

Isolated ileocecal valve atresia

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Ileocecal valve atresia is a rare variety of gastrointestinal atresia. There have been only 10 cases reported in the literature. In this paper, we report the case of a 4-day-old male preterm, who presented with signs of neonatal intestinal obstruction. Contrast enema showed microcolon with no reflux of the contrast into the terminal ileum. Exploratory laparotomy showed an isolated ileocecal valve atresia. The atretic segment was resected and an ileocolic anastomosis was performed. The histopathological report confirmed the diagnosis of ileocecal valve atresia. The postoperative recovery was uneventful. *Ann Pediatr Surg* 14:211–213 © 2018 Annals of Pediatric Surgery.

Introduction

Intestinal obstruction is a common cause of neonatal morbidity and mortality. Intestinal atresia accounts for one-third of all cases of neonatal intestinal obstruction [1].

The International Incidence of Intestinal Atresia is reported to be 1 in 5000 live newborns [2], in a local study done in Saudi Arabia the incidence was 1 in 5500 live newborns [3].

Intestinal atresia most commonly affect the jejunum and the ileum [4]. Isolated ileocecal valve atresia is a rare entity with only 10 cases have been reported in the literature [5–12].

In this paper, we discuss a case of isolated ileocecal valve atresia in terms of clinical presentation, radiological features, and different modalities of surgical options.

Case summary

A preterm male neonate was delivered vaginally at 36 week of gestation with a birth weight of 1700 g and Apgar score of 7/9 in the first minute. He was referred to the pediatric surgery on the second day of life due to failure to pass meconium associated with bile-stained vomiting and abdominal distension.

The abdominal radiograph showed dilatation of the small bowel loops with a prominent central intestinal loop (Fig. 1). A water-soluble contrast enema showed a microcolon with no reflux of contrast into the terminal ileum. The contrast delineated the obstructed ileocecal valve protruding into the cecum (Fig. 2); therefore, an obstruction at the level of the ileocecal junction was suspected.

He was underwent laparotomy which showed a mildly distended small bowel, and a remarkable distention and bulking of the terminal ileal loop; however, the ileum was blindly ended and it was bulging into the cecum. Nevertheless, the length of the small bowel was found to be normal; there was no defect in the mesentery (Fig. 3). The intraoperative diagnosis was an isolated ileocecal valve atresia. The dilated bulky part of the terminal

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ileum was resected along with the atretic segment (Figs. 4 and 5) and an ileocolic anastomosis was performed. The postoperative period was uneventful.

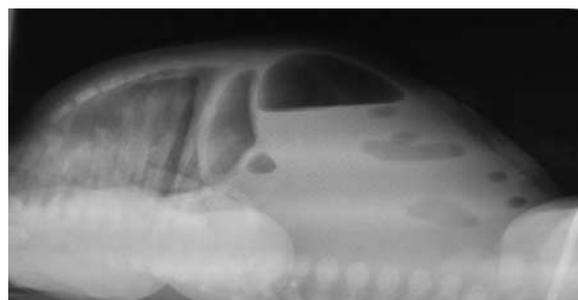
The resected ileal segment was sent to pathology. Gross examination demonstrated a common wall separating the ileum from the cecum indicative of an atresia of the ileocecal valve itself. The ileocecal valve was thickened, completely obstructed. Serial section of the ileocecal valve showed complete obstruction of the lumen with fibrosis thickening. The microscopic examination showed an ileal mucosa on one side of the obstructed valve and a colonic mucosa on the other side and there was mild submucosal and subserosal congestion.

Discussion

Intestinal atresia is a common cause of bowel obstruction in the newborn and it can occur at any point in the gastrointestinal tract. Ileocecal valve atresia is a rare entity with only 10 cases having been reported in the English literature [5–12].

Numerous variants of the atresia have been described including failure of development of the entire ileocecal region to a membranous obstruction of the valve with the

Fig. 1



Radiograph shows small bowel loops dilatation, with a significant prominent central intestinal loop.

Fig. 2



Contrast enema: microcolon with no reflux of contrast into the terminal ileum and the bulging of the obstructed ileocecal valve in the cecum.

Fig. 3



Intraoperative picture shows the microcolon (C), the bulky dilated terminal ileum (B), and the appendix (A).

presence of the appendix [6,7]. Our reported case represents the variation of fibrous thickening of the ileocecal valve and complete membranous obstruction with normal looking ileocecal junction, normal mesentery, and presence of the appendix.

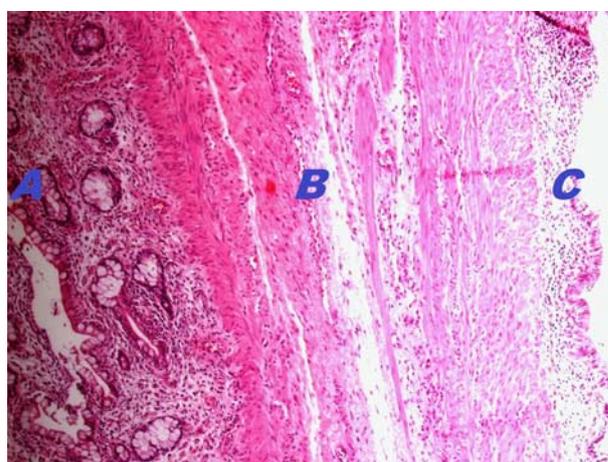
The reported cases presented as neonatal intestinal obstruction which include abdominal distension, bilious vomiting, and failure to meconium. The diagnosis of the previously reported cases of iliocecal valve atresia was made intraoperatively because of lack of specific preoperative radiological findings. However, the diagnosis in our case was suspected preoperatively as the contrast study showed intestinal obstruction at the level of the ileocecal valve. The contrast enema showed microcolon with no reflux of contrast material into the terminal ileum, delineating the obstructed ileocecal valve. The obstructed part of the ilium at the ileocecal

Fig. 4



Resected segment of the terminal ileum with obstructed ileocecal valve.

Fig. 5



Histological Section at the level of the ileocecal valve level showed complete obliteration of the lumen .
A :the obstructed ileal lumen .
B: the seromuscular layer .
C ; the ceceal mucosa fromthe other side.

Histological section at the level of the ileocecal valve showed complete obliteration of the lumen.

valve was bulging in the cecum. This radiological finding was not demonstrated in the previous reported cases.

Different surgical modalities were proposed for the reported ileocecal valve atresia cases depending on the intraoperative findings. Cacciari *et al.* [7] resected the middle part of the atretic ileocecal valve followed by ileocecal valve repair. In our case, the small bowel had a normal length so we performed a resection of the atretic segment followed by an ileocolonic anastomosis, the same surgical approach was applied by others authors [5,8,9,12]. We think that, as in all cases of intestinal atresia, the bulbous dilated preanastomotic bowel should be removed to avoid a longue post-operative ileus due to malfunctioning of this segment.

The reported outcomes of these different cases were good in our case as well.

Conclusion

This paper sheds the light on this rare entity and adds to the very small body of the literature.

It is unlikely to make a preoperative diagnosis of ileocecal valve atresia; however, a proper barium enema may make the diagnosis suspected. Therefore, the intraoperative examination of the intraluminal portion of the ileocecal region is mandatory for the diagnosis of these cases.

Conflicts of interest

There are no conflicts of interest.

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