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Case Report

A Rare Congenital Anomaly: Jejunal Duplication Cyst

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I N F O

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A B S T R A C T

Gastrointestinal duplication cysts are a group of rare congenital anomalies with incidence of 1 in 4500 births. They are encountered mainly during first 2 years of life or antenatally. Here we report a case of non-communicating duplication cyst of jejunum in a 29 days old infant who presented with complaints of bilious vomiting. Most of the cases are associated with some other congenital anomaly. Completely isolated jejunal duplication cyst is rare. This case is presented to create an awareness of this rare entity which can occur as different presentations causing significant morbidity and even mortality if timely management is not provided.

Keywords: Congenital Anomaly, Duplication Cyst, Jejunal Cyst, Gastrointestinal

Introduction

Intestinal duplication cysts are uncommon and rare developmental malformation of digestive system which is mainly detected during infancy or early years of life but seldom in adults.[1] With incidence of 1 in 4500 births, they may be associated with other congenital anomalies of spine or genitourinary tract.[2] Clinically they can be asymptomatic or present with symptoms like vomiting, abdominal distension, pain or obstruction.

Awareness and diagnosing these duplication cysts are essential as, though a simple lesion, they can have lethal consequences if not treated on time.[3] Antenatal scans and postnatal radiological modalities help in the early diagnosis of duplication cyst but final diagnosis is proved on histopathological examination.

We are presenting here a rare case of non-communicating jejunal duplication cyst in 29 days infant who presented with complaints of bilious vomiting.

Case Study

A 29 days old female infant presented with complaints of bilious vomiting, projectile in nature since 5 days. Infant took normal feeds since birth.

Antenatal ultrasound scan at 20 weeks was suggestive of cystic lesion? Mesenteric cyst of size 2.1 x 1.6 cm. Second antenatal ultrasound scan at 27 weeks was suggestive of mesocolic cyst of size 2.8 x 2 cm.

Postnatal scans were suggestive of 5.9 x 4.6 cm cystic lesion in right lumbar region extending into Right hypochondriac region closely abutting and splaying bowel loops around it and mild dilation of proximal bowel.

CT scan reports was suggestive of a large well defined, thin walled cystic lesion measuring approx. 6.6 x 5.1 x 5 cm in umbilical and left lumbar region likely to be a benign cystic lesion most likely a mesenteric cyst (Figure 1).

Clinically infant was stable. The abdominal examination

revealed soft abdomen with minimal tenderness in Right hypochondriac region and a palpable cystic swelling. There was no associated spinal or genitourinary tract anomaly.

Infant underwent exploratory laparotomy through Right supraumbilical transverse incision. The laparotomy revealed jejunal duplication cyst measuring 8 x 8 x 4 cm in size approximately 10 cm distal to duodenojejunal junction in the mesenteric border along with malrotation of the bowel with duodenojejunal junction to the right of midline. Resection of ~10 cm of jejunum with the duplication cyst was done followed by anastomosis of jejunum. Ladd's procedure was done to rectify the malrotation of the bowel.

The specimen was sent for histopathological examination. We received a specimen of jejunum measuring 9 cm in length with a cyst measuring 7 x 6.5 x 5 cm (Figure 2). The serosal surface of the jejunal loop was congested. The cyst wall was grayish white in color with congested and dilated blood vessels. On opening the jejunal cyst around 20 cc sero-mucinous fluid oozed out. On opening the cyst was unilocular, non-communicating with smooth inner wall. The jejunal mucosa was unremarkable. Multiple sections were taken from the cyst wall, jejunum, surgical margins and the cyst with jejunal wall.

Sections from resected margins showed unremarkable jejunal lining. Sections from adjoining cyst also showed similar type of jejunal mucosal columnar lining (Figure 3). There was a common muscle layer between the jejunal wall and the cyst wall confirming the diagnosis of jejunal duplication cyst along with clinical and radiological correlation (Figure 4).

Discussion

The term intestinal duplication was first coined by Fitz RH[4] but was popularized in 1930s by Ladd WE[5] and further classified by Gross RE in the 1950s.[6]

Duplication of gastrointestinal tract, a rare congenital anomaly, is found in about 0.2% of all the children.[7,8] Age of presentation with enteric duplication cyst is mainly within first 2 years of life but can remain undetected until adulthood.[9] Exact etiology is unknown but several theories have been suggested like split notochord theory, aberrant intestinal lumen recanalization or persistent embryological diverticula. In the split notochord theory, the neural tube traction mechanism results in intestinal duplication along with anomalies of spinal cord and vertebra.[10-12]

Duplication cyst can occur at different sites most common being the abdominal cavity (75%) followed by intrathoracic (20%) and remaining by thoracoabdominal (5%). Amongst abdominal duplication cysts jejunal lesions are the most common (50%), followed by ileum (44%), colonic (13%), gastric (7%), duodenal (6%), rectal (4%) and oesophageal (2%). Other extra abdominal sites include mediastinal (18%) and cervical (1%) lesions.[1] Duodenal, jejunal and ileal are the three small bowel sites for duplication cysts. Our case study presents jejunal duplication cyst. Duplication cysts are called 'duplication' because of the attachment to normal intestinal tract and their common blood supply.

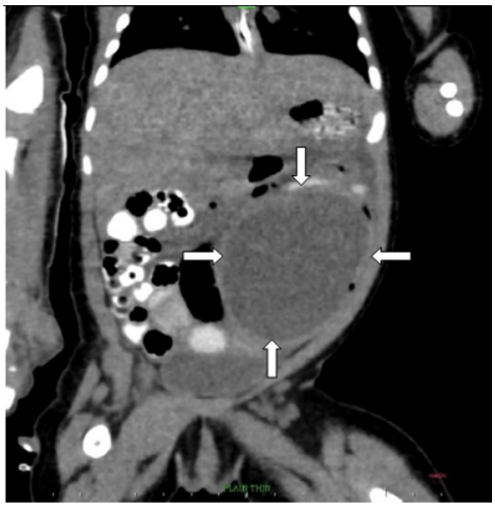

Figure 1.CT scan image of the duplication cyst in coronal section marked with arrows

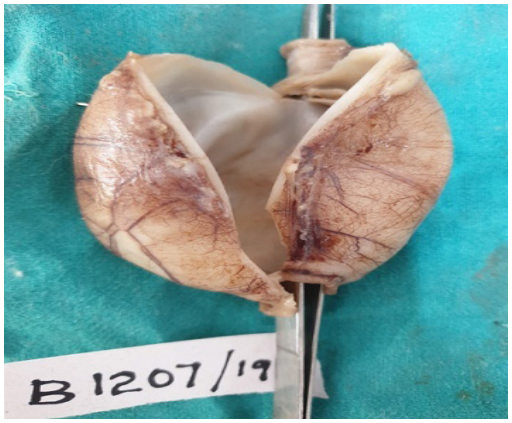

Figure 2.Gross specimen: A specimen of jejunum with non-communicating duplication cyst

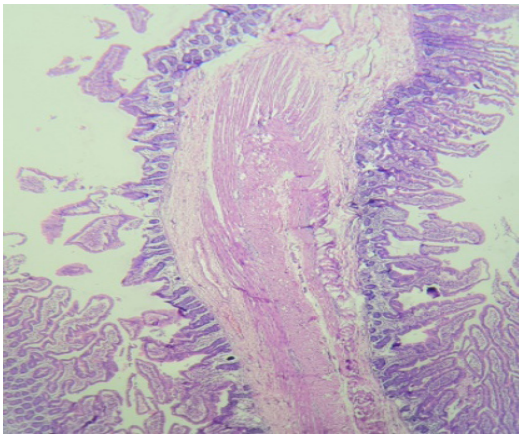

Figure 3.Photomicrograph shows adjoining cyst having similar type of jejunal mucosal lining (H and E x100)

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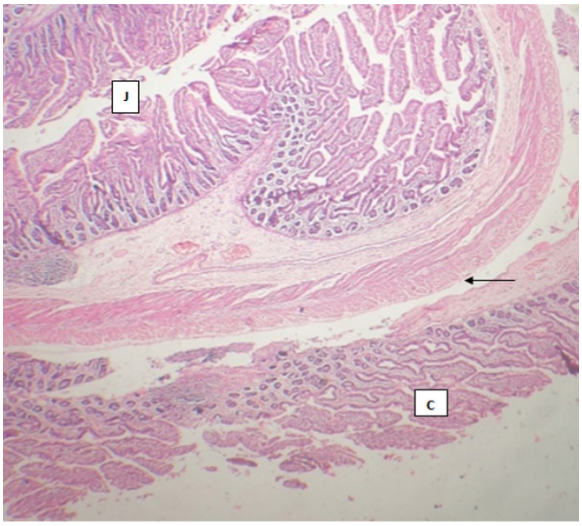

Figure 4.Jejunal Duplication Cyst. Photomicrograph shows a common muscle layer (arrow) between the jejunal wall (J) and the cyst wall (C) (H and E x400)

Duplications are located on dorsal aspect of intestine usually mesenteric but sometimes intramural.[13] The size of cyst varies from few millimeters to approximately 10 to 15 cm or more in diameter. They can be classified into localized duplication cyst, duplication in association with spinal cord or vertebral malformation and duplication of colon.[10] Rai BK et al. reported enteric duplications with ectopic mucosa like pancreatic tissue.[13] Duplication cysts may also be cystic (80%) or tubular (20%).[14] Tubular duplications may be attached parallel to lateral border of intestine which is then known as double barrel duplication. Rarely if the duplication has separate mesentery and blood supply it is then known as loop duplication. Often the duplication is in communication with lumen of intestine and can be at proximal or distal end or at both ends of duplication.[15] Duplication cysts can also be classified as typical or classical and atypical or unusual cysts.[16] Typical cysts are named so because they have intimate contact with any part of the gastrointestinal tract from mouth to anus, have smooth muscles in its wall and the lining mucosa is like that of any part of GIT. Atypical, on the other hand differ in the feature that they don't possess intimate contact with the GIT. Our study case presents a localized, non-communicating, typical duplication cyst present on the mesenteric border.

Jejunal duplication cysts can cause variety of symptoms depending upon its location, size and other factors such as presence of ectopic mucosa, inflammation or its communication with adjacent bowel. Symptoms include abdominal pain, abdominal distension, vomiting, constipation, bleeding, volvulus, intussusceptions and small bowel obstruction.[10,13] In our study the patient presented with complaints of only bilious vomiting.

Imaging modalities like barium studies, USG or CT scans are usually done to diagnose such cases. Imaging modality of choice is USG for evaluating any abdominal mass in neonate. It demonstrates the nature and location of the abdominal mass. Endoscopic ultrasound helps to distinguish between the solid and cystic lesions. It also establishes the location of cyst relative to the surrounding tissues. Duplication cyst is seen as anechoic, homogenous lesions having regular margins that arises from submucosal layer or is extrinsic to the gut wall.[14] Antenatal scans can allow early detection, treatment and avoidance of possible complications.[17] On CT scan duplication cysts are seen as smooth, round, fluid filled cysts or a tubular structure with slightly enhancing thin cyst wall.[10]

The confirmation of the duplication cyst is done through histopathological examination of the resected specimen.[1] On microscopy, intestinal duplication cysts contain two mucosal layers that share a common muscle layer. Jejunal duplication cysts show mucosa, submucosa, muscularis propria, and lining that contains jejunal mucous glands.[14]

Intestinal duplications, when symptomatic, require urgent surgical intervention. Usually segmental resection of the cyst along with the adjacent intestine is the treatment method for small cystic or short tubular duplication. For a long tubular duplication, the management is by mucosal stripping through series of multiple incisions. Management of asymptomatic cases remains controversial, though it is recommended to remove the duplication to avoid complications that may occur later in life.[14] In this study the infant underwent exploratory laparotomy followed by resection of the jejunum duplication cyst and resection and anastomosis of the adjacent jejunum segment.

The differential diagnosis can be all the causes of neonatal bowel obstruction like intussusceptions, volvulus, mesocolic cyst, mesenteric or omental cyst.[18] In a female patient ovarian cyst should also be taken into consideration.[17]

Conclusion

Intestinal duplication cysts are rare congenital entity that can have a variety of clinical presentation and complications with significant risk of morbidity and mortality. This explains the importance of early diagnoses and treatment of this congenital anomaly. Surgical resection of the duplication cyst along with the adjacent segment is the treatment of choice with excellent prognosis.

Conflict of Interest: None

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