A RARE CASE OF ISOLATED PYLORIC ATRESIA

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INTRODUCTION

• Congenital gastric outlet obstructions (GOO) involve the antrum or pylorus, with a large differential including pyloric atresia
• Cause of pyloric atresia are still being postulated, but one thought is genetic mutation affecting foregut segmentation
• Pyloric atresia is observed in 1:100,000 births
• Can be identified prenatally by imaging versus postnatally with surgery

CASE DESCRIPTION

• Prenatal ultrasound at 36 5/7 weeks showed double bubble sign and polyhydramnios, concerning for duodenal atresia (Figure 1)
• Family history remarkable for father with intestinal malrotation and maternal uncle with duodenal atresia
• Repeat ultrasound at 37 4/7 weeks negative for double bubble sign, only dilated fetal stomach
• Delivered via planned vaginal delivery at 39 weeks with pediatric surgical team on standby
• Postnatal testing included abdominal x-ray (Figure 2) and upper GI series, both consistent with GOO; lower GI was normal
• Diagnostic laparoscopy at day of life 2 demonstrated pyloric thickening (Figures 3a and 3b)
• Laparoscopy was converted to open procedure for pyloric web, consistent with Type 1 pyloric atresia
• No complications intraoperatively
• After 7 days of bowel rest and parenteral nutrition in the NICU, was transitioned to enteral feeds
• Discharged without feeding issues within a month after birth

FIGURES

DISCUSSION

• Congenital pyloric atresia (CPA) is a rare form of GOO that may present postnatally with non-bilious, non-bloody emesis, upper abdominal distension, lack of stool, failure to thrive and electrolyte disturbance
• Common prenatal findings include of CPA include single bubble sign and polyhydramnios
• Differential for CPA includes duodenal atresia, malrotation of midgut volvulus, gastric or pyloric duplication cysts, antral web, mucosal valve or diaphragm or retrograde duodenogastric intussusception and aberrant pancreatic tissue plugging pylorus
• There is familial occurrence of isolated CPA documented within siblings, but no known inheritance pattern or genetic mutation within families of various GOOopathologies
• Prenatal diagnosis of CPA is performed by comprehensive ultrasound looking for polyhydramnios and single bubble sign; double bubble sign more consistent with duodenal atresia
• If intestinal obstruction suspected, performing continuous observation of whole stomach configuration during 60-minute ultrasound to detect periods of active and absent gastric peristalsis can help discern between pyloric and duodenal obstruction
• During gastric peristalsis, double bubble sign may disappear and reappear when gastric peristalsis stops

CONCLUSION

• Polyhydramnios and single or double bubble sign should raise suspicion for GOO
• Prenatal imaging is helpful to prepare for management at the time of delivery while postnatal imaging is helpful in establishing diagnosis of GOO
• Ultimately, only surgical exploration can give confirmatory diagnosis
• Although there has been literature discussing isolated CPA between siblings, there is no literature extrapolating possible relationship between pyloric atresia and other GOO pathologies

REFERENCES