Pediatr Surg Int (1998) 14: 100–101

CASE REPORT

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Gastric pneumatosis associated with preduodenal portal vein, duodenal atresia, and asplenia

Accepted: 6 January 1998

Abstract An 8-day-old newborn presented with non-bilious vomiting, upper abdominal fullness, and failure to pass meconium. Plain radiographs revealed gastric pneumatosis (GP). At operation, he was found to have a non-obstructive preduodenal portal vein, preampullary duodenal atresia, asplenia, and malrotation. The baby was treated by duodeno-duodenostomy without mobilizing the portal vein and correction of the malrotation according to Ladd's procedure. He made an uneventful recovery and the GP resolved spontaneously. The malformative process was believed to have occurred at or soon after the 5th week of gestation, and the GP probably resulted from intramural air tracking through mucosal tears caused by high intragastric pressure.

Key words Gastric pneumatosis · Preduodenal portal vein · Duodenal obstruction · Neonate

Introduction

Gastric pneumatosis (GP) is a rare occurrence and has been seen as a radiographic manifestation of gastric-outlet obstruction, usually secondary to hypertrophic pyloric stenosis (HPS), in infants [1]. It has, however, not been reported with duodenal atresia (DA). The case presented here had additional rare and interesting findings in the form of asplenia, DA, and malrotation.

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Case report

A full-term male neonate was referred at the age of 8 days with non-bilious vomiting and failure to pass meconium. He was small for dates, weighing only 1.45 kg, and did not have dysmorphic features. The mother was a 21-year-old gravida III, para II who was detected to have polyhydramnios at 32 weeks' gestation. At admission the baby was dehydrated and had upper-abdominal fullness with visible gastric peristalsis. Abdominal radiographs revealed a massively dilated stomach, a double bubble, and intramural air (GP) (Fig. 1). No air was seen in the bowel distally.

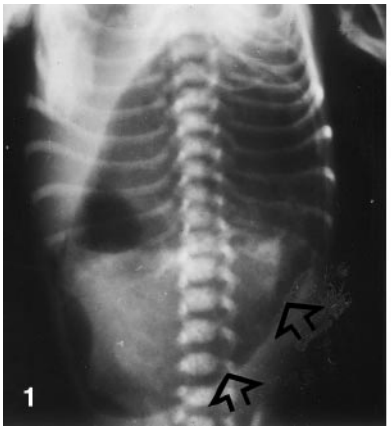
At laparotomy, the stomach was grossly distended and showed evidence of subserosal intramural air. The baby had a preampullary DA in the second part with a preduodenal portal vein (PDPV) running across the first part of the duodenum (Fig. 2). The vein was confirmed to be non-obstructive by pressing gas from the stomach into the duodenum. An associated malrotation with Ladd's bands crossing the third part of the duodenum was also observed. The spleen was absent. No intrinsic duodenal obstruction could be identified distal to the DA, and patency of the rest of the small bowel was confirmed. A duodeno-duodenostomy was performed without mobilizing or compressing the PDPV. A thin transpyloric feeding tube was placed in the proximal jejunum and was used to start feeds 48 h postoperatively.

Oral feeds were started on the 7th postoperative day and the child was discharged 3 weeks after admission when he had shown satisfactory weight gain. The GP resolved spontaneously. Echocardiography did not reveal any cardiac malformations. Asplenia was confirmed by ultrasound and technetium Tc-phytate scintigraphy. The child was doing well at last follow-up, weighing 8.5 kg at 10 months of age.

Discussion

Gastric pneumatosis is a very uncommon but characteristic plain-film radiographic sign. In infancy, the occurrence of isolated GP has been reported to be rare in gastric outlet obstruction secondary to HPS [1], neonatal necrotizing enterocolitis, usually with intestinal pneumatosis [2], and as a complication of gastrostomy catheter placement [3]. In adults, GP is caused either by gas-forming bacterial infection (emphysematous gastritis) or in association with bullous emphysema [4]. This is the first reported case of GP associated with DA. Speculating on the mechanism of GP in gastric outlet obstruction, Holgersen et al. [4] suggested that high intraluminal pressure may force gas into the wall of the stomach through superficial mucosal tears. Such tears are likely to occur due to high intragastric pressure associated with excessive vomiting, as seen in our case, and may have been responsible for the GP.

PDPV is a rare anatomic anomaly, with 63 cases reported in the



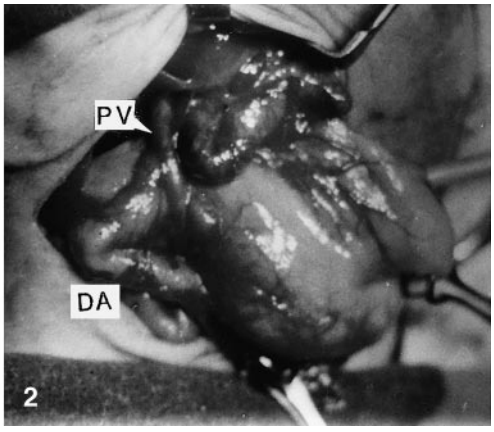


Fig. 1 Plain X-ray film showing gastric pneumatosis (*arrow*)

Fig. 2 Operative photograph showing duodenal atresia (*DA*) with preduodenal portal vein (*PV*)

literature upto 1990 [1]. It was first reported by Knight in 1921 [5], who also explained the embryologic basis of the anomaly as persistence of a preduodenal vitelline communicating vein. The development of the portal venous system was later reviewed and, on this basis, it is believed that this malformation occurs between the 5th and 10th weeks of gestation and results from persistence of the right vitelline vein and the third anastomotic bridge instead of the left vitelline vein and the second anastomotic bridge [6]. Of all patients reported with PDPV, 64% were children, two-thirds of whom were detected in the 1st week of life due to an intrinsic duodenal anomaly, malrotation, or Ladd's bands [7]. In our patient the PDPV was clearly non-obstructive and proximal to the area of obstruction.

Other anomalies associated with PDPV include biliary atresia, splenic malformations (asplenia/polysplenia), preduodenal common bile duct, and cardiovascular anomalies [1 9]. Interestingly, the development of the spleen and the rotation of the stomach also begin at the 5th week of gestation in an almost contiguous area to the one in which the portal vein (PV) is developing, and duodenal vacuolation commences after the 7th week, also in the same area [10]. Our patient had asplenia but no inferior vena cava or cardiac anomalies associated with the PDPV and DA. It is possible that a mesodermal failure occurring at or around the 5th week of gestation caused the PDPV and asplenia. Although the duodenum is an endodermal derivative, its close proximity to the site of PV development and the timing of the malformation suggest that it may have a common factor or that the anteriorly placed PDPV may have caused the DA. The DA was, however, found to be distal to the PDPV. This could be because duodenal growth subsequent to the atretic process caused the DA to lie distal to the PDPV. Since the PV and spleen are mesodermal derivatives, it is also possible that the DA occurred as an isolated incident subsequent to the event that caused the PDPV and asplenia.

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