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Anomalous pancreatic ductal system allowing distal bowel gas with duodenal atresia

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ABSTRACT

A twelve-day-old male presented with non-bilious, non-bloody emesis 2 hours after feeding since two-days-old, with flatus and light stools since birth. An upper gastrointestinal series (UGI) at an outside institution was reported as normal. Abdominal radiographs demonstrated distal bowel gas with proximal duodenal dilation. Abdominal ultrasound demonstrated gas in the main pancreatic duct and a dilated, fluid-filled duodenum. An UGI revealed duodenal atresia with enteric bypass by congenital anomaly of the pancreaticobiliary system. At thirteen days old the patient underwent uncomplicated duodeno-duodenostomy. The post-operative course was uneventful and the patient subsequently tolerated oral feeds.

Bypass of the atretic duodenal segment through an anomalous pancreatic ductal system is a rare anomaly described in the literature in only a handful of cases. This case report highlights the importance of considering duodenal atresia and pancreaticobiliary enteric bypass in the differential diagnosis of neonates presenting with partial duodenal obstruction. On ultrasound, the presence of gas in the biliary tree or pancreatic duct should alert the physician to the possibility of duodenal atresia with congenital pancreaticobiliary duct anomalies that allow for bypass of enteric contents, including air, into more distal bowel, thereby creating a gas pattern aptly described as double bubble with distal gas.

Duodenal atresia normally occurs in 1 of 10,000 live births but in very rare cases can be masked by bypassing anomalous biliary or pancreatic ducts. Congenital anomalies of the pancreaticobiliary tree may allow bypass of enteric contents and air beyond the duodenal atresia and into more distal bowel, thereby delaying the diagnosis of duodenal atresia.

1. Case report

A twelve-day-old infant presented with non-bilious, non-bloody emesis 2 hours after each feeding since two-days-old. The infant was born at 40 weeks gestation by an uncomplicated caesarian section. According to the clinical history provided at the time of admission, he was passing flatus and had light stools since birth. The remainder of the prenatal history, birth history and neonatal course was otherwise normal.

The patient had undergone an outside upper gastrointestinal series (UGI) that was reported as normal and eventually presented to our institution for further evaluation. The physical examination was unremarkable and abdominal radiographs were obtained, demonstrating a normal bowel gas pattern aside from proximal duodenal dilation and a distended stomach (Fig. 1A). Since the vomiting was consistently reported as non-bilious, an abdominal ultrasound was obtained as a simple non-invasive screening tool to exclude the remote possibility of early pyloric stenosis (felt to be very unlikely due to the history of emesis since day two of life and the abundance of distal gas) and to evaluate for other etiologies related to the working differential diagnosis, which also included malrotation with volvulus and/or Ladd's bands (unlikely based on the clinical history but included due to the duodenal dilatation), and duodenal stenosis or web. The ultrasound demonstrated echogenic foci with dirty shadowing (gas) in the main pancreatic duct and a dilated, fluid-filled duodenum (Fig. 1B). No direct ultrasonographic signs of volvulus were seen and the superior mesenteric artery and vein relationship was normal.

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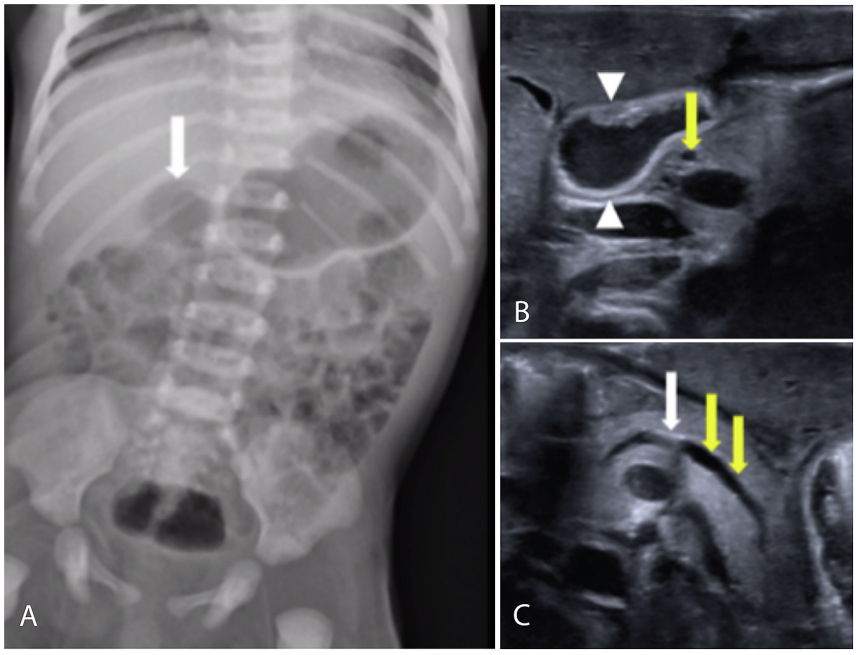

Fig. 1. A Supine and left lateral decubitus views of the abdomen demonstrate a normal gas pattern aside from proximal duodenal dilatation (arrow). **B** Transverse ultrasound demonstrated a dilated fluid filled duodenum (arrowheads) and **C** air (white arrow) in a dilated pancreatic duct (yellow arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Upon the recommendation of pediatric radiology, an UGI was performed to look for a cause of the suspected partial duodenal obstruction. The gas seen in the pancreatic duct was concerning and so water soluble contrast was used instead of thin liquid barium. The UGI revealed complete duodenal obstruction with filling of the main pancreatic duct mimicking the distal duodenum and normal duodenojejunal junction (Fig. 2). There was also filling of a system of anomalous pancreatic ducts that allowed the contrast to bypass the complete proximal duodenal obstruction, fill the main pancreatic duct and exit into more distal duodenum and proximal jejunum. A diagnosis of duodenal atresia with enteric bypass by a congenital anomaly of the pancreaticobiliary system was made. Further work-up of the patient included an echocardiography, which was within normal limits with no evidence of congenital heart disease.

Prior to surgery a nasogastric tube was placed to avoid further spillage of enteral contents into the pancreatic duct. Intravenous fluid hydration was initiated and the patient started on total parenteral nutrition. The lipase was 142 U/L on the day of presentation, and trended down to 112 U/L within 24 hours.

At thirteen-days-old, the patient underwent an uncomplicated duodenoduodenostomy for duodenal atresia with congenital anomalies of the pancreaticobiliary system. Intra-operatively, the duodenojejunal junction was identified to the left of the spine. The cecum and appendix were noted in the right lower quadrant, confirming normal rotation. A 5 French *trans*-anastomotic nasal corpak feeding tube was placed deep into the jejunum to allow for early post-operative feeding.

The patient's post-operative course was uneventful. The nasogastric and corpak tubes were discontinued on post-operative day 10, and he was discharged home at 27 days old tolerating oral feeds at goal.

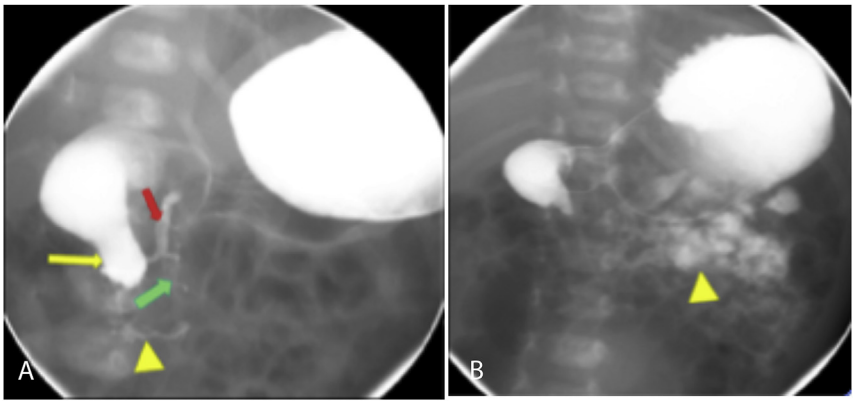

Fig. 2. A Water soluble contrast (dilute Omnipaque 300) UGI demonstrates duodenal obstruction (yellow arrow), filling of the main pancreatic duct mimicking distal duodenum (red arrow), anomalous pancreatic ducts (green arrow), and contrast in third portion of the duodenum (arrowhead). **B** Duodenal obstruction with contrast in the distal duodenum and jejunum (arrowhead). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)