True atresia of the ileocecal valve: Diagnosis and management of an extremely rare entity

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A B S T R A C T

True atresia of the ileocecal valve is extremely rare, with fewer than 10 cases having been reported in the literature. The case presented is that of a preterm newborn female with the diagnosis of a neonatal bowel obstruction in whom an exploratory laparotomy revealed an atresia of the ileocecal valve itself. As the patient had a normal length of intestine otherwise, an ileocecectomy with a primary anastomosis between the terminal ileum and right colon was performed. The patient recovered uneventfully and was discharged without complication.

Congenital intestinal atresia is a significant cause of bowel obstruction in the neonatal population. The most commonly affected segments of intestine are the jejunum and ileum, while colonic atresia is rare [1]. Atresia specifically of the ileocecal valve itself appears to be the rarest form. The management of this particular type of atresia varies because of the important role that the ileocecal valve plays in cecoileal antireflux [2]. This case report describes an infant with true atresia of the ileocecal valve presenting as a neonatal bowel obstruction following a course of medically managed necrotizing enterocolitis (NEC). We describe the preoperative work-up which confirmed the bowel obstruction. We also discuss the intraoperative maneuvers utilized in order to pinpoint the exact location of the lesion. Unique operative and histologic images are included.

1. Case report

A 29 and 3/7 weeks’ gestation preterm female infant with a birth weight of 1275 g was born at an outside hospital due to premature rupture of membranes. Feeds were started and advanced gradually. By report, at 12 days of life she was treated medically for NEC, which resolved without complication and without the need for surgical intervention. Feeds were eventually re instituted, and following fortification of the formula to increase the caloric content, the patient had an episode of bloody stool. Abdominal radiographs demonstrated pneumatosis intestinalis consistent with recurrent NEC. Thus, at 3 weeks of life she was transferred to our institution for further management. Antibiotics had been started and nasogastric tube decompression was instituted. Subsequent serial abdominal radiographs demonstrated resolution of the pneumatosis intestinalis. Once again the patient recovered without the need for surgical intervention. Contrast studies were obtained following the completion of medical management of the second episode of NEC to evaluate for a stricture. A water-soluble contrast enema revealed no luminal irregularity from the rectum to the cecum, and no stricture was identified (Fig. 1). There was, however, no reflux of contrast into the terminal ileum. An upper gastrointestinal series with small bowel follow through revealed normal rotation, minimally dilated proximal small bowel loops, and a dilated distal small bowel segment without passage of contrast into the colon, even on delayed imaging (Fig. 2). Follow-up abdominal plain films revealed no further passage of contrast, prompting an exploratory laparotomy for a presumed post-NEC stricture in the distal small intestine. Upon entering the abdominal cavity, there were no noticeable adhesions or scarring in the abdomen. It was noted that the proximal small bowel appeared decompressed and that the distal small bowel was dilated to the level of the ileocecal valve. Attempts to manually pass small bowel contents into the colon were unsuccessful. The small intestine was then transected just proximal to the ileocecal junction. An attempt to pass a catheter...
from the ileum to the cecum failed. The proximal ascending colon was then transected, and the catheter failed to pass from the cecum to the ileum, and so the ileocecectomy was completed and a primary ileocolic anastomosis was performed. Upon evaluation of the specimen in the operating room, there was a common mucosal layer where the ileocecal valve should have developed (Fig. 3). The appendix appeared normal. Gross examination by the Pathologist demonstrated a common wall separating the ileum from the cecum, indicative of an atresia of the ileocecal valve itself. Further histologic examination demonstrated ileal mucosa on one side of the obstructing membrane and colonic mucosa on the other, without inflammation or scarring whatsoever (Fig. 4). The cecum was probe-patent to the appendix. The patient progressed well postoperatively and eventually was advanced to full enteral feeds. She made an uneventful recovery and was discharged home passing normal stools. She continues to do well 6 months following the operation.

2. Discussion

The ileocecal valve forms at the junction of the small and large intestine just proximal to the cecal bud, which itself develops within the distal midgut loop at about the 6th week of gestation. The valve serves an important role in humans, limiting the reflux of colonic contents, including bacterial flora, into the small intestine. In addition, it helps with increasing absorption time of nutrients, including chyme, which is of even greater importance in cases when extended segments of bowel have been resected [3–5].

Jejunoileal atresia occurs from as low as 1.3 to 2.25 cases per 10,000 live births in Spain, Latin America, and France to as high as 2.9 cases per 10,000 live births in various regions of the United States [1]. While duodenal atresia is thought to be due to failure of recanalization of the intestinal lumen, jejunoileal and colonic atresia are likely the result of an in utero vascular accident. The most commonly affected segment of intestine is the jejunum followed by the ileum. Colonic atresia is comparatively rare [1]. Atresia specifically of the ileocecal valve itself appears to be the rarest form of all. Upon review of the literature, there are less than 10 cases of atresia of the ileocecal valve that have been documented. Numerous variants of the atresia have been described including failure of development of the entire ileocecal region to a membranous obstruction at the level of the valve with a normal appendix [6–9].

The case presented here is similar to the latter, as are the majority of cases that have been reported. Optimal treatment of each case depends on the situation noted intraoperatively. Cserni [7], Ein [8], and Srivastava [10] all proceeded with resection of the ileocecal segment and primary end-to-end anastomosis, as was done in our case. Cacciari et al. described their approach to a unique situation in a case of an infant with an atresia as well as multifocal NEC necessitating a more extensive bowel resection than would be needed for an uncomplicated ileocecal valve atresia [9]. They performed an enteroplasty of the valve where the center of the common wall was excised and the lips were then sutured together to prevent recurrence or stricture. Mousavi and Sarparast proceeded with opening of the obstructing mucosal membrane and performing a tapering enteroplasty of the dilated distal ileum [11]. Thus, considering the importance of the ileocecal valve, it is necessary to

Fig. 1. Preoperative contrast enema reveals no luminal irregularity from the cecum to the rectum. There was no reflux of contrast into the terminal ileum.

Fig. 2. On delayed imaging following the UGI contrast study, there is a large smooth distal small bowel segment without further passage of contrast distally.
alter the treatment of the atresia to the specific disease and condition of the patient. In our case, the patient was otherwise healthy at the time of surgery, and she only required a short segment resection with primary anastomosis. The patient has not had any further consequence following resection of the valve, confirming that an ileocecectomy is preferred in uncomplicated cases of ileocecal valve atresia.

3. Conclusion

This report of a preterm newborn female with true atresia of the ileocecal valve, as confirmed by the histologic images demonstrating ileal and colonic mucosa on opposing sides of the obstructing membrane and the absence of inflammation or scar, adds to the very small body of literature regarding this very rare entity. Making an exact diagnosis preoperatively is unlikely. Intraluminal examination of the ileocecal region itself is mandatory so as not to overlook the lesion. If there is a history of significant prior bowel resection or the need for concurrent resection, then an attempt to preserve bowel length is preferred to optimize absorption. However, if the patient is otherwise healthy and has a normal length of intestine, ileocecectomy with primary anastomosis is the best option.

Disclosures

This manuscript does not contain patient identifying information. The authors have no conflicts of interest to disclose.

References