Duodenal atresia type III showing distal bowel gas and atypical malrotation in association with gastric and bile duct duplications

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Abstract

Association of duodenal atresia (DA) with gastric duplication cyst is an extremely rare entity. Furthermore, DA type III with the presence of distal bowel gas is very rare. We report the case of a 13-day-old preterm male with neonatal intestinal obstruction. Radiological evaluation revealed duodenal obstruction at the second part with the presence of few distal bowel gas shadows. Laparotomy revealed dilated gastroduodenum, atypical malrotation, and a cystic structure (duplication cyst) identical to the stomach attached to its greater curvature along with type III atresia of the second part of the duodenum. Duodenotomies revealed the presence of bile along with air at either blind ends of the duodenum which could be explained by the presence of bile duct duplication at the distal end. We propose that this association is due to “defective mesenchyme formation during organogenesis,” which may explain multiple duplication abnormalities and DA.

Keywords: Atypical malrotation, distal bile duct duplication, distal bowel gas, duodenal atresia type III, gastric duplication cyst

INTRODUCTION

Duodenal atresia (DA) is a common cause of neonatal intestinal obstruction, while gastric duplications are rare alimentary tract duplications.[1,2] We present herein a neonate with DA type III with distal bowel gas due to bile duct duplication and atypical malrotation in association with gastric duplication cyst, which is extremely rare and not described in literature till date. We propose that this association in this index case is due to “defective mesenchyme formation during organogenesis,” which may explain multiple duplication abnormalities and intestinal atresia(s).

CASE REPORT

A 13-day-old preterm (32 weeks) male neonate weighing 1650 g presented with bilious vomiting and epigastric fullness since birth. Antenatal ultrasounds were not done. On examination, the neonate was dehydrated, jaundiced, and hemodynamically unstable; pulse rate was 142/min and respiratory rate was 48/min. The abdomen was soft, and epigastric fullness was present. Laboratory investigations revealed indirect hyperbilirubinemia (total bilirubin – 12 mg/dl) and C-reactive protein levels >6 mg/L. Abdominal ultrasound was suggestive of duodenal...
obstruction with the presence of sludge in the gallbladder. An abdominal X-ray revealed duodenal obstruction at the second part with the presence of few distal bowel gas shadows [Figure 1]. An upper gastrointestinal contrast series showed complete obstruction at the second part of duodenum [Figure 1]. Preoperative optimization was performed. Laparotomy revealed large, dilated gastroduodenum; atypical malrotation with the presence of Ladd’s bands over the second part of the duodenum, ligament of Treitz to the left of the midline, relatively narrow mesenteric base, and cecum in the left hypochondrium. A cystic structure (duplication cyst) identical to the stomach wall, attached to its greater curvature, noncommunicating with the lumen of the stomach, and sharing common blood supply was appreciated [Figure 2]. Division of Ladd’s bands and kocherization of the duodenum revealed type III atresia of the second part of duodenum. Noncollapsed small bowel (suggestive of air and meconium filled) loops were present. Duodenotomies revealed the presence of bile along with air at either (blind) ends of the duodenum. Widening of the mesenteric base and Kimura’s duodeno-duodenostomy were performed [Figure 2]. Excision of large noncommunicating gastric duplication cyst was deferred as the neonate had an episode of intraoperative desaturation. Furthermore, the neonate was premature, was of low birth weight, and there was sepsis. The postoperative course was uneventful. The patient became jaundice free and is doing well, gaining weight at 2-month follow-up.

DISCUSSION

DA is a common cause of neonatal intestinal obstruction with an estimated incidence of between 1 in 6000 and 1 in 10,000 births. Type III DA is two blind ends of the duodenum separated by a gap with a V-shaped mesenteric defect. Usually, all types of DA are limited to the second (postampullary) part of the duodenum, and bilious vomiting is the main presenting symptom. Plain radiography shows “Double Bubble” sign and is the most valuable diagnostic tool in all cases of DA. Exceptions from this sign are those with incomplete obstruction due to duodenal web (type I) or distally located duodenal webs. In our case, radiological evaluation revealed duodenal obstruction at the second part with the presence of few distal bowel gas shadows. A differential diagnosis of DA was made. Furthermore, DA type III with the presence of distal bowel gas is very rare, as seen in our case. This unusual presence of distal bowel gas could be explained by the presence of partial duplication of distal end of the bile duct associated with DA as reported in some series. Distal end of the bile duct with partial duplication may be “T” or “Y” shaped. Anomalous bifurcated bile duct conduit with distal bowel gas was demonstrated by preoperative upper gastrointestinal series in only 8.77% of patients in a large series of DA. In our index case, partial duplication of bile duct could not be demonstrated by upper gastrointestinal series, which may be due to smaller lumen of the bile duct, allowing only passage of air through the bifid duct into the distal bowel. This was confirmed intraoperatively by noncollapsed small bowel (suggestive of air and meconium filled) loops, as shown in Figure 2.

Gastric duplications are rare, constituting about 2%–7% of all alimentary tract duplications. Majority of them are noncommunicating cysts, arise at the greater curvature, and present as large lump in an early age. Delayed presentation with complications such as acute abdomen, peritonitis and pancreatitis due to hemorrhage, infection, perforation, volvulus, and intussusception and compression effects of the cyst is reported. Intestinal duplications have sporadically been reported with intestinal atresia, but the association of gastric duplication with DA is extremely

Figure 1: Abdominal X-ray (images on the left) showing gastric and duodenal gas shadows up to the second part of the duodenum along with the presence of few small distal bowel gas shadows (indicated by red stars); also seen are two separate gas shadows in the right upper abdomen (arrows). Upper gastrointestinal contrast study (images on the right) revealing dilated stomach and duodenum up to its second part with the absence of contrast distally.
Intraoperative images showing dilated stomach, cystic structure (indicated by yellow arrows) identical to the stomach wall, attached to its greater curvature, and sharing common blood supply from omental vessels; atypical malrotation with cecum present in the left hypochondrium (pointed by forceps) is appreciated. Noncollapsed small bowel (suggestive of air and meconium filled) loops in the left upper and right image (before duodenotomies) are also seen. The left lower image shows completed Kimura’s duodeno-duodenostomy.

In conclusion, DA type III with distal bowel gas due to distal bile duct duplication and atypical malrotation in association with gastric duplication cyst is extremely rare. We propose that the association of complete and partial duplication abnormalities and associated intestinal atresias is due to “defective mesenchyme formation during organogenesis.”

Declarations of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the guardians have given their consent for the infant’s images and other clinical information to be reported in the journal. The guardians understand that the names and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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