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Isolated Enteric Duplication Cyst: A Rare Congenital Anomaly

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ABSTRACT

Introduction: Gastrointestinal tract duplications are uncommon congenital abnormalities, commonly present as cysts. By definition, they are located in or adjacent to the wall of part of the gastrointestinal tract, have smooth muscle in their walls, and are lined by alimentary tract mucosa. Duplication Cysts are most commonly located in the distal ileum; sometimes, they are associated with other visceral or skeletal anomalies. They are frequently discovered during childhood, following a bowel obstruction or abdominal pain.
Materials and methods: A 19-day-old Indian boy presented with abdominal distension and a chief complaint of regurgitation of feeds since birth. Contrast-enhanced computed tomography showed a large hypodense cystic lesion in an abdominal cavity.
Results: Surgical exploration and pathologic specimens resulted in the diagnosis of an isolated enteric(ileal) duplication cyst.
Discussion: An enteric duplication cyst must be ruled out for repeated episodes of feeding regurgitation, especially in an infant. Diagnosing this case was challenging, and imaging tests could not identify whether the mass was in the retroperitoneum or the anterior peritoneum. For such cases, surgical exploration is necessary to make a definitive histopathological diagnosis of this rare congenital anomaly as it can present in various clinical forms. It can cause significant morbidity and even mortality if left untreated by causing life-threatening complications.

Keywords: congenital, duplication cysts, surgical exploration, histopathological

INTRODUCTION

Enteric duplication cysts are rare developmental congenital anomalies, originating anywhere along the alimentary tract from the tongue to the anus. The reported incidence is 1:4,500 births [1]. Most duplications are detected in infants and children (antenatally or within the first two years of life) [2,3]. They pose a diagnostic dilemma and therapeutic challenge, as the presenting symptoms are non-specific and variable, closely related to

the cyst's type, size, and location. Approximately 75% of duplications have been reported to be located within the abdominal cavity, whereas the remaining is intrathoracic (20%) or thoracoabdominal (5%). Ileal lesions are the most common (53%), followed by mediastinal (18%), colonic (13%), gastric (7%), duodenal (6%), rectal (4%), oesophageal (2%) and cervical (1%) lesions [4]. In terms of appearance, they are tubular or spheric cysts that are situated inside or close to a portion of the digestive system. They tend to be situated on the mesenteric aspect of the alimentary canal, sharing a common muscular wall and blood supply but having a separate mucosal lining [5]. Microscopically, gastrointestinal tract duplications contain smooth muscle in their walls and are lined with alimentary tract mucosa. The lining mucosa, however, is not necessarily that of the adjacent segment of the gastrointestinal tract. The walls of these lesions at various levels of the gastrointestinal tract include ectopic tissue, including ganglion cells, lymphoid aggregates resembling peyer patches, pancreas, gastric, squamous, transitional, and ciliated Mucosa [6]. On this basis, gastrointestinal tract duplications are named according to the portion of the gastrointestinal tract that they are adjacent to, rather than by their lining mucosa [7]. They may present as solid or cystic tumors, intussusception, perforation, or gastrointestinal bleeding. Therefore, in these situations, a high index of suspicion is necessary. Although enteric duplication cysts can be discovered accidentally during surgery, prior radiological detection is usually attainable in most cases. [4]. Sufficient surgical intervention is necessary, and the attending surgeon must possess knowledge of the pathophysiology and clinical attributes of these uncommon cysts. Due to the rarity of this condition, few case reports have been published from India.

CASE PRESENTATION

Here we report a case of a 19-day-old male, who was admitted to the hospital, with a chief complaint of abdominal distension, and regurgitation of feeds since birth. An abdominal CT scan revealed a large cystic mass measuring 11.3×11.3×5.5 cm **(Figure 1)**, extending from epigastrium to hypogastrium, attached to the ileal segment responsible for an ileal obstruction. No genitourinary or spinal abnormalities were detected, nor was there any contrast enhancement. During the surgical procedure, the patient underwent exploratory laparotomy with a complete resection of the cyst along with the attached ileal segment, followed by an ileo-ileal anastomosis.

Gross examination of the resected segment showed a thin-walled cystic mass measuring 20 cm in length, and filled with mucinous fluid, closely attached to a 05 cm long intestinal segment **(Figure 2, 3);** No perforation, atresia, or communication between the cyst and the ileal lumen on evaluation of the cystic wall or the normal ileal segment identified. Histological examination of the cyst wall shows the presence of enteric (ileal) mucosa **(Figure 5)** along with well-defined muscular walls (muscularis propria), nerves, and ganglion cells (myenteric plexus) **(Figure 4).** Diagnosis of an Enteric duplication cyst was made. On follow-up, the infant is doing well after surgery.

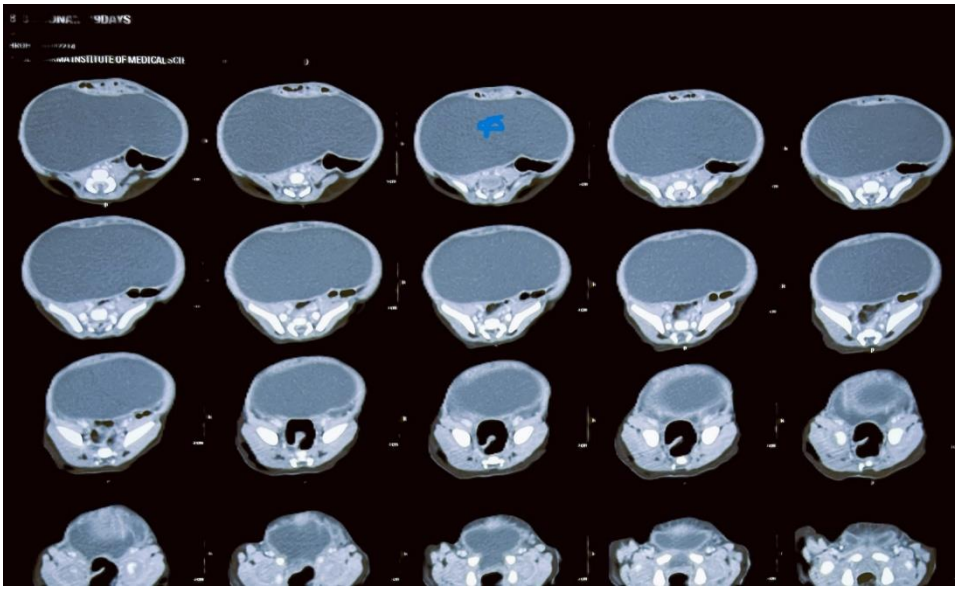

Figure 1: Abdomino-pelvic CT scan showed a 11x11x5.5 cm sized, non-enhancing, thin-walled, fluid-filled, cystic lesion (blue asterisk) in the lower abdomen.

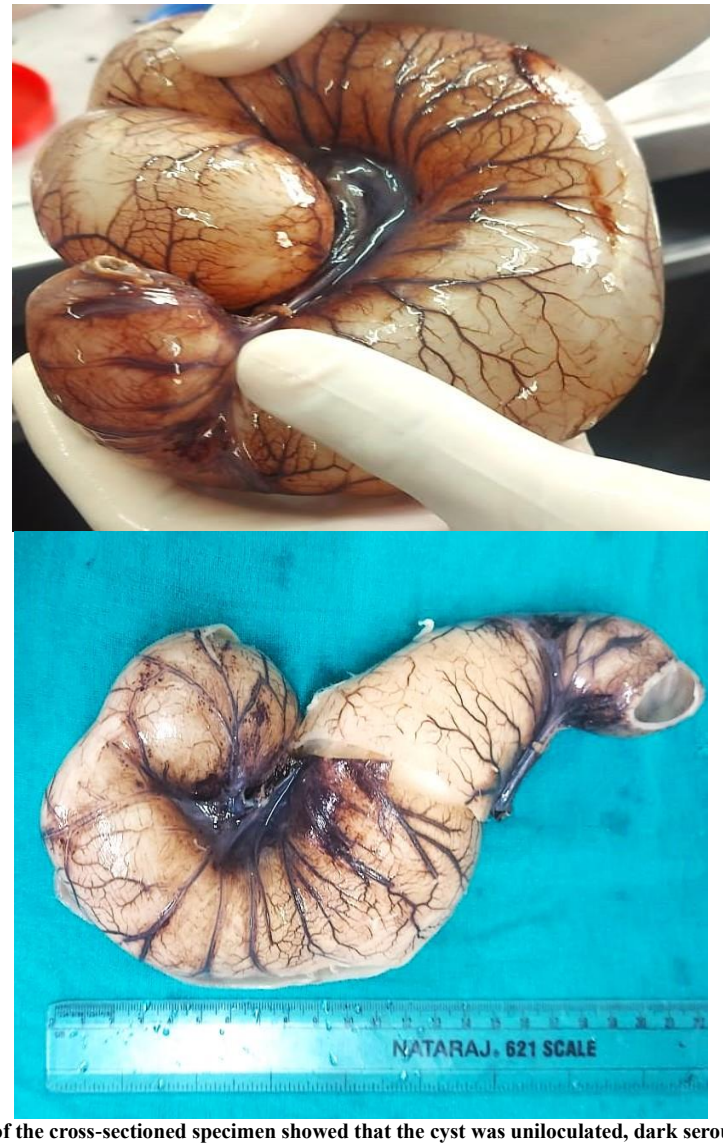

Figure 2,3: Gross finding of the cross-sectioned specimen showed that the cyst was uniloculated, dark serous fluid-filled, and attached to the ileum without solid portion or irregular wall thickening.

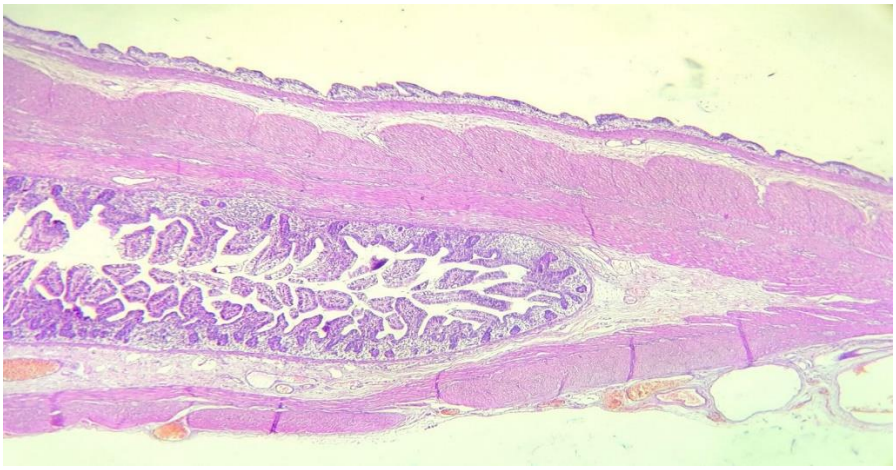

Figure 4: Microscopic findings: Two distinct muscular layers with myenteric plexus and a serosal layer attached to the small bowel wall (H&E, ×100).

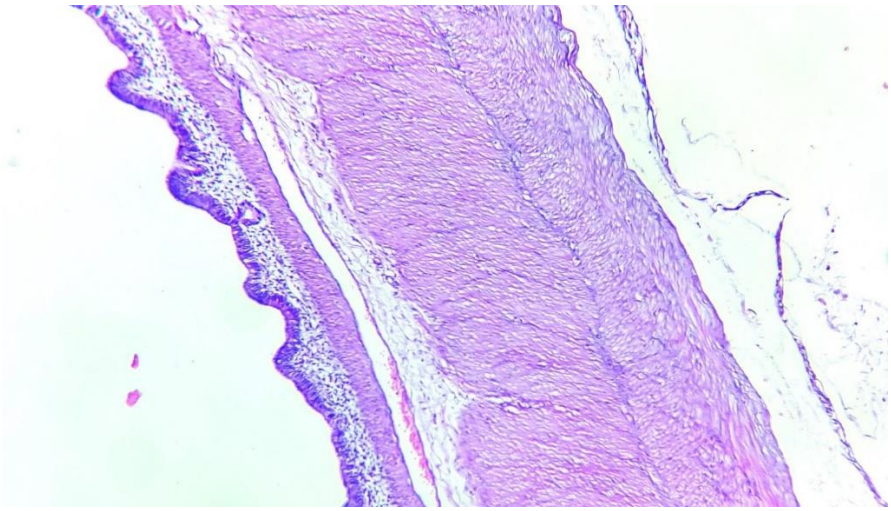

Figure 5: The cyst lined by tall columnar cells (H&E, ×200).

DISCUSSION

Intestinal Duplication Cysts (DCs) are rare cystic congenital malformations that communicate rarely with the intestinal lumen [8]. They can be diagnosed in ultrasound antenatally in the early weeks of gestation [9]. DC could be found at any segment along the digestive tract from the oral cavity to the anus [10], it occurs most commonly in the distal ileum, followed by the colon, esophagus, stomach then the duodenum [11]. Depending on its size, location, and the kind of epithelial lining, it can cause a wide range of digestive symptoms; clinical manifestations include mild stomach pain, nausea, vomiting, and abdominal mass to serious symptoms such as gastrointestinal ulcerations, bleeding, or anemia; It could also be revealed due to complications such as bowel obstruction, perforation, or peritonitis [6]. Most cases are revealed during the first two years [9]. DCs are subdivided into tubular and cystic types, the latter is more frequently encountered in the small intestine. They can be communicating or non-communicating with the adjacent intestinal segment [12]. Skeletal malformations are frequently associated with foregut DCs, whereas genitourinary and other gastrointestinal anomalies are associated with midgut and hindgut DCs [6]. Duplication cyst in our case was non-communicating to the adjacent ileal segment, with no evidence of skeletal or genitourinary deformities.

Histologically, the alimentary epithelium lining the cyst wall most often resembles that of the adjacent digestive segment; heterotopic linings that are distinct from the adjacent segment are less prevalent. The rest

of the cyst is composed of a smooth muscle layer, most often shared with the adjacent digestive segment [12]. Digestion-related DCs have been explained by several ideas, such as aberrant recanalization and partial twinning, which account for the DCs' occurrence in both the esophagus and anorectal regions as well as their connection to genitourinary tract abnormalities [13]. Persistent diverticula theory explains small intestine DCs. The split notochord theory explains associated spinal anomalies with enteric DCs [14].

The differential diagnosis includes all causes of neonatal bowel obstruction including volvulus, intussusceptions, mesenteric or omental cysts, a pancreatic pseudocyst, choledochal cyst, [5,15] and abdominal lymphangioma. Diagnosis usually uses imaging modalities such as barium studies, USG, or CT scans. Upper gastrointestinal study and barium enema demonstrate filling defect or rarely a communication between the cyst and normal bowel [16]. The preferred imaging technique for assessing an abdominal mass in a neonate is ultrasound, which can demonstrate its nature and location. Using scans during pregnancy results in a higher rate of antenatal detection of duplications (approximately 30%), allowing early treatment and avoiding possible complications [3]. CT scans are more useful in demonstrating the precise anatomical relationship between the cysts and surrounding structures [17].

The histopathological examination helps us to confirm the diagnosis. In the present report also, histopathological examination played an important role in confirming the diagnosis. Intestinal duplications often require urgent surgical intervention. Treatment of asymptomatic cases remains controversial, though they have to be removed to avoid late complications like malignant change. In the present case, the patient was managed by surgical resection, followed by anastomosis. The post-operative recovery and follow-up period was uneventful.

CONCLUSION

Digestive DCs are rare developmental abnormalities, affecting the pediatric population more frequently, they are most commonly located in the distal intestine and usually lined by an alimentary epithelium It is crucial to be aware and make a definitive clinical and histopathological diagnosis of this rare congenital anomaly as it can present in various clinical forms and can cause significant morbidity and even mortality if left untreated by causing perforation, obstruction, bleeding, and malignancy.

Declaration by Authors

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