Print ISSN: 2321-6379
Online ISSN: 2321-595X

Case Report

Enteric Duplication Cysts: Variations in Presentation and Management

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Abstract

Enteric duplication cysts (EDC) are rare congenital malformations that are formed during the embryonic development of the gastrointestinal tract (GIT). They can be present anywhere along the GIT and hence give rise to their varied presentations. The aim of this study is to observe the varied presentations of EDCs and different modalities of treatment available for the same. This is a single institution case series analysis where six cases of EDCs that presented from January 2018 to January 2023 are being retrospectively analyzed. This study concludes by stating that every case of EDC should have an individualized approach and curative management can be done with surgical resection either through an open or the more advanced minimally invasive laparoscopic or thoracoscopic approach to avoid immediate and long-term complications.

Key words: Congenital malformations, Enteric duplication cysts, Foregut duplication cysts, Minimally invasive

INTRODUCTION

As per Ladd, the term "Enteric duplication cyst" will be applied if a congenital lesion has a coat of smooth muscle, gastro-intestinal type epithelial lining and an intimate anatomical location with some part of gastro-intestinal tract.[1] These cysts can arise from any part of gastrointestinal tract (GIT) from oropharynx to anus. Depending on the site of origin, they are classified into foregut, midgut, and hindgut cysts.[1,2] Enteric duplication cysts (EDC) pose a diagnostic dilemma and a therapeutic challenge, as the presenting symptoms are non-specific and variable and are closely related to the type, size, and location of the cyst. Most commonly EDCs are found in the ileum (33%), followed by the esophagus (20%), colon (13%), jejunum (10%), stomach (7%), and duodenum (5%).[3-6] In our study, we have six cases, four of which are foregut duplication cyst and two are midgut duplication cyst. Two among the six cases had presented to the emergency department as acute intestinal obstruction and right lung abscess, respectively. The aim of our study is to highlight the varied presentations, possibility of ectopic tissues, and multiple duplication cysts along with optimum results obtained with minimally invasive surgeries.

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www.ijss-sn.com

Month of Submission : 11 - 2024
Month of Peer Review : 12 - 2024
Month of Acceptance : 01 - 2025
Month of Publishing : 01 - 2025

MATERIALS AND METHODS

This is a type of descriptive retrospective study, carried out in the Department of General surgery and Department of Pediatric Surgery, St Martha's Hospital, Bangalore, India, from the period of January 01st, 2018, to January 01st, 2023. We did a retrospective analysis of six patients diagnosed with EDC based on radiological features, operative findings, and histopathological report. The data were collected from the medical records department and analyzed accordingly.

CASE SERIES

Case 1

A 10-month-old male infant diagnosed antenatally of cystic lesion in the abdomen presented with intermittent abdominal pain. On examination, fullness of the left flank was noticed. Ultrasonography (USG) abdomen reported a cystic mass of approximately 8 × 8 cm with possibility of EDC or a mesenteric cyst. On elective laparotomy, a jejunal duplication cyst of 8 × 8 cm, over the mesenteric

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border, sharing a common wall with the adjacent normal bowel was seen. Subserosal dissection of the cyst was done leaving behind the serosal layer, which was in continuation with jejunum [Figure 1a and b]. Histopathology (HPE) was consistent with jejunal duplication cyst. The patient is doing well on follow-up.

Case 2

A 3-month-old male infant presented to the emergency with history of vomiting, abdominal distension, and decreased activity for 1 day. On examination, abdomen was distended with palpable intestinal loops. X-ray abdomen showed multiple air fluid levels and contrast-enhanced computed tomography (CECT) abdomen pelvis reported a retroperitoneal cyst with features of small bowel obstruction. The patient was taken up for an emergency laparotomy with findings of two small bowel EDC, one being 5 × 3 cm ileal duplication cyst, 10 cm proximal to the ileocaecal junction causing volvulus and a gangrenous bowel segment. Resection and anastomosis of the gangrenous bowel segment with the duplication cyst was done. Another 4 × 3 cm jejunal duplication cyst was seen 20 cm distally from the duodeno-jejunal flexure, which was excised [Figure 2a and b]. HPE confirmed, ileal duplication cyst attached to the gangrenous segment as well second jejunal duplication cyst. This case also highlights the possibility of multiple duplication cysts. The patient has been doing well on follow-up.

Case 3

A 2-year-old girl presented with complaints of intermittent episodes of abdominal pain since 2 weeks with an antenatal USG report suggestive of an EDC. CECT abdomen pelvis showed a gastric duplication cyst along the greater curvature of the stomach with no luminal communication. The patient was planned for elective laparoscopic procedure, which showed a 4.8 × 4 cm cyst over the greater curvature compressing the duodenum. Subserosal dissection was done leaving the serosa of the cyst with the gastric wall behind [Figure 3a and b]. HPE was consistent with gastric duplication cyst also showing ectopic pancreatic tissue. The patient is doing well on follow-up.

Case 4

A 3-year-old boy presented with history of fever, productive cough, and vomiting for 3 months. On examination, child was severely malnourished with decreased air entry of the right lower lobe of lung. Computed tomography (CT) scan revealed a right lower lobe of lung abscess. Child was planned for the right lower lobectomy and intraoperatively, Bengal gram seeds with other vegetative materials were seen within the abscess cavity, which was involving the whole of lower lobe. Exploration revealed a foregut duplication cyst, communicating with the esophagus and eroding into the right lower lobe of the lung. A right lower lobectomy with excision of the duplication cyst and repair of the esophageal wall was done [Figure 4a and b]. HPE report was consistent with esophageal duplication cyst. The patient is doing well on follow-up.

Case 5

A 8-month-old female infant came with two episodes of pneumonia treated conservatively in an outside center and was evaluated for recurrent lower respiratory tract infections. The patient was referred to our hospital with CECT chest

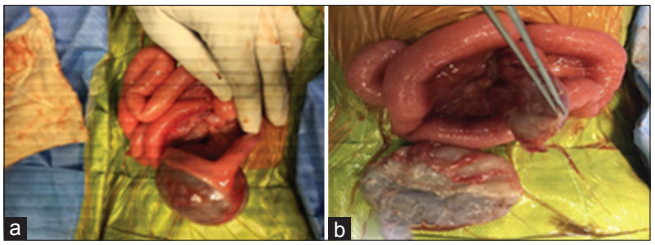

Figure 1: (a) Jejunal duplication cyst over the mesentric border. (b) Resected specimen of the cyst after subserosal dissection

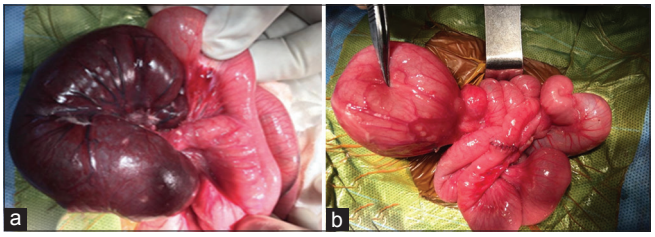

Figure 2: (a) Gangrenous ileal duplication cyst proximal to the IC junction volvulus. (b) Jejunal duplication cyst

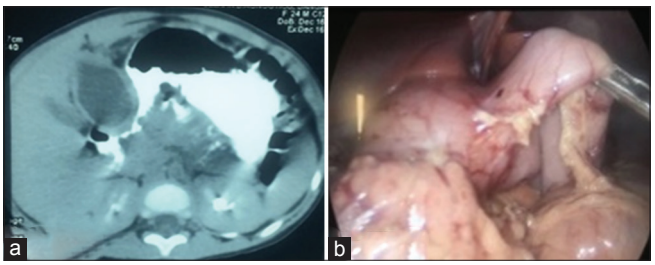

Figure 3: (a) Computed tomography abdomen showing a cystic lesion along the greater curvature and (b) intact gastric wall after subserosal dissection

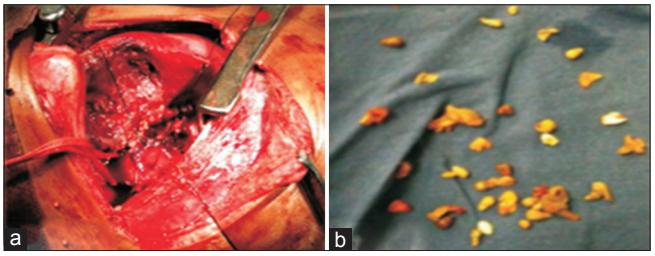

Figure 4: (a) Right lung abscess eroding the whole of lower lobe and (b) black channa seeds with other vegetative material found in the abscess cavity

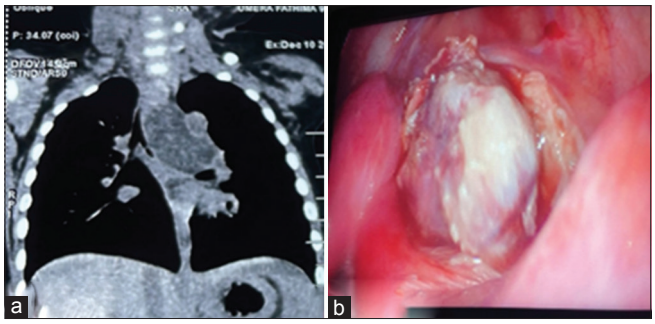

Figure 5: (a) Esophageal duplication cyst in the upper mediastinum compressing trachea and the bronchus and (b) esophageal duplication cyst after dissection from adjacent vital structures

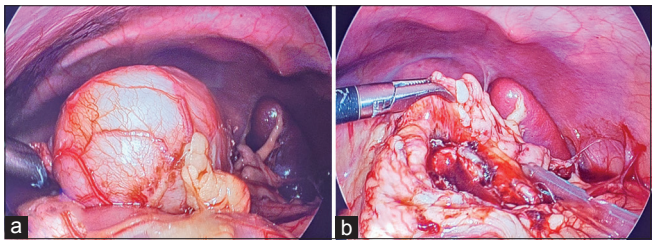

Figure 6: (a) Duplication cyst seen over the posterior wall of the stomach (b) status post subserosal dissection

showing a solitary well-defined fluid filled lesion, in the visceral compartment of the mediastinum with mass effect on trachea and compression of the left main bronchus [Figure 5a]. Flexible bronchoscopy showed complete external compression of the left main bronchus without any internal communication. Child was then planned for video-assisted thoracoscopic surgery (thoracoscopic excision of the cyst). Intraoperatively, duplication cyst arising from the esophagus was seen adjacent to the arch of aorta. Partial decompression of the cyst was done before proceeding with posterior dissection [Figure 5b]. Cyst was excised and the serosa of the cyst adhered to esophagus left behind. HPE was consistent with esophageal duplication cyst. The patient is doing well on follow-up.

Case 6

A 2-year-old boy presented to the outpatient department with intermittent episodes of colicky abdominal pain for past 3 months. No history of hematemesis. The patient is a known case of tetralogy of Fallot with pulmonary atresia with large subaortic ventricular septal defect with confluent branch pulmonary artery with patent ductus arteriosus post intracardiac repair. USG abdomen shows cystic lesion with turbid contents anteromedial to the kidney. CT kidneys, ureters, and bladder confirm 2.8 × 2.4 × 4.6 cm cystic lesion in the tail region of pancreas with findings suggestive of possibility of pseudocyst. Serum amylase and lipase values were normal. The patient was then planned for laparoscopic removal of the cyst, which was seen around the tail of pancreas intraoperatively. Complete excision of the cyst was done after dissecting it from the tail of pancreas. HPE analysis confirmed features of gastric duplication cyst [Figure 6a and 6b]. Post-operative period was uneventful and child is doing well on follow-up.

DISCUSSION

The incidence of EDCs is 1 in 4500 births and is found in 0.2% of all children, with a slight male predominance.[7,8] In our study, we had two cases of esophageal duplication cysts, two gastric and two small bowel duplication cyst.

EDCs are believed to occur between the 4th and 8th weeks of embryonic development. Their etiology is still unknown, though several theories have been proposed to explain their pathophysiology, no single hypothesis can justify all duplications location and associated anomalies. Split notochord theory is often postulated.[9] The luminal recanalization theory explains duplications in those portions of the GIT that have a solid stage including the esophagus, small bowel, and colon; nevertheless, it does not explain duplications at other levels. Incomplete or partial twinning theory could explain the colorectal duplications that are associated with duplication of genital and urinary structures. Persistent embryonic diverticula theory suggests that small diverticula, usually transient along the antimesenteric border of the intestinal wall, persist and develop intestinal duplications, although most EDCs are in the mesenteric border. These different theories lead to a thinking that the origin of EDCs can be multifactorial.[3,6,10-12] Incidence of multiple duplication cysts has also been observed but are rather uncommon (1–7%).[6,14] In our study, we have one case of 3-month-old infant with multiple duplication cyst involving the ileum and the jejunum. The variable location of the EDCs in our study corroborates with the fact that they can be located anywhere along the GIT and should be considered as a differential in a young child presenting with non-specific symptoms and it is imperative to investigate and inspect entire bowel during surgery to rule out association of multiple cyst.

The clinical presentation or onset symptoms of these malformations range from infancy and early childhood to adulthood. Almost 70% of EDCs present symptoms within the 1st year of life and 85% in the second one.[5,12,14] The signs and symptoms depend on the type and location of the duplication. Oral and esophageal cysts may cause respiratory distress or dysphagia. Retrosternal pain, hemoptysis, and infection can occur in case of large cysts with rapid growth.[15,16] Gastric and intestinal duplications may produce nausea, vomiting, abdominal distention, or

palpable abdominal mass. Recurrent abdominal pain is one of the most frequent forms of presentation and is usually attributed to high pressure inside the duplication cyst due to the accumulation of secretions. Extrinsic compression of the adjacent bowel is also possible, which causes obstruction.[10,11,13,15,17] In our study, we had two cases of esophageal duplication cyst. A 8-month-old infant had presented with recurrent lower respiratory tract infections due to cyst occluding the left bronchus lumen and the other case of 3 years old male who had presented with the right lower lobe lung abscess, as the duplication cyst had ruptured and eroded into the right lower lobe of lung. We had one case of multiple duplication cysts in the ileum and jejunal duplication cyst, where the ileal duplication cyst had caused the volvulus leading to acute small bowel obstruction. There were two cases of gastric duplication cyst and one of jejunal duplication cyst which had all presented with history of intermittent abdominal pain.

The most serious complications of duplication cyst can arise when an ectopic functional gastric mucosa is present within the cyst, leading to inflammation, bleeding, ulceration, and even perforation or ectopic functional pancreatic mucosa causing unexplained hypoglycemic episodes. We had one case of gastric duplication cyst with ectopic nonfunctional pancreatic tissue.

A prenatal diagnosis from an USG examination is possible for these lesions. However, the sensitivity is only 20–30%. USG of the abdomen shows a classical five-layered cyst wall with alternating hyperechoic and hypoechoic layers. This is known as the gut signature sign and is pathognomonic for alimentary tract duplication. Although USG can also be used to diagnose foregut duplications located in the neck, its sensitivity is poor for intrathoracic lesions. Transesophageal echocardiogram USG can be used for these lesions, but it is not readily available at all centers, especially in developing countries. CECT can be used to localize the lesions in these scenarios. Magnetic resonance imaging provides better cross-sectional anatomy in these cases and also helps in the diagnosis of duplication cysts involving the hindgut.[7]

In our study, CECT abdomen pelvis and high-resolution CT chest was used as a diagnostic modality to confirm diagnosis and to aid in the planning of the surgical resection of the cyst, though the HPE report has proved to be the gold standard for the diagnosis of EDC.[5] In our study, the HPE report of a 2 years old girl who had presented as gastric duplication cyst showed ectopic pancreatic tissue which supports the use of histopathological report for definitive diagnosis.

The definitive treatment for a duplication cyst is surgical resection of the cyst. The surgical approach can be open (laparotomy or thoracotomy) or minimally-invasive (laparoscopic or thoracoscopic) and depends on the expertise of the surgeon. In our study, among four cases of abdominal duplication cyst, two underwent laparotomy and laparoscopic excision of gastric duplication cyst was done in other two cases. We had two cases of esophageal duplication cyst, in which one was managed with thoracoscopic excision and other with thoracotomy with lower lobe lung resection. The confirmatory diagnosis of all the cases was done on the basis of HPE report.

CONCLUSION

EDCs are one of the rare congenital abnormalities. Most of the EDCs are asymptomatic and are detected incidentally in the first 2 years of life. EDCs can be multiple and occasionally present as acute emergencies due to volvulus, infection, impending perforation due to obstruction or have serious clinical manifestations due to functional ectopic gastric and pancreatic mucosa. EDC can be multiple. Cross sectional imaging like CECT abdomen pelvis or thorax for further evaluation, though not mandatory but is a valuable investigation which aids in diagnosis and in better surgical planning. EDCs have an excellent prognosis if detected early and excised intoto. Excision was traditionally done through open procedures but is now being successfully performed through minimally invasive surgeries, with meticulous subserosal dissection and thus avoiding the need for resection and anastomosis. Surgical excision is the only curative treatment for EDCs and intervention is advised early to avoid complications such as infection and malignant transformation.

REFERENCES

1. Liaqat N, Latif T, Khan FA, Iqbal A, Nayyar SI, Dar SH. Enteric duplication in children: A case series. Afr J Paediatr Surg 2014;11:211-4.
2. Blank G, Königsrainer A, Sipos B, Ladurner R. Adenocarcinoma arising in a cystic duplication of thesmall bowel: Case report and review of literature. World J Surg Oncol 2012;10:55.
3. Hur J, Yoon CS, Kim MJ, Kim OH. Imaging features of gastrointestinal tract duplications in infants and children: From esophagus to rectum. Pediatr Radiol 2007;37:691-9.
4. Berrocal T, Hidalgo P, Gutiérrez J, De Pablo L, Rodríguez-Lemos R. Imagen radiológica de las duplicaciones del tubo digestivo. Radiología 2004;46:282-92.
5. Sharma S, Yadav AK, Mandal AK, Zaheer S, Yadav D, Samie A. Enteric duplication cysts in children: A clinicopathological dilemma. J Clin Diagn Res 2015;9:8-11.
6. Macpherson RI. Gastrointestinal tract duplications: Clinical, pathologic, etiologic, and radiologic considerations. Radiographics 1993;13:1063-80.
7. Nebot CS, Salvador RL, Palacios EC, Aliaga SP, Pradas VI. Enteric duplication cysts in children: Varied presentations, varied imaging findings. Insights Imaging 2018;9:1097-110.
8. Tiwari C, Shah H, Waghmare M, Makhija D, Khedkar K. Cysts of gastrointestinal origin in children: Varied presentation. Pediatr Gastroenterol Hepatol Nutr 2017;20:94-9.

9. Qi BQ, Beasley SW, Williams AK. Evidence of a common pathogenesis for foregut duplications and esophageal atresia with tracheo-esophageal fistula. Anat Rec 2001;264:93-100.
10. Di Serafino M, Mercogliano C, Vallone G. Ultrasound evaluation of the enteric duplication cyst: The gut signature. J Ultrasound 2016;19: 131-3.
11. Letelier AM, Barría CM, Beltrán MS, Marcelo A, Moreno CH. Intestinal duplication: Diagnosis and treatment of an unusual condition. Revista Chilena De Cirugía 2009;61:171-5.
12. Palacios A, DeVera M, Martínez-Escoriza JC. Prenatal sonographic findings of duodenal duplication: Case report. J Clin Ultrasound 2013;41(Suppl 1):1-4.
13. Tong SC, Pitman M, Anupindi SA. Ileocecal enteric duplication cyst: Radiologic-pathologic correlation. Radiographics 2002;22:1217-22.
14. Górecki W, Bogusz B, Zajac A, Soltysiak P. Laparoscopic and laparoscopy assisted resection of enteric duplication cysts in children. J Laparoendosc Adv Surg Tech A 2015;25:838-40.
15. Okur MH, Arslan MS, Arslan S, Aydogdu B, Türkçü G, Goya C, *et al*. Gastrointestinal tract duplications in children. Eur Rev Med Pharmacol Sci 2014;18:1507-12.
16. Cuch B, Nachulewicz P, Wieczorek AP, Wozniak M, Pac-Kozuchowska E. Esophageal duplication cyst treated thoracoscopically during the neonatal period. Medicine (Baltimore) 2015;94:e227089.
17. Erginel B, Soysal FG, Ozbey H, Keskin E, Celik A, Karadag A, *et al*. Enteric duplication cysts in children: A single-institution series with forty patients in twenty-six years. World J Surg 2017;41:620-4.

How to cite this article: Sikerwar E, Chandrashekar V, Zameer M, Rajan CS. Enteric Duplication Cysts: Variations in Presentation and Management. Int J Sci Stud 2025;12(10):1-5.

Source of Support: Nil, **Conflicts of Interest:** None declared.