Case Report

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Noncommunicating multiple intra-abdominal enteric duplication cysts

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ABSTRACT

A very rare case of noncommunicating multiple intra and retroperitoneal enteric duplication cysts (EDCs) is reported and discussed. Two large noncommunicating EDCs, one within the mesentery of proximal jejunum causing complete luminal obstruction and other isolated cyst in retroperitoneal area displacing duodenum and extrahepatic biliary system, were resected successfully in a 2-day-old neonate along with correction of malrotation.

Keywords: Enteric duplication cyst, gut development, malrotation of gut, retroperitoneal cyst

INTRODUCTION

Enteric duplication cysts (EDCs) are rare (1 in 4,500), congenital anomalies that may originate anywhere in the gastrointestinal tract (GIT).[1] EDCs are hollow, epithelial-lined, and cystic or tubular structures that are attached with the wall of the gut and supplied by mesenteric vessels. EDCs are more common in small bowel, either run parallel to the small bowel (tubular) or may lie within the mesentery (spherical) and may or may not be communicating with the adjacent bowel segment.[2] EDCs in retroperitoneum are very rare and till date only seven cases have been reported in English literature.[3-8] These cysts are isolated, do not communicate with GIT, and have their own blood supply.[9] Concomitant occurrence of intra and retroperitoneal EDCs is extremely rare and as per knowledge has not been reported yet. We present first case of noncommunicating intra and retroperitoneal EDCs associated with malrotation of gut in a 2-day-old neonate.

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CASE REPORT

A preterm twin, 35-week gestation age, was delivered in our hospital. Antenatal scan showed bubble stomach and there were three intra-abdominal cystic lesions near to the stomach. At birth, baby's APGAR was 6 at 1 min and 8 at 5 min. He was transferred to neonatal intensive care unit (NICU) for monitoring and observation. His abdominal examination was unremarkable and he passed small amount of meconium immediately after delivery. He had moderate volume of light yellow color aspirate from his nasogastric tube. His plain abdominal X-ray showed dilated stomach, duodenum, and proximal small bowel with no gas in the rest of the abdomen [Figure 1]. His clinical and radiological findings were consistent of proximal small bowel obstruction (small bowel atresia). He underwent laparotomy, which showed a 5×7 cm fluid filled cyst in the mesentery of proximal jejunum, compressing small bowel lumen and causing complete obstruction [Figure 2]. There was another 6×3 cm isolated cyst located in the right upper quadrant, which was also fluid-filled and extending in the retroperitoneum, displacing the duodenum forward. The cyst was also causing anterior displacement of

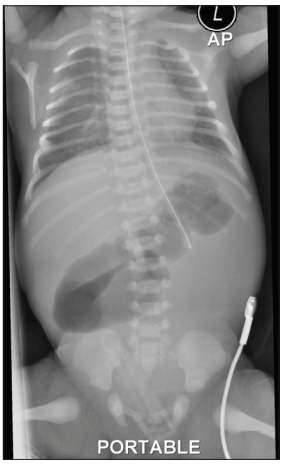


Figure 1: Plain X-ray of abdomen showing gas distended stomach and duodenum; and no evidence of gas filled bowel loops distal to it

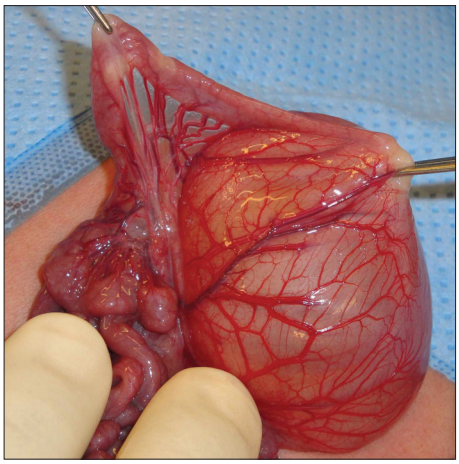


Figure 2: Enteric duplication cyst located at the mesenteric border of proximal jejunum

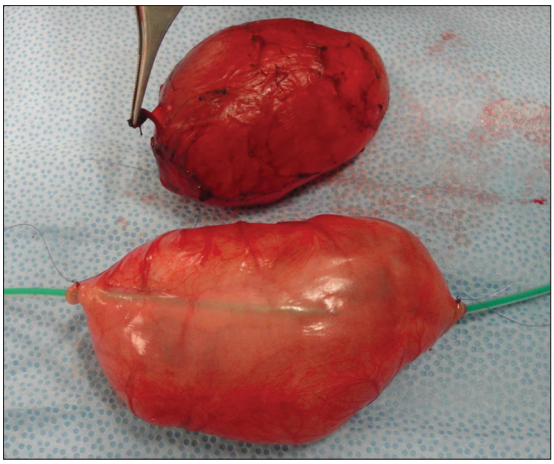


Figure 3: Completely resected retroperitoneal enteric duplication cyst (top) and enteric jejunal duplication cyst including resected segment of jejunum and feeding tube is passed through the jejunal segment showing no communication of gut with the cyst (bottom)

the extrahepatic biliary system but not occluding the lumens of ducts. The retroperitoneal cyst had a small vascular pedicle coming from mesenteric vessels. The caecum and appendix were malpositioned, lying in the midline. The proximal jejunal mesenteric cyst was excised along with the attached piece of bowel followed by an end-to-end anastomosis. The isolated retroperitoneal cyst was also excised completely [Figure 3] and Ladd's procedure was performed to correct malrotation. To confirm the communication between jejunal mesenteric cyst with the lumen of resected jejunal segment attached with cyst, a size 8 Fr feeding tube was passed from one end to other and no communication was noted [Figure 3]. Patient's postoperative course was uneventful. An ultrasound abdomen was done a week postoperative to delineate hepatobiliary anatomy and also to document any residual/additional retroperitoneal cysts, which did not reveal any abnormality. The histological examination of both cysts revealed cyst wall lined by well formed small bowel type mucosa, submucosa, circular, and longitudinal layers of muscularis mucosa confirming small bowel EDCs.

DISCUSSION

EDCs are rare congenital anomalies, which can occur anywhere in the GIT from mouth to anus, although there is greater predilection towards the ileal region.[10] The hallmark of diagnosis is the close proximity of the cyst to some part of the alimentary tract with sharing its blood supply. In isolated EDCs, there is no definite communication or connection with the nearby GIT. However, the presence of the typical histopathological features like distinct muscle wall layers and lining epithelium would qualify for the diagnosis of noncommunicating duplication cysts.[11] In our case, the findings correlate with the above explanations of EDCs, as we encountered two EDCs, one situated within the mesentery of proximal jejunum with no luminal communication, and other cyst was located in retroperitoneal area with its separate vascular pedicle and not connected with the nearby gut segment.

Embryogenesis of alimentary tract duplications cysts is unclear. Several hypotheses for formation of EDCs have been postulated including diverticulization, canalization defects, intrauterine vascular accident theory, split notochord theory, and the traction hypothesis between intestinal endoderm and the overlying structures.[12-14] These do not explain the formation of extraperitoneal EDCs. We propose that a part of fetal gut may get sequestered at a very early stage of development in the extraperitoneal space such as retroperitoneum or chest to develop into retroperitoneal or mediastinal EDC and take its vascular supply from gastrointestinal vascular tree. We also suggest that in intraperitoneal EDCs, occurrence of malrotation of gut may be a contributory factor by causing lymphatic obstruction of a part of embryonic gut, thus resulting in formation of noncommunicating cyst. This hypothesis may well explain the occurrence of two noncommunicating EDCs at two different anatomical locations in our patient, one in the retroperitoneum with separate blood supply and other within the mesentery of small bowel along with associated malrotation of gut.

The clinical presentation of EDCs depends on anatomical location, size, and inner lining of the cyst wall. In previously reported seven cases of retroperitoneal EDCs, 60% of these cysts were picked up and managed in neonatal period[3,6,10], whereas the remaining 40% cases presented late with delayed complication of obstruction and jaundice in adulthood.[4,8,9] In all these cases, the cysts were located in upper left quadrant and contained gastric mucosa. Compared to previously reported cases,

in our patient the location of retroperitoneal EDC was on right side close to duodenum and did notcontain gastric mucosa. Unlike other previously reported cases of retroperitoneal EDCs, in our case, there was a second noncommunicating circular EDC in the mesentery of proximal jejunum and also associated malrotation of gut.

The treatment of all intra and extraperitoneal EDCs is complete resection either by open transperitoneal or retroperitoneal approaches. Recently laparoscopic techniques have been used successfully in the excision of intraperitoneal EDCs in infants.[15] However, in our case we used open transperitoneal approach to completely excise two EDCs at a time.

Awareness of varied types and possible anatomical locations of EDCs is essential and would be helpful to choose optimal early surgical intervention.

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