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Case Report

Apple-Peel Intestinal Atresia Along with Cystic Ileal Duplication Cyst in a Newborn – Successful Management of Rare Congenital Malformation

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Abstract

Association of apple peel type of intestinal atresia with duplication cyst which is rare is reported in this case.

Keywords: Alimentary tract duplications; Intestinal obstruction; Jejunoileal atresia

Introduction

Apple-peel type of intestinal atresia and non-communicating ileal duplication cyst are rare congenital malformations. A six days old neonate admitted with signs of small bowel obstruction had both the anomalies occurring simultaneously during laparotomy and was managed successfully. We are reporting this case since occurrence of both of these congenital anomalies is rare.

Case Presentation

A six-day-old male child, full-term, 2.5 kg, 1st by birth order brought to casualty with complaints of abdominal distension, bilious vomiting and non-passage of meconium since birth. On admission, child was sick and dehydrated. Abdomen was distended with visible bowel loops. No visible peristalsis.

Resuscitation was done with intravenous fluids, oxygen and Nasogastric (NG) tube aspirations. NG tube was draining bile. Erect abdominal x-ray revealed dilated intestinal loops and multiple air-fluid levels (Figure 1). After initial stabilization of hematological and biochemical parameters, emergency exploratory laparotomy was planned. Proximal jejunum was dilated and ending blindly. It was about 10 cm from duodenojejunal flexure. There was a mesenteric defect followed by type III b (apple-peel) atresia with single arterial supply and short bowel due to atresia (Figure 2).

A fluid-filled cystic lesion measuring 4 cm in diameter was found at distal end of atretic ileal segment about 10 cm proximal to ileocaecal junction. The lesion was seen to be adherent to ileal wall and shared a common blood supply (Figure 3).

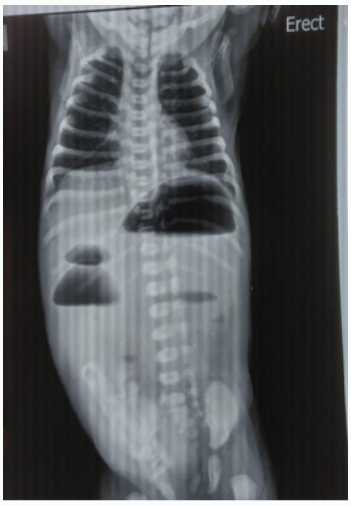

Figure 1: Abdominal x-ray showing air fluid levels with paucity of gas.

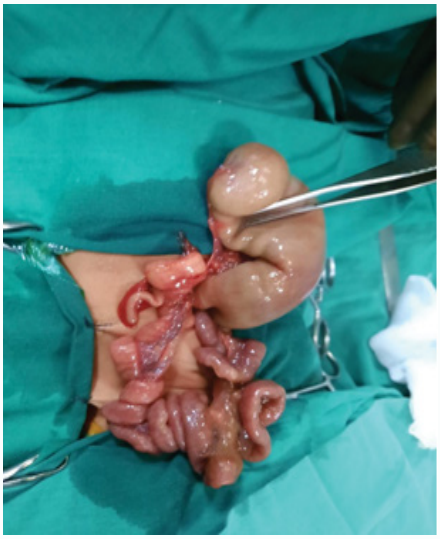

Figure 2: Showing dilated proximal jejunal loop blind ending with wide mesenteric defect.

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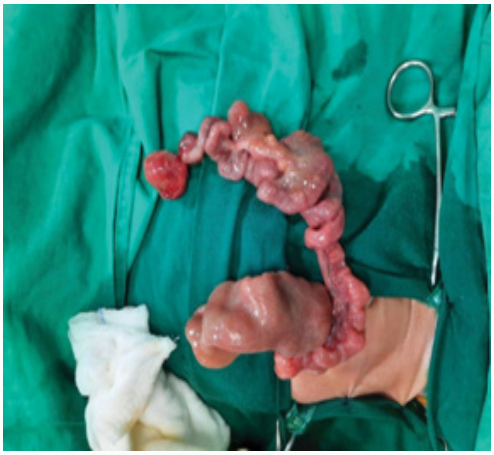

Figure 3: Showing apple peel atresia with duplication cyst.

Proximal dilated jejunum along with the duplication cyst and atretic distal ileum which was about 25 cm of small bowel was excised. The distal atretic small bowel was opened at its distal end and flushed with warm normal saline to confirm the patency. End-to-end anastomosis was done between proximal jejunum and distal atretic segment of ileum using 5-0 polyglactin sutures in single layer. It was done over feeding jejunostomy since child would remain nil per oral for a long time post-surgery due to associated hampered motility of bowel .Wide mesenteric defect was closed without hampering the blood supply. Gross pathology suggested collapsed unilocular cystic structure measuring 4 cm attached to serosal aspect of intestinal wall. The cyst contained mucoid material and cyst wall along with excised bowel was sent for histopathology. Histopathology revealed small intestinal mucosa and the wall showed organized both inner circular and outer longitudinal smooth muscle bundles (Figure 4).

Child was started on jejunostomy feeding on post-operative day five. Developed bile staining from glove drain on post op day 7 with abdominal distension. Dye study was done after putting dye through nasogastric tube which revealed dilated proximal jejunal segment which was a dynamic and a minor leak of dye in peritoneum with dye reaching up to rectum. Child was managed conservatively for five days. After septic work up was negative, child was again started on jejunostomy feeds. Gradually feeds were started from nasogastric tube. Child tolerated feeds with no vomiting or abdominal distension. Passed formed stools. Nasogastric tube was removed after the output decreased and became non-bilious. Child was discharged on twenty ninth postoperative days on full breast feeds. The baby is under follow up since last three months with nutritional advice. He is tolerating breast feeds and gaining weight. Feeding jejunostomy tube was removed on follow up.

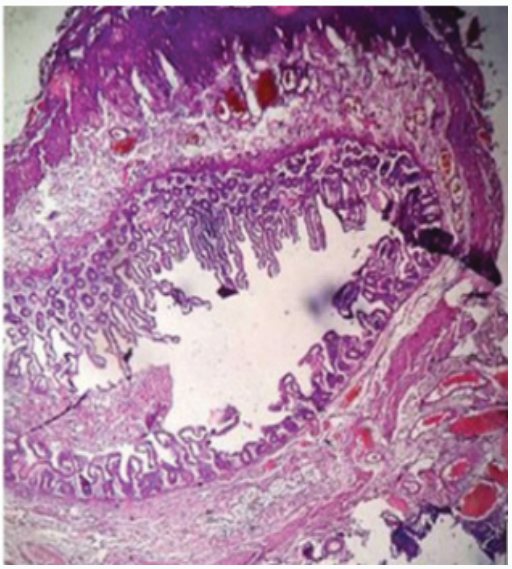

Figure 4: Microscopic picture of duplication cyst revealed normal small intestinal mucosa with flattened villi and the wall showed organized inner circular and outer longitudinal smooth muscle bundles.

Discussion

The incidence of bowel duplication cysts along with jejunoileal atresia is 0% to 4.7%. Conversely, the incidence of jejunoileal atresia in series of jejunoileal duplications is about 10%.

About third of cases of intestinal obstruction in neonates are due to intestinal atresia. As we all know, apple-peel or type IIIb Jejuno Ileal Atresia (JIA) is an uncommon variant in Grosfeld classification system [1]. It is also called Christmas-tree or Maypole deformity. There is proximal jejunal atresia along with large mesenteric defect with loss of bowel leading to short bowel [2]. The distal atretic small intestine has a helical appearance around one vessel in a retrograde fashion which may be inferior mesenteric artery in most of the cases. It has familial pattern of association and may occur with other congenital malformations. This variety of atresia has highest mortality rate (71%) among all types of JIA [3]. Prognosis in JIA has improved due to advances in management, but it is a challenge due to its wide defect in mesentry [4].

Alimentary tract duplication is rare congenital malformations that can arise anywhere in the gastrointestinal tract, the most common location being ileum. It is epithelial-lined structure attached to the intestinal wall and supplied by mesenteric vessels [4]. It may or may not communicate with intestinal lumen depending on the type of duplication. Duplication cysts will lead to intestinal volvulous, perforation and there are chances of malignancy as well [5]. Most of them are located on the mesenteric side of bowel as opposed to Meckel diverticulum, which occur on antimesenteric side of bowel. Mesenteric cyst is another differential diagnosis. Difference can be made on histopathology since mesenteric cysts have flattened to cuboidal epithelium without organized muscle coat and absence of nerve plexus, whereas duplication cysts generally shows organized muscle coats [6]. Excision of duplication cyst along with the bowel is the standard operative management due to their common blood supply. A duplication cyst leading to mesenteric volvulus followed by vascular compromise has been proposed in the pathogenesis of intestinal atresia [7]. Apple-peel type of atresia may also occur due to antenatal vascular accidents [8].

Management of ileal atresia is laparotomy with excision of dilated segment and either end to end or end to side anastomosis [9]. In cases of duplication cyst associated with ileal atresia with lumen disparity, Bishop-Koop ileostomy is a better option along with resection anastomosis at duplications sites in cases where the site of intestinal atresia is far away from the site of duplication.

Conclusion

Apple-peel type of intestinal atresia may occur along with duplication cyst. A non-communicating duplication cyst needs to be

differentiated from mesenteric cysts. Good surgical technique and postoperative care are essential for salvage of these neonates. The separate resection-anastomoses may be done in a few cases where there may be multiple atresias with normal bowel in between. Decision about management has to be taken based on individual case findings on exploration, condition of baby and gestational age of neonate.

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