaments, with the possibility of gastric volvulus increased in patients with abnormal large amounts of subdiaphragmatic space [5]. Because the main symptoms of chronic gastric volvulus are recurrent projectile vomiting, abdominal distention, and failure to thrive overlap those of HPS, the differential diagnosis can be difficult [5]. In our case, the patient presented earlier, at 10 days old with nonprojectile milky vomiting since birth. The coexistence left diaphragmatic eventration with gastric outlet obstruction and abnormal position and configuration of the stomach, which masked the HPS and raised the diagnosis of acute gastric volvulus that required emergency surgical exploration.

Although the association between HPS and other anomaly is rare, it should be kept in mind when facing similar conditions.

Conclusion

We report a 10-day-old boy who presented with nonprojectile milky vomiting since birth. Radiological study showed stomach rotation and lying below the eventrated left diaphragm with gastric outlet obstruction, which raises a range of differential diagnosis at the top gastric volvulus, HPS, intestinal malrotation, and pyloric web. The diagnosis of HPS was established during surgery. We recommend intaoperative UGI in such cases to rule out unexpected associated other anomalies.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Curt SK, Mark W. Lesions of the stomach. In: George W, Patrick J, editors. *Ashcraft's pediatric surgery*. 5th ed. USA: Elsevier Inc.; 2010. pp. 391–395.
2 Ward E, Easley D, Pohl J. Previously unsuspected infantile hypertrophic pyloric stenosis diagnosed by endoscopy. *Dig Dis Sci* 2008; **53**: 946–948.
3 Huang IF, Tiao MM, Chiou CC, Shih HH, Hu HH, Ruiz JP. Infantile hypertrophic pyloric stenosis before 3 weeks of age in infants and preterm babies. *Pediatr Int* 2011; **53**:18–23.
4 Pandey A, Gangopadhyay AN, Upadhyay VD. Malrotation associated with infantile hypertrophic pyloric stenosis in a neonate. *J Paediatr Child Health* 2009; **45**:71–74.
5 Anagnostara A, Koumanidou C, Vakaki M, Manoli E, Kakavakis K. Chronic gastric volvulus and hypertrophic pyloric stenosis in an infant. *J Clin Ultrasound* 2003; **31**:383–386.
6 Oğuzkurt P, Senocak ME, Hiçsönmez A. A rare coexistence of two gastric outlet obstructive lesions: infantile hypertrophic pyloric stenosis and organoaxial gastric volvulus. *Turk J Pediatr* 2000; **42**:87–89.
7 Velaoras K, Bitsori M, Galanakis E, Charissis G. Hypertrophic pyloric stenosis in twins: same genes or same environments? *Pediatr Surg Int* 2005; **21**:669–671.
8 Piroutek MJ, Brown L, Thorp AW. Bilious vomiting does not rule out infantile hypertrophic pyloric stenosis. *Clin Pediatr (Phila)* 2012; **51**:214–218.
9 Glatstein M, Carbell G, Boddu SK, Bernardini A, Scolnik D. The changing clinical presentation of hypertrophic pyloric stenosis: the experience of a large, tertiary care pediatric hospital. *Clin Pediatr (Phila)* 2011; **50**:192–195.
10 Kotobi H, Auber F, Otta E, Meyer N, Audry G, Hélardot PG. Acute mesenteroaxial gastric volvulus and congenital diaphragmatic hernia. *Pediatr Surg Int* 2005; **21**:674–676.
11 Karabulut R, Türkyilmaz Z, Sönmez K, Karakus SC, Basaklar AC. Delayed presentation of congenital diaphragmatic hernia with intrathoracic gastric volvulus. *World J Pediatr* 2009; **5**:226–228.