Intra-thoracic hypertrophic pyloric stenosis with late onset presentation: a case report

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ABSTRACT

Background: Hypertrophic pyloric stenosis (HPS) is a common cause of gastrointestinal obstruction in childhood.

Case Presentation: We report a 3-month-old male child with vomiting and weakness that had been started 3 days before being referred to the hospital. We used ultrasonography to examine this patient and, while the results from the laboratory tests were normal; abdominal ultrasound examination showed HPS with hiatus hernia and abnormal mesenteric vessels pathway.

Conclusion: We concluded that HPS with presentation in thorax is a rare condition that can be diagnosed with meticulous ultrasound scanning of diaphragmatic hiatus, pyloric canal, and mesenteric vessels pathway.

Keywords: Hypertrophic pyloric stenosis, hiatus hernia, infant, ultrasound.

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Background

Hypertrophic pyloric stenosis (HPS) is an inherent condition in which failure in relaxation of the prepyloric antrum leads to prolonged pylorospasm, it is associated with elongation of the pyloric canal and circumferential thickening of the pyloric muscle [1]. This condition usually presents between 2 and 10 weeks after birth, and is associated with symptoms like projectile vomiting and metabolic disorders [2]. The incidence of this disorder is 0.9–5.1 per 1,000 cases and presents with non-bilious projectile vomiting [3]. The cure for this disorder is pyloromyotomy.

Although etiology of HPS remains unknown, diagnosis of its clinical manifestation and treatment have improved considerably over the last few decades. The relationship between HPS and partial hiatus hernia is uncommon and a few reports are available in literature [4–5]. We present a 3-month-old baby with late onset HPS with complete intra-thoracic stomach, which was first diagnosed on abdominal ultrasound.

Case Presentation

A 3-month-old male infant presented to the hospital with vomiting, diarrhea, weakness, and fever for 3 days. He was a term vaginal delivery and the first child of the family with the birth weight of 3,400 g, which had increased to 3,800 g at the time of his admission to the hospital. His past medical history was significant for glucose-6-phosphate dehydrogenase (G6PD) deficiency and surgical history was significant for inguinal hernia repair 1 month after his birth. There was no relevant family history. On physical examination, there was a fever of 38°C and a respiratory rate of 48 rpm. All laboratory tests were within normal limits. Chest X-ray showed right para-cardiac lucency indicative of air. Subxiphoid transhepatic abdominal ultrasound showed protrusion of pylorus and stomach from diaphragmatic hiatus into thorax as hiatus hernia.

The dimensions of pyloric canal in diaphragmatic hiatus were significantly abnormal with length of 24 mm and wall thickness of 5.9 mm. In addition, abnormal mesenteric vessels pathways associated with the diaphragmatic hernia were detected by ultrasound scanning. In upper gastrointestinal (GI) series, there was distended stomach in right hemithorax with a delay in transit of barium to small bowel. As ultrasound criteria of HPS were conclusive, we did not look for other radiologic signs of the disease. Surgical management (hiatal hernia repair and myotomy of pyloric muscle) was performed with no complication. The baby thrived afterward, and he showed clinical improvement during his 3-month follow-up examinations.

Discussion

HPS is a disorder of gastric outlet seen in infants with an average age of 5 weeks in which, the abnormal thickness of antropyloric portion of the stomach causes gastric obstruction. The clinical diagnosis depends on palpation of the thickened pylorus called “olive.” Abdominal palpation is not always successful as it depends on many factors such as the experience of the physician, the presence of gastric dilatation, and a quiet infant. When clinical examination is unsuccessful, additional imaging techniques that are highly accurate can be used to facilitate the diagnosis process. Therefore, radiologists have an important role in the initial diagnosis of these disorders in infants and referring them to

surgical treatments [2]. An association between HPS and partial hiatus hernia has not been reported frequently in the past. It was believed by many scientists that the obstruction at the pylorus leads to high intragastric pressure that pushes the stomach towards the chest [5].

In 1960, Johnston through his assessment of a series of 76 cases of hiatus hernia in childhood detected eight cases with HPS (10.5%) [5]. He concluded that gastric emptying disorder results in a functional pyloric obstruction. It was believed that the raised intragastric pressure leads to displacement of the cardia towards the chest [5]. Between 1988 and 1994, Lijima et al. [6] were able to cure three patients of HPS associated with hiatus hernia, and two of them had been diagnosed preoperatively.

Recently, Winckworth et al. found a neonate with iniencephaly clausus, a rare neural tube defect that had HPS with a congenital ultrashort esophagus. The results from ultrasound during pregnancy showed a right-sided thoracic cystic lesion, and further assessments afterbirth helped to detect an intact diaphragm with intra-thoracic stomach and spleen, which then developed into HPS [7].

Ultrasound provides a direct method to determine the thickness of the pyloric muscle and real-time assessments of contraction of the pyloric canal [2]. Using ultrasound, we can assess the pyloric canal morphology and behavior dynamically. In ultrasound evaluation, the criteria which are used to diagnose HPS include pyloric canal length of 14–20 mm, pyloric muscle wall thickness greater than 3 mm, and pylorus muscle length greater than 12 mm. These measurements depend on the patient's age, the quietness of the baby, sonographic technique, and the experience level of the examiner.

Late onset of the presentation of HPS is a rare condition, and the age of presentation in our case was 3 months after the birth. In our patient, the pyloric canal length was 24 mm with the wall thickness of 5.9 mm (Figure 1). Location of pyloric canal was in diaphragmatic hiatus, which was observed with subxiphoid transhepatic ultrasound.

Typical string sign was detected in upper GI series that is a filiform passage of contrast material through the mucosal interstices; by several linear tracts of contrast material separated by the intervening mucosa, called the double-track sign (Figure 2). Figure 2 shows the radiologic findings of malrotation and midgut volvulus including low location of duodenojejunal junction, corkscrew appearance of the duodenum and jejunum, right side position of jejunum, and abnormal position of cecum below transverse colon.

Such features of HPS versus pylorospasm have been identified by Haran et al. [1]. Pylorospasm is the muscle spasm that leads to closure and delayed passage of gastric contents to the duodenum. This condition is usually related to peptic ulcers. Fluoroscopic investigation shows vigorous active peristalsis like a caterpillar that causes an abrupt stop at the pyloric antrum and outlining the external thickened muscle as an extrinsic impression that is called the shoulder sign. In our patient, there was a distended stomach in the right hemithorax with a delayed transit of barium to small bowel. It should be noted that as ultrasound criteria of HPS were conclusive, in order to minimize the radiation exposure to the infant, we did not try to find other radiologic signs of the disease.

Another interesting finding of this case was the abnormal pathway of mesenteric vessels. Normal mesenteric vessels (superior mesenteric artery and superior mesenteric vein) in their proximal parts have a downward direction approximately parallel to abdominal aorta. In this patient, the direction of mesenteric vessels was towards the left anterior side of the abdomen. This abnormal pathway of mesenteric vessels

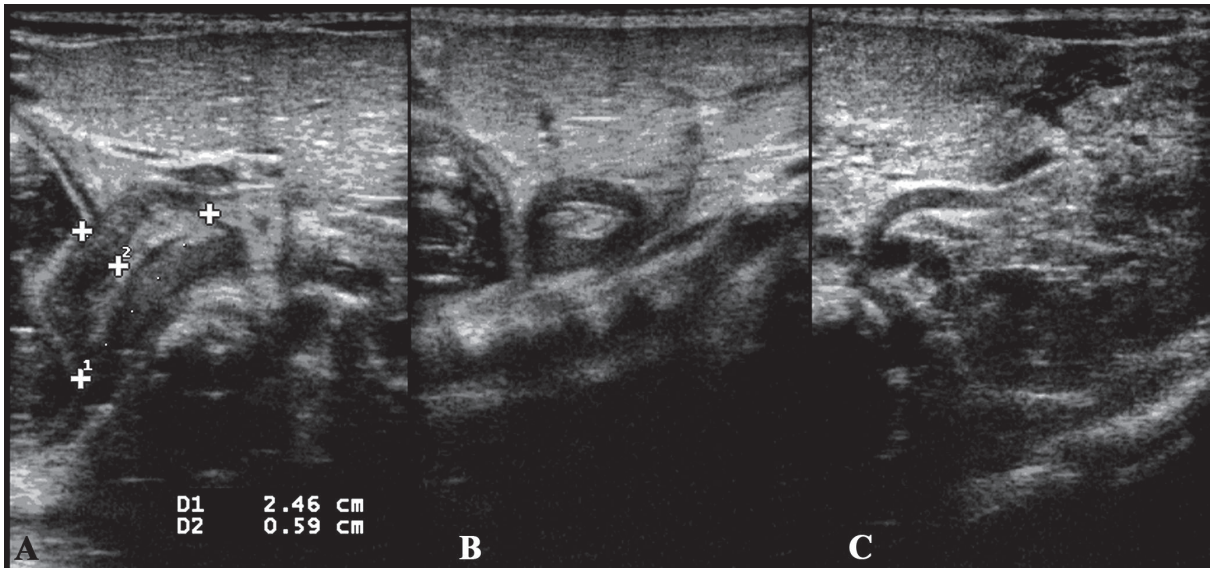

Figure 1. *Gray-scale ultrasonography using sonographic 10 MHz linear probes, (A) the longitudinal view of pyloric canal shows a bulky pylorus with pyloric canal length of 24.6 mm and wall thickening of 5.9 mm, (B) the transverse view of pyloric canal shows a target sign, and (C) the abnormal pathway of mesenteric vessels towards anterior and left upper quadrant due to associated diaphragmatic hernia.*

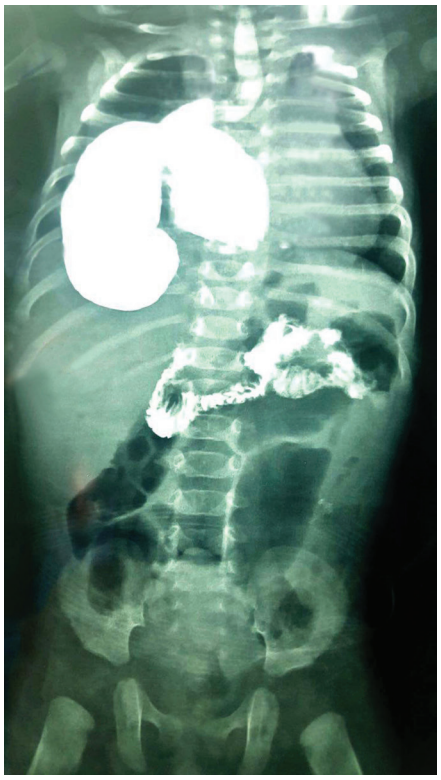

Figure 2. *The thoracic-abdominal-pelvic view of upper GI series shows a hiatal hernia with distended stomach in right hemi-thorax. The C loop of duodenum has normal pattern without honey comb pattern and relatively less gas in abdomen and especially rectum is seen.*

has been observed in association with a number of GI disorders such as midgut volvulus, internal hernia, and Bochdalek hernia [8]. These cases show that the abnormal direction of mesenteric vessels could be observed in hiatus hernia cases.

Conclusion

Although the HPS in an intra-thoracic stomach is a rare condition, but with meticulous ultrasound scanning of diaphragmatic hiatus, pyloric canal, and mesenteric vessels, it can be diagnosed.

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List of abbreviations

GI series Gastrointestinal series
HPS Hypertrophic pyloric stenosis

Summary of the case

Patient (gender, age)	1	Male, 3 month old
Final Diagnosis	2	Intra-thoracic hypertrophic pyloric stenosis
Symptoms	3	Vomiting, diarrhea, weakness, and fever
Medications (generic)	4	Correction of water-electrolyte imbalance
Clinical Procedure	5	hiatal hernia repair and myotomy of pyloric muscle
Specialty	6	Ultrasonography

Consent for publication

Informed consent was taken from the parents of the patient.

Ethical approval

Ethical approval is not required at our institution to publish an anonymous case report in a medical journal.

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