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CASE REPORT

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Ileal atresia associated with a congenital vascular band anomaly: observations on pathogenesis

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Abstract We report the case of a newborn, who developed intestinal obstruction soon after birth. Exploratory laparotomy revealed a congenital vascular band anomaly extending from the antimesenteric border of the terminal ileum to the gallbladder in association with ileal atresia. Surgical intervention was performed for correction of the disorder. A review of the embryology and congenital vascular bands is presented together with discussion as to possible etiopathogenesis leading to small bowel atresia.

Keywords Jejunoileal atresia · Congenital vascular band · Vitelline remnants

Introduction

Congenital bowel atresia occurs in 1/330 to 1/1500 live births. Causes include mesenteric vascular accidents, failure of recanalization of the solid stage of the embryological bowel, genetic abnormalities, or hypercoagulable states [1, 2]. Occasionally, cases of congenital vascular band anomalies have been reported as causing intestinal obstruction, but none causing small bowel atresia have been reported [3, 4]. An unusual case of a congenital vascular band anomaly in association with ileal atresia is described with the evaluation of the relevant etiopathogenesis.

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Case report

A 3200-g girl was born at 36 weeks gestation to a 40-year-old healthy multigravida. The pregnancy was complicated with polyhydramnios, diagnosed by ultrasound examination at 28 weeks gestation. The follow-up ultrasound examination suggested a bowel obstruction. At birth, the Apgar scores were found to be 8 both at 1 and 5 minutes. Physical examination showed a well-appearing baby in no distress, with typical dysmorphic features of Down's syndrome. Soon after birth, she developed signs and symptoms indicative of intestinal obstruction. Abdominal radiographs suggested a low intestinal obstruction. Barium enema showed microcolon, consistent with low ileal atresia.

Exploratory laparotomy revealed type-II ileal atresia and congenital vascular band, adjacent to a Meckel's diverticulum. The proximal bowel terminated in a bulbous blind end, which was connected to the collapsed distal bowel by a very short fibrous cord along the edge of an intact mesentery (Fig. 1). A thick fibrous congenital band extended from the antimesenteric border of the terminal ileum to the distal tip of a normal appearing gallbladder, overlying and compressing the atretic segment. The band was ligated at its margins and resected. The atretic segment was resected and the continuity was restored with primary end-to-end anastomoses. Sections of the band revealed that it was composed of firm connective tissue containing blood vessels. There were no calcifications noted in the abdominal cavity as is occasionally observed with intestinal atresias.

Total parenteral nutrition was administered for a week. She was started on oral feedings after return of intestinal function. The clinical course of the patient was uneventful and she was discharged free of complaints on day 11 postoperatively.

Discussion

Intrauterine vascular accidents are commonly believed to be the cause of most small bowel atresias [1, 2]. Experimental studies have shown that interference within small bowel vasculature can cause atresia [1, 2]. A positive correlation was found between the degree of ligation of the arterial branches of small bowel and the extent of atresia [1, 2]. Moreover, it was shown that partial obstruction of the artery resulted in stenosis of the small bowel, whereas complete obstruction led to small bowel atresia [1, 2]. In the present case, however, there was no strict evidence of vascular insufficiency.

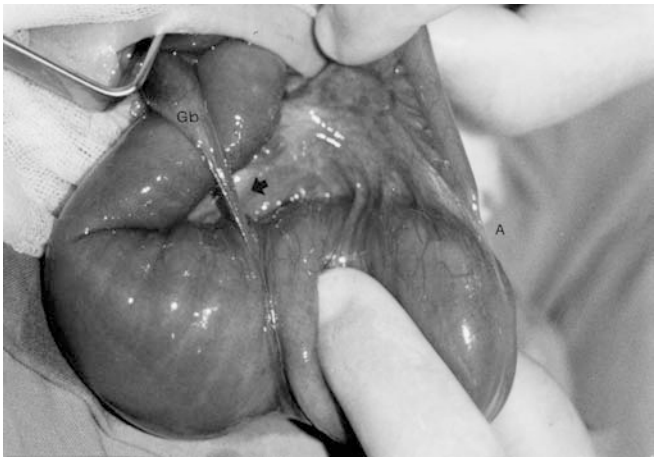

Fig. 1 Operative view of the ileal atresia (*A*). The proximal bowel is connected to the collapsed distal bowel by a very short fibrous cord along the edge of an intact mesentery. *The arrow* indicates the congenital band extending from the gallbladder (*Gb*) to the terminal ileum

The intact bowel wall and mesentery may contribute to our suggestion regarding vascular insufficiency. Also, an association with Down's syndrome may indicate that the atresia might be due to a developmental error, occurring during the early weeks of gestation, whereas vascular accidents occur at a much later stage of development [5]. Overall, vascular accidents have been reported to account for only 25% of small bowel atresias [2]. In the present case, the coincidence of the vitelline vein remnant and small bowel atresia, which has not been previously reported to our knowledge, might indicate that a mechanical factor could be another etiologic factor in small bowel atresia.

Congenital bands are not frequently encountered in pediatric surgical practice. A review of the literature suggests that these bands are mostly consistent with the persistence of the omphalomesenteric (vitelline) duct and accompanying vitelline vessels. During the fifth week of fetal life, the paired vitelline veins form a plexus around the midgut and proliferate in the cephalic direction. The growing liver cords interrupt the course of the veins and together form the hepatic sinusoids. The proximal and distal portion of the right vitelline vein forms the hepatocardiac portion of the inferior vena cava and the portal vein, respectively, while the left vitelline vein disappears during embryogenic development. The omphalomesenteric duct also gradually becomes obliterated and disappears [6]. In the present case, it was suggested that the persistence of the left vitelline vein resulted in the congenital vascular band, extending from the terminal ileum to the gallbladder, and that the failure of regression of the omphalomesenteric duct resulted in the Meckel's diverticulum. The location of the congenital vascular band was similar to those documented previously [3, 4].

Occasionally, vitelline vein remnants producing intestinal obstruction has been reported [4]. However, unlike any of these cases, the vitelline vein remnant in our case was additionally in association with ileal atresia. This unexpected association raised questions on the etiology of jejunoileal atresia. The atresia in the present case was attributed to the compression of the ileum by the vitelline vein remnant. Several authors have similarly suggested a disruptive mechanical pathogenesis. They implied that destruction of a previously normal formed bowel might lead to ischemic necrosis and subsequent resorption of the affected segment resulting in jejunoileal atresia [2, 7, 8]. It was thought most likely that the primary effect of the vitelline vein remnant on the ileum operated through just such a mechanism. Experimental work has shown that tying a ligature round the intestine of a fetal puppy can cause an intraluminal diaphragm [5]. This concept was also supported by the clinical observations of Hasegawa and Petrovski, confirming the role of mechanical accidents as the cause for some jejunoileal atresias. The authors pointed out that a persistent omphalomesenteric duct might be a cause of jejunoileal atresia by its continuous compression of the intestine [2, 7].

Although an unrelated occurrence of a congenital band anomaly and ileal atresia in this child cannot be ruled out, it seems to be very unlikely. The present case suggests that jejunoileal atresia may be caused in rare circumstances by mechanical accidents.

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