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Case Report

Atresia of the Ileocecal Junction with Agenesis of the Ileocecal Valve and Vermiform Appendix: Report of a Case

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Abstract
Intestinal atresia involving the ileocecal region is a very rare intestinal malformation, and the presence or absence of the ileocecal valve influences its surgical management. We report the case of a male newborn with a provisional diagnosis of distal ileal atresia, in whom laparotomy revealed that the entire ileocecal region was atretic with an absent ileocecal valve and appendix vermiformis. We resected the dilated terminal ileum together with the atretic segment and performed an ileocolic anastomosis between the terminal ileum and the transverse microcolon without valve reconstruction. When last seen, 8 months after the operation, the baby was developing normally. Ileocolic anastomosis without valve replacement appears to be sufficient if an ileocecal valve is completely absent and only a short segment of the terminal ileum is lost.

Key words Ileocecal atresia · Ileocecal valve agenesis · Ileocecal valve reconstruction

Introduction

Jejunoileal atresia occurs in 1:330 to 1:1500 live births.[1] Although about one third of intestinal atresias are localized in the distal ileum, atresia of the ileocecal junction and atresia of the ileocecal valve are rare. To the best of our knowledge, only four such cases have been reported in the English-language medical literature.[2–4] Agenesis of the vermiform appendix is also a rare condition, found in approximately 1 in 100000 laparotomies performed for suspected acute appendicitis.[5] Again, only a few cases have been reported in the English-language medical literature.[6,7] We report a case of intestinal atresia involving the entire ileocecal junction, with agenesis of the ileocecal valve and a vermiform appendix. We discuss the reasons for the decisions we made intraoperatively.

Case Report

A male neonate weighing 4200 g was born at 41 weeks' gestation by Cesarean section for breech presentation to a nulliparous, 25-year-old-woman with no remarkable family history. The prenatal ultrasound scan showed dilated intestinal loops, but no evidence of polyhydramnios. Abdominal distension with no passage of meconium was reported at 24 h. An erect plain film of the abdomen showed dilated loops of bowel with fluid levels on the right side and an absent bowel gas pattern in the lower abdomen (Fig. 1). A contrast enema showed microcolon up to the mid-transverse colon, with no passage of contrast beyond this point. Based on these findings we made a provisional diagnosis of terminal ileal atresia, and performed an emergency laparotomy. Laparotomy revealed that the distended terminal ileum was connected to the transverse microcolon with a 6-cm segment of bowel atrophic bowel (Fig. 2). No tenia coli or appendix was identified in this atrophic segment. We also noted a V-shaped defect in the mesentery supplying the ileocecal region.

We resected the grossly distended last 8 cm of distal ileum together with the 6-cm segment of atrophic intestine. After irrigating the lumen of the transverse colon with normal saline to exclude distal obstructions, we performed an end-to-oblique Dennis–Brown anastomosis. Closer examination of the mesentery and small bowel showed evidence of normal rotation. The baby had an uneventful postoperative course. He passed meconium on postoperative day 2 and commenced oral feeding on postoperative day 5. When last seen at his

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8-month follow-up, he was developing well with height and weight within the 30th percentile.

Histopathological examination of the resected segments revealed complete absence of the ileocecal valve and a 1.5-mm deep blind sac on the tip of the atrophic bowel specimen. Microscopy showed evidence of an atrophic large bowel mucosa and the blind sac showed evidence of an atrophic appendix and lack of the tenia coli.

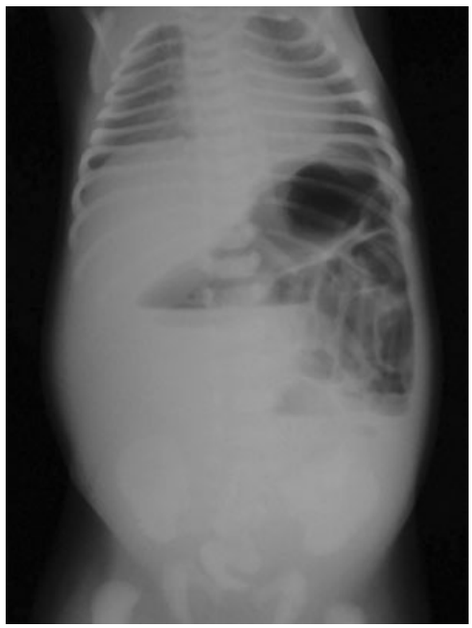

Fig. 1. Erect plain X-ray film of the abdomen showing dilated loops of bowel, fluid levels, and an absent bowel gas pattern in the lower abdomen

Discussion

Our review of the English-language medical literature revealed only four cases of atresia of the ileocecal junction.[2–4] Ein et al.[2] and Cacciari et al.[3] reported three cases of atresia of the ileocecal valve in which the ileocecal valve was not completely absent, but a common wall between the ileum and the cecum sealed the outlet of the valve. In these three infants the appendix was normal. Ein et al. resected the atretic segment and performed ileocolonic anastomosis without valve replacement. In contrast, Cacciari resected only the middle part of the atretic ileocaecal valve, followed by valve reconstruction. The fourth case, reported by Grassi et al.,[4] was found in a 20-year-old, who had atresia of the ileocecal region and complete agenesis of the ileocecal valve. The presence or absence of the appendix was not mentioned in this report. This patient was also treated by resection and ileocolic anastomosis with no valve reconstruction, followed by a good long-term outcome.

Our patient had atresia of the ileocecal region with complete absence of the ileocecal valve and appendix, which is unlike any of the above four cases. We found 21 cases reported in PubMed of isolated agenesis of the appendix. The etiology of the congenital absence of the appendix is unknown, although it was often seen in babies whose mothers took thalidomide during pregnancy.[8] There was no record of our patient's mother having taken any medication during pregnancy.

Clinical and experimental evidence suggests that ileal atresia could result from ischemic injury after the midgut has returned to the coelomic cavity.[9] This and the V-shaped defect observed in the ileocecal mesentery of our patient led us to postulate that a vascular accident

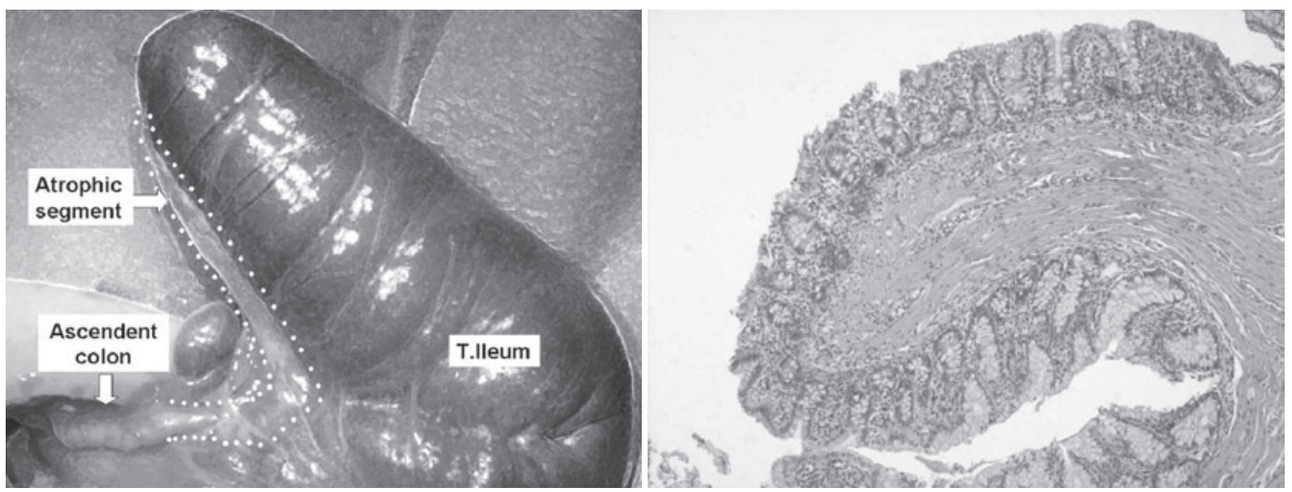

Fig. 2. **a** Intraoperative photograph showing the atrophic large bowel marked with *white dots*, and an absent appendix. **b** H&E-stained section of the resected intestine

causing ischemia of the entire ileocecal region may have resulted in the intestinal malformation.

Intraoperatively, the first difficulty we encountered was identifying the ileocecal junction in the absence of an identifiable vermiform appendix. The second difficulty was deciding how much bowel to resect. The necessity to preserve all viable native bowel in a newborn, even if it requires multiple anastomosis, has been highlighted in the past.[10–12] Although a lumen was palpable within the abnormal cecum and the colon ascendens was resected, because only a small segment of bowel was involved and the external appearance was grossly abnormal, we decided to remove the entire segment of bowel involved. The third difficulty was deciding whether to reconstruct the ileocecal valve.[13] Since only about 14 cm of small bowel was lost we decided not to attempt reconstruction.

In summary, atresia of the ileocecal junction is an extremely rare finding; however, the presence or absence of the ileocecal valve influences the surgical management. Reconstruction of the ileocaecal valve is a valid alternative to intestinal resection when an abnormal ileocecal valve is present.[3] Ileocolic anastomosis without valve replacement or reconstruction appears to be sufficient when the ileocecal valve is completely absent and only a short segment of terminal ileum is lost.

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