CASE REPORT

Case of hepatic portal venous gas in an infant with hypertrophic pyloric stenosis

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SUMMARY

Hepatic portal venous gas is the presence of gas within the portal vein and its branches. A 4-week-old male infant presented with 1-week history of non-bloody, non-bilious projectile emesis. Examination was significant for an olive-shaped mass in the abdomen. Bloodwork showed hypokalaemic metabolic alkalosis. Abdominal ultrasound and radiograph was significant for portal venous gas and did not meet radiographic criteria for pyloric stenosis. He underwent upper endoscopy, which showed a narrowed, hypertrophic pylorus. The child underwent pyloromyotomy with resolution of his emesis. Hepatic portal venous gas (HPVG) is very rare and can be seen in the setting of hypertrophic pyloric stenosis. It is believed that an increase in intraluminal dilation and pressure subsequently moves gas from the intestinal mucosa venous system and lymphatics into the portal veins. The presence of HPVG in a well-appearing patient can be benign and should not prompt further testing nor delay treatment.

(UGI) series also did not show evidence of pyloric stenosis or other anatomical abnormalities. Measurements of pylorus' length or width did not meet radiographic criteria for pyloric stenosis. Given imaging findings, the patient was admitted to paediatric intensive care unit for close observation with concerns of bowel ischaemia. Antibiotics were initiated and he was empirically treated for presumed bacteraemia. His emesis persisted. AUSs were repeated on the second and third day, respectively. Results remained inconclusive for pyloric stenosis and showed resolution of HPVG. He ultimately underwent an upper endoscopy, which showed a narrowed, hypertrophic pylorus (figure 1). He then had another UGI series, which showed findings confirming pyloric stenosis.

OUTCOME AND FOLLOW-UP

He underwent a pyloromyotomy with resolution of his emesis.

BACKGROUND

Hepatic portal venous gas was initially described in 1950 as a radiographic sign due to increased mortality in infants with necrotising enterocolitis (NEC).¹ Its presence is now associated with multiple diseases which may be benign or potentially devastating, requiring immediate diagnosis and management. We report a case of a 4-week-old male infant with non-bilious emesis and radiographic finding of portal venous gas on abdominal ultrasound (AUS) and abdominal x-ray (AXR) secondary to hypertrophic pyloric stenosis.

CASE PRESENTATION

A 4-week-old male infant presented to the emergency department with a 1-week history of multiple episodes of non-bloody, non-bilious projectile emesis occurring with every third or fourth feed. Physical examination was significant for a well-appearing infant with an olive-shaped mass palpated in the mid-upper abdomen. Blood testing revealed a sodium of 132 mmol/L (135–146 mmol/L), potassium of 2.5 mmol/L (3.5–5.1 mmol/L), chloride of 80 mmol/L (96–107 mmol/L) and bicarbonate of 37 mmol/L (21–31 mmol/L). His vitals were all stable. He also had significant weight loss; he lost close to 500 g in 10 days.

INVESTIGATIONS

Initial imaging with AUS and AXR were significant for portal venous gas. Upper gastrointestinal

DISCUSSION

Hepatic portal venous gas (HPVG) is an extremely rare finding, which can be seen in the setting of hypertrophic pyloric stenosis (HPS). The presence of HPVG on imaging was previously thought to be an ominous sign warranting urgent intervention. However, with the improved sensitivity of ultrasound imaging, it is being found in an increasing number of other conditions such as ulcerative colitis, intra-abdominal abscess, small bowel obstruction and gastric ulcer.²

HPVG was first noted in a case series of paediatric patients with NEC in the 1950s. The diagnosis of HPVG can be made by various methods including plain AXR, AUS with Doppler imaging or CT abdomen. Ultrasound is the preferred test for diagnosing HPVG since it is non-invasive and sensitive but is also operator dependent. Features of HPVG on ultrasound include highly echogenic particles flowing within the portal vein, or poorly defined, highly echogenic patches within the hepatic parenchyma (figure 2).

HPVG has been associated with high mortality in the past. The exact pathophysiology underlying HPVG is not clear. One of the many proposed theories includes the migration of gas produced from intestinal microbe or abscess, which eventually circulate to the portal system.³ Ischaemic bowel and inflamed mucosa, as in inflammatory bowel disease, have also been associated with HPVG.⁴ It is hypothesised that luminal air enters the capillary veins by

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Figure 1 Hypertrophic pylorus visualised on upper endoscopy.

an impaired epithelial barrier.⁵ It is believed that an increase in intraluminal dilation causes an increase in intraluminal pressure and subsequent movement of gas from intestinal mucosa

Figure 2 Abdominal ultrasound revealing the presence of gas in the portal vein.

veins and lymphatics into the portal venous system. This serves as an explanation for the presence of HPVG in cases of gastric outlet obstruction, as seen in HPS.⁶ There have been case reports describing the presence of HPVG as a result of increased intraluminal pressure from a colonoscopy as well.⁷ As seen in our patient, those with HPS, the HPVG is usually a transient process that resolves within days of gastric decompression, which has also been seen with the use of a nasogastric tube or following surgery.

Learning points

- ▶ Hepatic portal venous gas (HPVG) is a radiographic sign with increasing incidence in other conditions secondary to increased sensitivity of imaging modalities.
- ▶ It is important that clinicians recognise that the presence of HPVG in a stable and well-appearing patient can be benign and should not be considered an ominous sign.
- ▶ As illustrated in our patient who presented with HPVG in the setting of hypertrophic pyloric stenosis, it should not be considered pathological, leading to further testing or delay in treatment.

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