

Duodenal Atresia Associated with Apple-Peel Configuration of Distal Small Bowel in a Late Preterm Neonate: A Rare Case Report and Literature Review

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Abstract

Background: Duodenal atresia is a common congenital intestinal obstruction, whereas apple-peel (or Christmas tree) atresia is an extremely rare variant of duodenal atresia. Apple-peel atresia is itself a rare variant of jejuno-ileal atresia. So far, very few cases of duodenal atresia with jejuno-ileal apple-peel configuration have been reported worldwide.

Case Presentation: We report a late preterm (35+5 weeks) female neonate, antenatally suspected to have jejunal atresia, who presented with bilious vomiting and

mild abdominal distension from the second day of life. Postnatal abdominal ultrasound and echocardiography were normal, but plain radiography demonstrated a “double bubble” sign suggestive of duodenal obstruction. Exploratory laparotomy revealed duodenal atresia with apple-peel configuration of the distal small intestine. A duodeno-jejunosomy was performed with preservation of maximal small bowel length. The postoperative course was uneventful.

Conclusion: This case highlights the rare association of duodenal atresia with apple-peel configuration,

supporting a possible vascular etiology and underscoring the need for meticulous surgical planning to preserve bowel length and optimize outcomes.

Keywords: Duodenal Atresia, Apple-Peel Atresia, Jejunioileal Atresia, Neonatal Intestinal Obstruction, Vascular Etiology.

Introduction

Duodenal atresia (DA) occurs in approximately 1 in 5,000–10,000 live births and represents one of the most common causes of neonatal intestinal obstruction^{1,2}. It is classically attributed to failure of duodenal recanalization during embryogenesis. Apple-peel atresia, by contrast, is a rare form of jejuno-ileal atresia thought to result from intrauterine vascular insult leading to absence of much of the mesentery, with distal small bowel spiraling around a single vascular arcade³. The coexistence of duodenal atresia and jejuno-ileal apple-peel configuration is exceedingly rare, with only sporadic cases reported in the literature⁴⁻⁷. We present a case of a late preterm neonate with antenatal suspicion of jejunal atresia, in whom duodenal atresia and apple-peel distal bowel were diagnosed during laparotomy. This case report has been prepared in line with the Surgical Case Report (SCARE) criteria.

Case Presentation

A 2-day-old female neonate was born to a 28-year-old para 2 mother at 35 weeks + 5 days of gestation via cesarean section for prolonged PROM and failed induction. PICT and viral markers were negative. Antenatal ultrasonography had suggested jejunal atresia with polyhydramnios. The baby cried immediately after birth, weighed 2.4 kg, and had APGAR scores of 9 and 10 at 1st and 5th minutes, respectively, requiring no resuscitation.

On the second day of life, she developed frequent bilious

vomiting with mild abdominal distension. Otherwise, she had no history of failure to suck, yellowish discoloration of the body, cough, or fast breathing. On examination, she was well-looking and alert. Vital signs were PR = 150, RR = 54, T° = 37°C, and SpO₂ = 92% in room air. She had pink conjunctiva and non-icteric sclera. Chest and cardiovascular examinations were unremarkable. There was mild upper abdominal distension but no palpable mass or tenderness. Per-rectal examination showed a normal anal opening with some meconium-stained fluid. Other systems were normal. Investigations revealed an unremarkable abdominal ultrasound (no free fluid, no mass, normal hepatobiliary system). Echocardiography was also normal. Plain abdominal radiograph showed a “double bubble” sign suggestive of duodenal obstruction (see figure 1). The baby was stabilized with intravenous fluids, IV ampicillin 120 mg BID, IV gentamicin 9 mg daily, and nasogastric tube decompression. Written informed consent was obtained, and the patient was transferred to the OR for surgery.

Intraoperatively, there was type III duodenal atresia just distal to the ampulla of Vater. The 3rd and 4th parts of the duodenum, proximal part of the SMA, and ligament of Treitz were absent (see figure 2). The distal small bowel showed apple-peel configuration with a long segment of jejunum and ileum spiraling around a single vascular supply with absent mesentery (see figure 3). There was minimal peritoneal ascitic fluid. Other viscera appeared normal.

A diamond-shaped duodeno-jejunostomy was performed, preserving maximal small bowel length. Two tubes were placed through a single nostril — one for gastric decompression and the other as a trans-anastomotic feeding tube for early feeding initiation, as

total parenteral nutrition is unavailable in our setup. The apple-peel configuration was viable and left intact. Postoperatively, the neonate was managed with NPO and maintenance fluids for the first 36 hours. Preoperative IV antibiotics were continued. Trophic feeding started after 36 hours through the feeding NG tube. Two doses of Pergram amino acid infusion were given. On the 3rd postoperative day, the gastric tube was removed, oral feeds were gradually introduced, and tolerance improved over the following days. On the 5th postoperative day, the trans-anastomotic feeding tube was removed. Recovery was uneventful, and she was discharged in good condition. On her 3rd postoperative week, the wound was healed, she was nutritionally improved and alert with no complaint. She is currently on regular follow-up at the pediatric surgical referral clinic.



Figure 1: Plain abdominal X-ray showing double bubble sign suggestive of duodenal atresia



Figure 2: Intraoperative picture showing proximal dilated atretic duodenum with distal atretic end of jejunum and apple-peel configuration, later diamond-shaped duodeno-jejunal anastomosis following refreshing and spatulating of the jejunal atretic end was done. Mesenteric defect left open

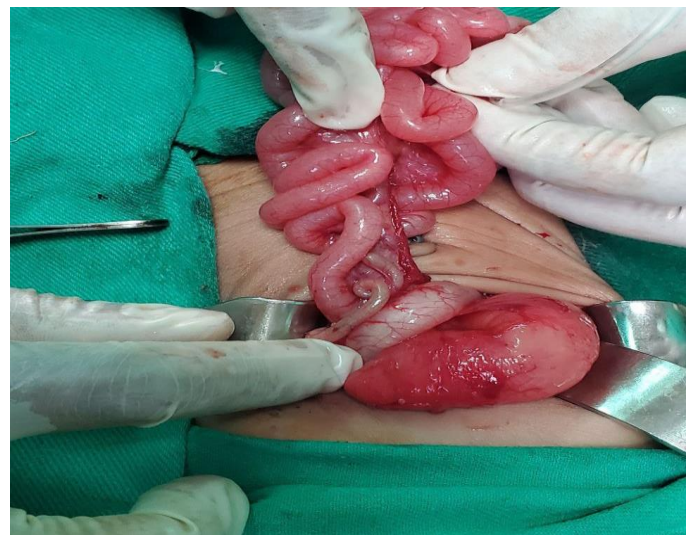


Figure 3: Picture taken just after saline infusion through the jejunal atretic end showing typical Christmas tree appearance; the rest of the bowel was checked for patency and found normal.

Discussion

Duodenal atresia is usually an isolated anomaly but can coexist with other congenital malformations, including trisomy 21, congenital heart disease, and annular

pancreas¹⁻². The association with apple-peel atresia is extremely rare and has been described in only a handful of cases⁴⁻⁷.

Classically, duodenal atresia is thought to result from failure of recanalization during the early embryonic period, while jejuno-ileal apple-peel atresia arises from a vascular insult to the superior mesenteric artery during late fetal life³. The coexistence of the two lesions challenges traditional embryological theories, suggesting that a vascular insult may also underlie some cases of duodenal atresia^{4,7}.

Clinically, both conditions present with bilious vomiting and abdominal distension. The “double bubble” sign is typical for duodenal obstruction, though the apple-peel configuration is rarely diagnosed before surgery^{2,6}. Surgical correction requires individualized planning. In our patient, duodeno-jejunostomy relieved the proximal obstruction, while preservation of the apple-peel distal bowel was crucial to avoid short bowel syndrome. Postoperative outcome depends on residual bowel length, presence of associated anomalies, and neonatal stability⁵⁻⁷.

Our case adds to the limited literature supporting a vascular basis for at least some cases of duodenal atresia and coexistence of duodenal atresia with Christmas tree abnormality of the distal small bowel. It also highlights the importance of meticulous intraoperative assessment to preserve bowel length.

Conclusion

The coexistence of duodenal atresia associated with apple-peel configuration of the distal small intestine is extremely rare. This unusual association supports a possible vascular etiology of duodenal atresia. Early recognition, prompt surgical management, and bowel-preserving strategies are essential for favorable

outcomes.

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