

Case Report

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Colonic and type IV jejunal atresias with apple-peel bowel segment as the content of right inguinal hernia: A case report

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KEYWORDS

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Apple peel atresia,
Inguinal hernia,
Volvulus

ABSTRACT

Background: Colonic atresia is an uncommon entity. It may be associated with multiple anomalies.

Case Presentation: We present a case, which was initially suspected to be jejunal atresia, but was found to have colonic atresia as well intra-operatively. The associated jejunal atresia with apple-peel configuration was present as the content of inguinal hernia.

Conclusion: This case report highlights the rarity of the condition and the postoperative issues in the management of such patients.

INTRODUCTION

Colonic atresia has a reported incidence of 1:20000 to 1:66000 live births and is associated with congenital anomalies in up to 47% of cases. These may be abdominal anomalies like multiple intestinal atresias, Hirschsprung's disease, malrotation, abdominal wall defect, or extra-abdominal anomalies involving the musculoskeletal system, eyes, or the face.[1]

We describe an interesting and rare case of colonic atresia, with multiple jejunal atresias, with distal small bowel apple-peel configuration as the content of the right inguinal hernia. On exhaustive search, such an association has not been described in English literature.

CASE REPORT

A 6-day male neonate, full-term normal delivery, weighing 1.61 kg, was brought with a history of abdominal distension, bilious vomiting since birth, and non-passage of meconium.

On examination, there was upper abdominal distention, and a non-obstructed reducible right inguinal hernia was present. The anal opening was present at the normal site. An infant feeding tube could be inserted into the anus and no meconium staining was observed on it. Nasogastric decompression yielded bilious aspirate.

X-ray abdomen showed few air-fluid levels which were suggestive of proximal jejunal atresia (Fig. 1). USG

abdomen, in addition to distended small bowel loops, surprisingly revealed a distended ascending colon which was filled with thick fluid suggestive of obstruction in the distal large bowel.

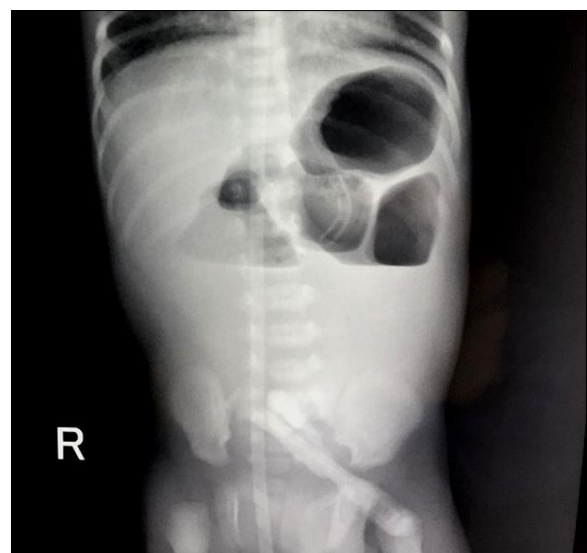


Figure 1. X-Ray Abdomen showing few air-fluid levels suggestive of jejunal atresia

After initial fluid resuscitation, an exploratory laparotomy was done. There was distended proximal jejunum, with a blind end situated 20 cm from the duodenojejunal flexure. Distal small bowel segments had apple-peel configuration based on ileocolic vessels. This apple-peel segment was herniating through the right internal inguinal ring. After reduction of con-

tents, the apple-peel segment had three more segments of the atretic bowel. Also, distally a volvulus was seen, with gangrenous changes involving terminal ileum, caecum, appendix, and ascending colon. The ascending colon was distended, and there was type III colonic atresia lying at the distal part of the ascending colon (Fig. 2). Distal to atresia, the colon was found collapsed but patent.

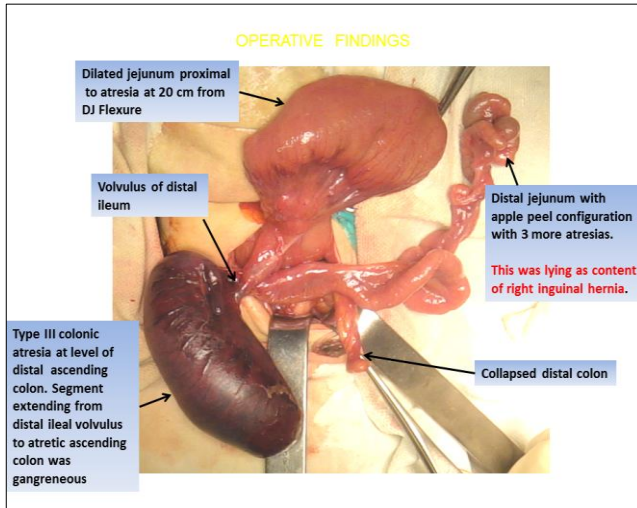


Figure 2. Intraoperative findings showing dilated jejunum, apple-peel segment, and gangrenous ileocolic segment with volvulus

Atretic distal segments were resected. Proximal dilated jejunum was imbricated and end-to-back jejunoileal anastomosis was done in a single layer using 5-0 silk. The gangrenous ileocolic segment including the colonic atresia was resected and end-to-end ileocolic anastomosis was done in a single layer using 5-0 silk. The right internal inguinal ring was closed with 3-0 polyamide using purse-string suture.

Post-operatively, the child had prolonged ileus and was kept on total parenteral nutrition. Ryle's tube feeding was initiated on postoperative day 12. Prokinetic drugs mosapride and erythromycin were also added. Feeds were gradually increased. Intravenous fluids were tapered and stopped on postoperative day 44. The patient was discharged on postoperative day 49, with a weight of 2.09 kg. Histopathology reports were suggestive of atresia and confirmed the presence of ganglion cells. The patient had a weight of 5.1 kg on follow-up after 5 months.

DISCUSSION

Colonic atresia was first described by Binniger in 1673. It accounts for less than 10% of cases of all intestinal atresias.[2] The pathogenesis of colonic atresia is not well understood. There are two main theories to explain this condition. One is the vascular incident theory, similar to one described by Louw and Barnard in 1955 for jejunoileal atresia.[3] Mesenteric vascular obstruction due to extrinsic compression from volvulus, internal hernia, or tight gastroschisis results in colonic atresia.[4,5] This may also result

from intrinsic occlusion of the vessels due to thromboembolic events originating from the placenta.[6] These are events occurring in the late gestational period.

The other theory postulates deranged morphogenesis in the early gestational period, as suggested by the absence of lanugo, bile pigments, and squamous epithelium distal to atretic segment. The field-effect of disturbed morphogenesis in the early gestational period explains the occurrence of multiple associated anomalies.[7] Other theories include defect in the fibroblast growth factor 10 (FGF10) pathway as suggested by Fairbanks in 2005.[8]

Being uncommon, colonic atresia requires a high index of suspicion if a preoperative diagnosis is to be made. Clinical presentation, X-Ray, and Ultrasound findings mimic small bowel atresia. Associated small bowel atresia as seen in our case will also influence the preoperative diagnosis. So, diagnosis is confirmed mostly on exploration.

Apple-peel atresia was first described in 1961 by Santulli and Blanc.[9] It is characterized by proximal dilated jejunum, foreshortened distal small bowel with an absence of dorsal mesentery, and absence of distal part of the superior mesenteric artery. The segment is supplied in retrograde fashion by a branch of the ileocolic artery, right colic artery, or inferior mesenteric artery. The bowel loops spiral around this feeding vessel as there is no dorsal mesentery. Apple-peel atresia is considered an early gestational defect as it has a high incidence of associated anomalies.

The index case most likely falls in the category of an early gestational event as suggested by the presence of apple peel atresia, with colonic atresia either present as an associated anomaly (early event), or developing as a sequela of distal ileal volvulus which could be a late event.

Colonic atresia is frequently seen with associated anomalies. Though colonic atresia has been described in association with multiple small bowel atresias (type IV), the simultaneous presence of an apple-peel configuration of distal small bowel segment, as observed in our patient, has never been reported in English literature. Interestingly, this apple-peel segment in our case was lying as the content of the right inguinal hernia. Even after an extensive literature search, we could not find any similar description in English literature.

To conclude, colonic atresia is associated with other anomalies in almost half of the cases. However, colonic atresia, in combination with multiple jejunal atresias with apple peel configuration of the distal small bowel, lying in inguinal hernia sac as content, has not been reported to date.

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