

Case Report

An unusual cause of intestinal obstruction: Intramural cystic duplication of gut at Ileocecal Junction

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Abstract

Duplication of gut is a rare congenital anomaly of controversial etiology. Its presentation is variable and preoperative diagnosis is rare, even per-operative diagnosis may be difficult.

A 1 year 4 months old female child of consanguineous marriage with delayed mile-stone presented with features of acute intestinal obstruction. Past history of recurrent distention of abdomen and occasional vomiting were present. On exploration, intramural cystic mass in the terminal ileum was found to occlude the ileocaecal opening as a ball valve. No other mass was present in the gastro-intestinal tract. Resection and anastomosis were done. Histological study showed a cystic type of duplication of gut. Sequestration of intestinal mucosa is likely cause.

Keywords: Duplication of gut, duplication of intestine, intramural cyst of intestine, intestinal obstruction

1. Introduction

Acute intestinal obstruction is a common surgical emergency in children. Bands, adhesions, intussusceptions, volvulus etc are the usual causes. Duplication of gut is rare¹. Pre-operative diagnosis is difficult. Ultrasound scan is recommended. Out of 676 cases of pain abdomen in children one was diagnosed as duplication of gut by ultrasound study². Correct preoperative diagnosis by means of imaging techniques is rare¹.

2. Case report

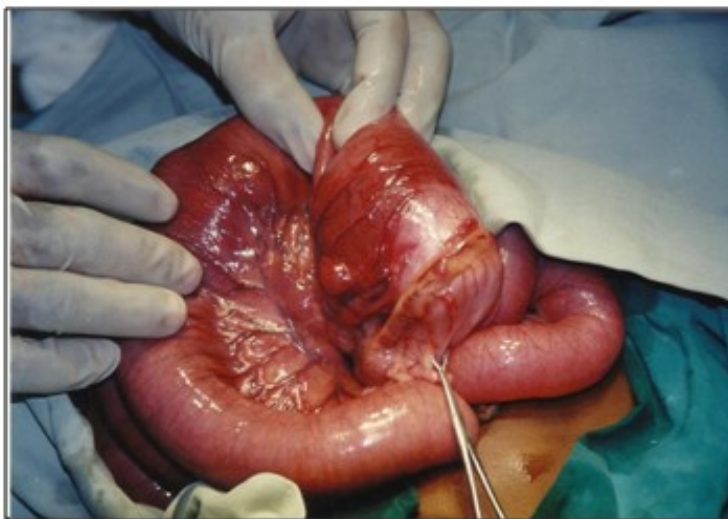
A 1 year 4 month old female child of consanguineous marriage with delayed milestone presented with complaints of intermittent distension of abdomen associated with vomiting for 7 days. Past history of recurrent distension of abdomen and occasional vomiting were present.

On examination she was anemic. Abdomen was distended and tender with increased bowel sounds. There was no localization of tenderness.

2.1 Investigations: Haemoglobin was 7 gm%. X-ray abdomen showed multiple fluid levels with distended small intestine. Ultrasound study confirmed the diagnosis of acute intestinal obstruction but failed to detect any cyst separately from the fluid filled intestine.

2.2 Surgery: On exploration site of obstruction was located at ileocecal region. There was no intussusception and mass was felt in the terminal ileum causing intestinal obstruction. (Figure 1)

Figure 1- Location of obstruction at ileocecal region



Resection of terminal ileum and caecum was done and ileo colic anastomosis was carried out.

Histopathology revealed, resected specimen consists of 7 cm of terminal ileum, 8 cm caecum and 6 cm long appendix with 0.3 cm diameter. There was a 3 cm diameter cyst at ileocecal junction projecting into the ileum at mesenteric border. The mucosa of the ileum, caecum and the luminal surface of the cyst showed multiple superficial ulcerations. The cyst contained about 10 ml of mucoid material. The cyst wall was smooth and glistening with a few areas showing congested mucosa like folds. (Figure 2,3,4)

Figure 2- Resected specimen consists of part of terminal ileum, caecum and appendix

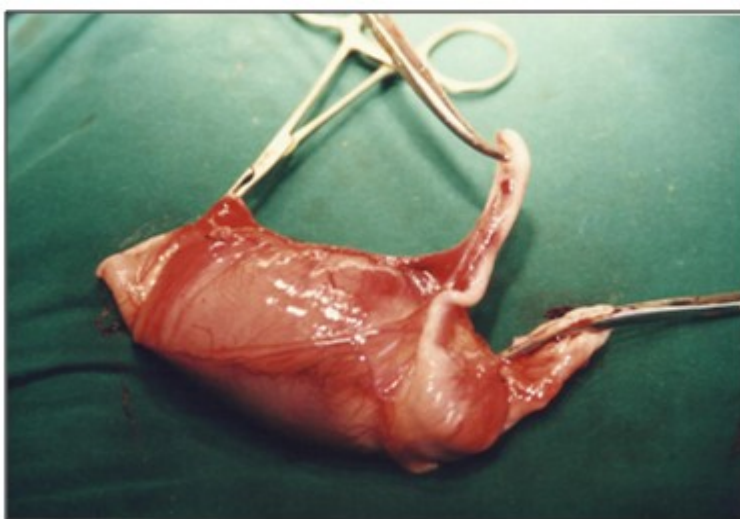


Figure 3- Cyst at ileocecal junction projecting into the ileum at mesenteric border

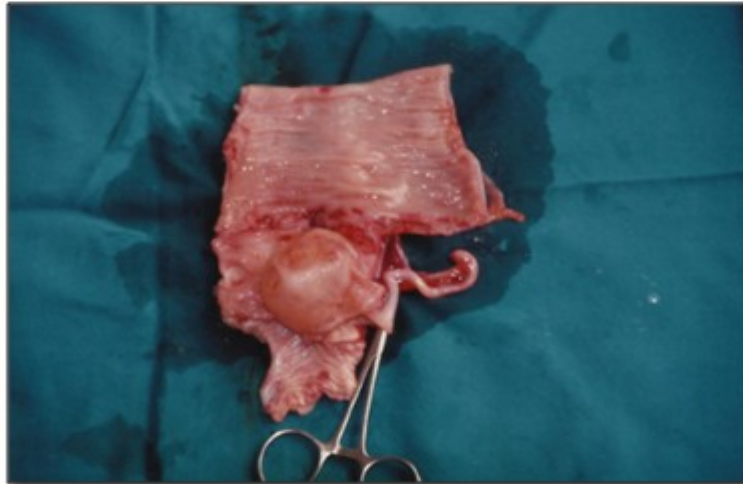


Figure 4- Aspiration of the Cyst



Microscopic examination showed the cyst lined mainly by flattened epithelial cells. At places columnar epithelium with glands (intestinal epithelium) was seen. Submucosa and muscular coats were normal. The luminal portion of the cyst showed central muscular layer with intestinal mucosa on both sides. Mucosa of the terminal ileum and caecum showed multiple nonspecific ulcers with mixed inflammatory infiltrate.

3. Discussion

Duplication of gut is cystic or tubular structures that are in intimate with portion of intestine³. The tubular duplication usually communicates with the adjacent intestines, whereas the cystic duplication tends to be completely separate. The mucosal lining is always representative of some part of the alimentary tract. The presence of gastric mucosa has the risk of peptic ulceration with the possible complication of haemorrhage or perforation. Cystic duplication is endothelium lined structure enclosed in a common muscular wall with its adjacent segment of the bowel. Triplication of gut-embryogenesis of tubular and cystic small intestine duplication has been reported⁴. Rarely cyst can attain huge size⁵.

There is no unified embryologic explanation for all duplications^{3,4}. Lewis and Thyng in 1907 postulated the remnants of the outpouching of the developing fetal intestine between sixth and eight weeks of intrauterine life between the leaves of mesentery. Bremer in 1944 suggested error of recanalization of epithelial plug. This theory is based on the

hypothesis that various portion of the alimentary tract become completely occluded by epithelial plug. Failure of canalisation by coalescence of vacuoles could conceivably result in the formation of duplication cysts. There is no convincing evidence to support the occlusion theory, and it fails to explain why duplications are restricted to the mesenteric aspect of the intestine. In 1954 Fallon et al proposed that duplication may result from failure of normal separation during the fourth week of development of the gut endoderm and the neural tube. Should complete separation of these two germ cell layers fail to occur, a cord of cells from the dorsal aspect of the developing gut will be pulled backward towards the developing vertebral column. This results into duplication of gut and explains the association of vertebral anomaly⁶. Rare association of tubular duplication of colon with subglottic stenosis is also reported⁷. In this case sequestration of intestinal mucosa is likely cause.

Resection and anastomosis is the choice of operation. This procedure is not applicable in situation in which excision will endanger structures like duodenum, where marsupialisation is carried out⁶.

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