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Dr. Subhasree Beura
Senior Resident, Department of Pediatrics, AMRI Hospital, Bhubaneswar, Odisha, India

Dr. Baikuntha Narayan Mishra
Consultant, Department of Pediatric Surgery, AMRI Hospital, Bhubaneswar, Odisha, India

Dr. Ranjit Kumar Joshi
Consultant, Department of Pediatrics, AMRI Hospital, Bhubaneswar, Odisha, India

Dr. Prabin Prakash Pahi
Consultant, Department of pediatrics, AMRI Hospital, Bhubaneswar, Odisha, India

Corresponding Author:
Dr. Subhasree Beura
Senior Resident, Department of Pediatrics, AMRI Hospital, Bhubaneswar, Odisha, India

Enteric duplication cyst in a neonate: A case report

Dr. Subhasree Beura, Dr. Baikuntha Narayan Mishra, Dr. Ranjit Kumar Joshi and Dr. Prabin Prakash Pahi

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Abstract
Duplication cysts of intestine are rare congenital malformations formed due to some developmental anomaly of gastrointestinal tract during embryonic period. They can occur in any portion of the gastrointestinal tract but are more commonly seen in small intestine. They have a variable clinical presentation depending on the size, site and type, or they may remain asymptomatic. In most of the cases they are detected during infancy or early childhood and uncommon in adults. Antenatal ultrasound can detect the condition in fetus and helps in follow- ups subsequently. Treatment of asymptomatic case is controversial, however early intervention prevents complications. Herein, we share our experience of managing a neonate with enteric duplication cyst, detected antenatally.

Keywords: Enteric Duplication Cyst, Congenital Anomaly, Newborn, Antenatal Screening

Introduction
The incidence of gastrointestinal (GI) duplication is about 1 in 4500 live births. They can occur along the entire length of the gastrointestinal system. Though enteric duplications can present at any age but 80% of them present within first two years and majority within first three months of life. Calderin in 1733 reported the first case of intestinal duplication cyst, followed by Fitz (1884) and Ladd in 1937. In about 75% duplications have been found in abdomen. They may be located in thorax in 20% cases, and in 5% thoraco-abdominal. Duplications in proximity of small intestine are the most common and in about 53% they occur in ileum [1]. They may be cystic or tubular [2]. They have heterogeneous clinical presentations ranging from an asymptomatic course to life threatening consequences of volvulus, GI bleeding, perforation, Intussusception [3]. Antenatal ultrasound (USG) may demonstrate an intra-abdominal mass during 2nd or 3rd trimester of pregnancy in 20-30% of cases [4]. It may be confused with meckel's diverticulum or intussusceptions [5]. Postnatal USG and contrast enhanced computed tomography (CECT) abdomen/ magnetic resonance imaging (MRI) may differentiate it from other intra-abdominal cystic lesions. Surgical management is recommended in all these cases as they may result in significant morbidity and mortality if left untreated.

Case report
A 19day male child was referred to our hospital for evaluation of an intra-abdominal mass, detected antenatally. An antenatal USG done at 33wk gestational age had revealed a well-defined, oval, cystic, anechoic mass lesion of size 5.3×2.3 cm in right lumbar region. The baby was first order, born at term gestation, was appropriate for gestational age with uneventful antenatal and perinatal history and was doing well postnatally. There was no presenting complaints on admission, an initial physical examination showed a soft abdomen with no palpable mass, rest of the systemic examinations were within normal limits. A repeat USG revealed well-defined cystic lesion of size 49×17×15 mm in right lumbar region displacing bowel loops peripherally with thick wall with a double line appearance and fine internal echoes. CECT abdomen revealed well-defined minimal enhancing thick walled cystic lesion of size 49×26×50 mm in right lumbar region with displacement of small bowel loops peripherally and ascending colon posteriorly, no significant communication seen to adjacent bowel loops and no evidence of traversing vessels seen (fig 1). The patient was taken for surgery. On exploratory laparotomy, a non-intercommunicating terminal ileal duplication cyst was found (fig 2). Resection of the involved segment followed by ileo ascending colon end to end anastomosis was

performed. The cyst was unilocular and containing clear yellow fluid and mucus. Histopathological examination revealed the resected cyst and contiguous portion of ileum shared a common muscular wall, although each had its own mucosal lining, confirming it to be a duplication cyst. The patient's post-operative course was uneventful and was discharged on 10th post-operative day. He is doing well on follow-ups after three months.

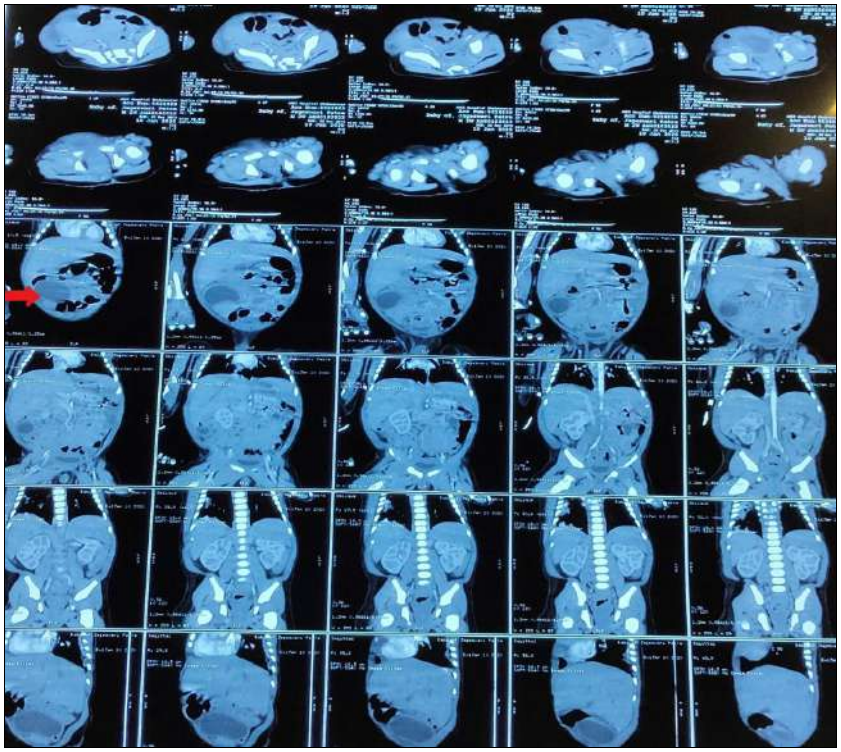

Fig 1: CECT showing enteric duplication cyst

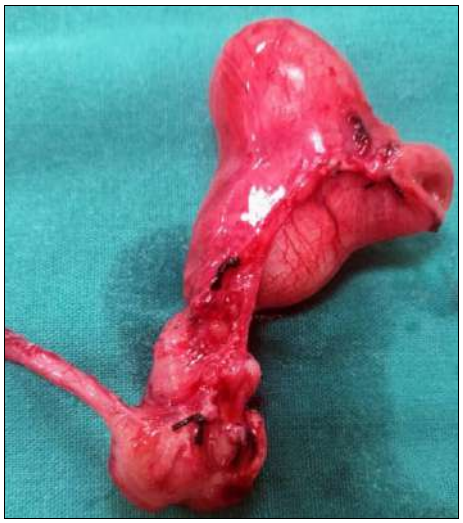

Fig 2: Gross specimen of excised ileal duplication cyst

Discussion

Duplication cysts are defined as spherical, or tubular structures that are firmly attached to at least one point in the alimentary tract, possess a well-developed coat of smooth muscle and have an epithelial lining resembling some part of alimentary tract [2]. Commonly duplication cysts are communicated with the intestinal lumen, involve the mesenteric border of the associated alimentary tract and share a blood supply with the native bowel. Many theories have been proposed about the origin of this disease; however, it continues to be unknown because of the inconsistencies in them. The "Diverticular theory" suggests that small transitory diverticula that are on the antimesenteric side of the intestinal wall of the embryo persists and turns into an enteric duplication cyst. Other theory is the lack of recanalization of the intestine after its solid stage during embryonic development. The split notochord theory states that during third to fourth week of gestation, gaps may appear in the notochord through which endodermal cells can herniate and form diverticula. Environmental factors theory postulates that stress hypoxia and trauma may induce duplications [1].

The location of enteric duplication cyst could be intramural or less frequently extramural. It could have ectopic gastric or pancreatic tissue with or without a common wall with adjacent organs. They are mainly located in the ileum (40%), but they have been encountered in the esophagus, colon, jejunum, stomach, duodenum and rectum. The presentation of this cyst is usually cystic (90%) but it may be also tubular (10%), single or multiple in up to 15% of the cases. Other malformations may be associated with it as spinal defects, intestinal malrotation, intestinal atresia, and other abnormalities of the urinary tract [6]. No such anomalies were found in our patient.

The clinical presentation of enteric duplication cyst varies greatly from asymptomatic to acute abdomen. However, in recent days, with the increasing use of prenatal USG scan, many cases are being identified in utero like in our case. The mode of presentation usually depends on the anatomic level of the duplication, size, mass effect of lesion, presence of heterotopic gastric mucosa within the duplication and communication with the adjacent bowel and inflammation. Rai *et al.* have also reported duplication cysts with pancreatic mucosa. Often they mimic other intra-abdominal conditions posing great diagnostic difficulty. The presenting symptoms are non- specific and may vary depending on its size, location and type. Abdominal pain, nausea, vomiting, bleeding, abdominal distension, mass, intestinal obstruction has been reported by Sonam *et al.* It may

also remain asymptomatic till adult hood and may be detected incidentally during surgery [1]. The differentiation with other abdominal cysts (ovary, mesentery or common bile duct) should be carried out in addition to other etiologies that could produce dilatation of bowel wall or urological problems. It is also difficult to differentiate enteric duplication cyst from ovarian cyst in particularly female patient [4]. Duplication can sometimes lead to complications which include perforation, intussusception, volvulus, and associated malignancy [7]. But malignancy arising from duplication cysts particularly in children is quite rare. Our case was asymptomatic and was detected antenatally as intra-abdominal mass and the same was confirmed by postnatal USG and CECT abdomen. We also did not encounter any complications in our case.

With the evolution in pre and postnatal diagnostic studies an early and more frequent diagnosis is possible. The imaging modalities commonly used to investigate duplication cysts is X-ray abdomen, USG, barium studies, CT scan and magnetic resonance imaging [8]. Plain X-rays are not specific as they only may reveal dilation of bowel loops or may show multiple air fluid levels. Technetium scintigraphy can also be helpful specially if there is ectopic gastric mucosa, but it does not distinguish it from Meckel's diverticulum. USG guides the diagnosis if a "double wall "image is identified, especially in the prenatal period and demonstrates location and nature of the mass. CT scan and MRI may demonstrate the mass more precisely. But definitive diagnosis is made following surgery and histo-pathological study of the surgical specimen [1, 4]. Enteric duplication cysts with ectopic gastric mucosa may present with bleeding and perforation, requiring emergency surgical intervention. Usually, the treatment of choice for cystic masses in children is complete surgical resection, through laparotomy, laparoscopy or laparo-assisted surgery [9, 10]. Total resection when possible should be the aim of the intervention because the partial excision contains high risk of recurrence [11]. Surgical management of asymptomatic cases still remains controversial, however due to the risk of potential complications, enteric duplication cysts should always be excised [11, 12, 13]. Segmental resection along with adjacent intestine is recommended in small cystic or tubular duplication. But in long tubular duplication mucosal stripping has been advocated to avoid short bowel syndrome [14]. In our case, on laparotomy we found a terminal ileal duplication cyst and performed a segmental resection including the ileo cecal junction, as it was in very close proximity to cecum, followed by end to end anastomosis. Duplication cyst was confirmed with histopathology. The contents were yellow oily fluid and mucus. There was no ectopic mucosa or evidence of malignancy. The post-operative course was uneventful and the baby was discharged on 10th post-operative day. He was on regular follow-ups and doing well after three months.

Conclusion

Enteric duplication cysts are rare congenital lesions, which may present with nonspecific symptoms or may remain asymptomatic. A pre-operative definite diagnosis of duplication cyst is not possible and may present with life threatening complications like intestinal obstruction, bleeding and perforation requiring urgent surgery. In children with an antenatal diagnosis of abdominal mass, enteric duplication cyst should be kept in mind. These babies should be followed-up with postnatal USG and evaluated further if the mass persists. Surgical management and histopathology should be considered to reach a definite diagnosis even in asymptomatic cases to avoid future complications.

Conflict of Interest

All authors declare that they have no conflict of interest.

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