“Inverse Type” Apple-Peel Syndrome Is Associated with Type III Colonic Atresia in a Neonate with Gastroschisis—A “New” Subtype of Colonic Atresia

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Abstract

The colon is an unusual site of intestinal atresia. Colonic atresia is subdivided into three phenotypes. Type III is the most common phenotype, where the proximal and distal blind sacs are not connected. Here, we report on the presence of colonic atresia with an “inverse apple-peel” appearance in a neonate with gastroschisis. The lack of mesenteric fixation of the entire small intestine, including the proximal colon, and the twisting around the vascular axis of the superior mesenteric artery led to intrauterine volvulus and hemorrhagic infarction of the ileocolic bowel at 34 weeks of gestation. According to the current nomenclature for small bowel atresia, we introduce type IIIB into the current colonic atresia classification. The occurrence of type IIIB has been mentioned in the literature, but no single cases have been reported until now. Patients with this type of atresia are predisposed to the loss of the ileocecal region.

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Keywords
Apple-Peel Syndrome, Colonic Atresia, Volvulus, Gastrochisis

1. Introduction
Colonic atresia (CA) is a very rare congenital malformation with a prevalence of 1 in 20,000 live births [1]. Prenatal ultrasonography typically shows enlarged loops of the small and large bowel. CA can be associated with anomalies of the heart, gastrointestinal tract, abdominal wall, and musculoskeletal system. Clinically, a newborn with CA develops signs of complete obstruction. Distension of the abdomen within 24 to 48 hours and delayed feculent vomiting are the leading symptoms. According to the recent literature, three different types are described. Type III is characterized by the complete interruption of the colonic continuity and the presence of a gap in the mesocolon [1]-[4]. For the more common types of small bowel atresia, the nomenclature encompasses four types, and type III is subdivided into two subtypes [5] [6]. According to the clinical phenotype, subtype IIIIB is referred to as “apple-peel” or “Christmas tree” atresia. The term “apple-peel” was first introduced in 1961 [7] for a special type of jejunal atresia with a discontinuity of the small bowel and a wide mesenteric gap. It describes a shortened and coiled small bowel around a retrograde perfusing artery, suggestive of a coiled apple peel. This retrograde perfusing artery of the apple-peel predominately compensates for the interrupted superior mesenteric artery [8]. Infants with complex gastrochisis carry an increased risk for small bowel atresia or CA [9] [10]. To the best of our knowledge, this is the first detailed report on the association of CA with an apple-peel phenotype affecting the small gut in combination with the ileocolic region.

2. Case Report
A prenatal ultrasound at 14 weeks of gestation revealed the extrusion of the fetal bowel out of the coelom associated with extra-abdominal bowel dilatation. At 34 weeks of gestation, the boy was delivered by Cesarean section (CS) due to fetal compromise. An ultrasound revealed pathological thickening of the bowel wall and an impaired cardiotocogram. The birth weight was 1820 g (10th percentile), and the Apgar scores were 6, 8 and 9 at 1, 5 and 10 minutes, respectively. The umbilical arterial pH was 7.35, and the amniotic fluid was stained with blood and meconium. Gastrochisis with the eventration of nearly the entire bowel and the stomach was confirmed. The lower parts of the infant’s body were immediately placed in a “bowel bag”. The infant was then stabilized with intubation, mechanical ventilation and intravenous fluid administration. A gastric tube was inserted, and aspiration revealed the presence of large amounts of bloody gastric content. The postnatal laboratory investigations revealed anemia with a hemoglobin (Hb) level of 12.8 g/dL, and an erythrocyte transfusion was required preoperatively. A surgical evaluation and primary closure of the abdominal wall were performed at the local neonatal intensive care unit (NICU). The abdominal defect was approximately 3 cm in diameter, and there was no fibrous coating over the extra-abdominal bowel. The protruded, non-fixed and twisted bowel was adherent to only a very small mesenteric pedicle. It consisted of the entire small bowel, the ileocecal region and 4 cm of the blind end of the cecum with the appendix vermiformis, and the ascending colon (Figure 1). There was a broad gap between the proximal colon and the tiny, non-used distal colon. The latter was densely adhered to the base of the mesentery (Figure 2). The derotation of the twisted mesenterial pedicle revealed the presence of a common ileocolic mesentery containing the blood supply to the small and the ileocecal bowels (Figure 3). In addition to the apple-peel deformity, the terminal portion of the extra-abdominal bowel was twisted at least 180° with a resulting hemorrhagic infarction. The resection of the proximal and atretic portion of the colon (15-cm length), including the terminal ileum, was required, and a temporary terminal ileostomy was created. The post-operative course was prolonged due to the inability for full enteral feeding for several weeks. Under parenteral nutrition, the infant tolerated increasing amounts of oral feeding. At 8 weeks of age, a temporary end-to-side anastomosis with a stoma of the proximal bowel according to Santulli was established [7]. During surgery, the relation of the bowel diameter was determined to be 6 to 1. The passage of increasing amounts of stool occurred after two weeks and allowed the closure of the enterostomy after five months with a body weight of 4520 g (below 3rd percentile). Oral nutrition by breastfeeding was tolerated increasingly, and weight gain 1.5 month after surgery was 1.45 kg. During a follow up of 11 months a slow catch up growth occurred.
Figure 1. The appearance of the dilated and blind-ending colonic atresia (asterisk). The unused colon is fixed to the base of the mesenteric stalk (arrow).

Figure 2. The vascular axis of the small bowel (arrows) together with the ileocecal region after detorsion.

Figure 3. The histology of the adherent small mesenteric rim with dilated blood (asterisks) and lymphatic vessels (hematoxylin/eosin, ×50).
3. Discussion

Colonic atresias account for 1.8% to 15% of all intestinal atresias, and they are subdivided into three phenotypes [1] [2] [5] [6] [11]-[13]. Type I is characterized by complete obstruction by a diaphragm, and in type II, the two blind ends are connected by a fibrous band. Type III is the most common phenotype, where the proximal and distal blind sacs are not connected. The occurrence of type IIIb has been mentioned in the literature, but no cases have been reported until now [5] [6] [12].

It has been shown that intrauterine events or “catastrophes”, such as volvulus, intussusception, herniation and snaring of the bowel during the fetal time period, may be responsible for the development of bowel atresia below the duodenum [2] [7] [11] [12] [14]. This “vascular theory” for the pathogenesis of bowel atresia was confirmed via animal experiments. In fetal puppies, the interruption of the blood supply to the bowel was able to induce the entire spectrum of different types of intestinal atresia or stenosis [14]. Therefore, with the exception of duodenal atresia, vascular disruption is responsible for the majority of the cases with bowel atresia.

The presence of nonrotation of the entire gut together with a narrow mesenteric pedicle predisposes patients with gastroschisis to develop intestinal atresia during the intrauterine period. Approximately 5% to 25% of these neonates are affected with intestinal atresia, and in 2.5% of the infants with gastroschisis, a CA can be expected [1] [10]. The coincidence of gastroschisis and the associated intestinal pathologies, such as intestinal atresia, perforation or volvulus (complex gastroschisis), is associated with an elevated risk for postsurgical morbidity and mortality [9] [13]. Early elective delivery for suspected complex cases of gastroschisis is normally advocated in order to prevent the development of bowel necrosis secondary to the abdominal events. However, a recently published multi-institutional study did not reveal any benefit to the elective delivery at <37 weeks of gestation [15].

In this case report, we present a “new” subtype of apple-peel malformation associated with CA in an infant with gastroschisis. In its classic form, apple-peel syndrome is associated with complete jejunal atresia with a broad gap between both blindly ending parts of the intestine and twisting of the distal small bowel (type IIIb) [5]. According to the vascular hypothesis, the occlusion of the superior mesenteric artery causes the absence of part of the jejunum, and the perfusion of the distal small intestine is maintained in a retrograde manner by the collateral vessels originating from the ileocolic artery [8].

In this “inverse” case of apple-peel syndrome, the atresia was located distally in the ascending colon and the “inverse” apple-peel affected the proximal small bowel and included the ileocolic region. The vascular supply of the entire small and ileocolic intestines was maintained by the completely preserved superior mesenteric artery. The described malformation can be explained by the interruption of the ileocolic, right, and middle colonic arteries. The disappearance of the middle portion of the colon, the tiny, non-used distal colon, and the absence of any bile stained content in the distal colon indicate that the atresia developed during early fetal development.

Fetal compromise at 34 weeks of gestation led to an emergency CS. In our case, the bowel infarction was not the result of a tight or closing abdominal wall defect [2]. Additionally, the volvulus was facilitated by the presence of only a small mesenteric stalk in the extracoelomic gut. The colonic atresia together with a broad mesenteric defect led to failed fixation and a perpendicular predisposition to the torsion of the gut. Concordant with the presented course, it has been described that the presence of meconium or blood-stained amniotic fluid and either prenatal or postnatal gastrointestinal complications is associated with an older age to reach full enteral feeds [16]. Therefore, the reported infant suffered from type 2 intestinal failure, requiring >28 days of parenteral nutrition [15].

4. Conclusion

In contrast to the classical type of apple-peel syndrome, the “inverse type” of apple-peel syndrome is associated with a high risk to the loss of the ileocecal region.

References


