Case Report

Isolated Duodenal Duplication Cyst in a Neonate

Riyazhussein Yakoob Hakda, Deepen V. Makwana, Ramendra Shukla[1], Urvish Parikh[1], Sudhir B. Chandna[1]

Department of General Surgery, [1]Department of Paediatric Surgery, SVP Institute of Medical Sciences and Research, NHL Municipal Medical College, Ahmedabad, Gujarat, India

Abstract

Duodenal duplication cysts are a rare subtype of alimentary tract duplications cysts, consisting of 7% of all the duplications. We report a rare case of neonatal duodenal duplication cyst presenting as a palpable abdominal mass and features of gastric outlet obstruction. A 27-day-old male child presented with complaints of icterus, non-bilious vomiting after every feed and right-sided abdominal lump for the last 15 days. A computed tomography scan of the abdomen revealed well-defined peripherally enhancing cystic lesion noted in the subhepatic region extending up to the right lumbar region. On surgical exploration, a cystic mass was found attached to the pyloric part of the stomach along the mesenteric border of the first, second and third part of the duodenum, which was marsupialised, and no communication was found with the duodenum. On histopathological analysis, a duodenal duplication cyst was diagnosed without any heterotopic mucosa. The literature was reviewed and the approach to duodenal duplication cyst in neonates is discussed.

Keywords: Duodenum, duodenal duplication cyst, neonate, neonatal abdominal lump, paediatric surgery

Introduction

Gastrointestinal duplications are rare congenital anomalies, of which duodenal duplication cysts are even rarer subtypes, comprising of 7% of all the duplications.[1] A duplication cyst is characterised by its location in or immediately adjacent to the wall of the GI tract, lying on the mesenteric side, sharing a common blood supply, containing a muscular wall with lining mucosa of any type, including ectopic gastric, pancreatic or respiratory tissue.[2,3] Several theories have been put forward to explain GI duplications, such as the abortive twinning theory, persistent embryologic diverticula theory, and the aberrant luminal recanalisation theory.[4,5] The enteric duplications are most commonly present in infancy or early childhood, presenting as early as infancy till up to 52 years of age. These could be asymptomatic like that in infants,[6] or more commonly present as vague abdominal pain, nausea and vomiting or a palpable abdominal mass.

We report a rare case of neonatal duodenal duplication cyst presenting as a palpable abdominal mass and features of gastric outlet obstruction.

Case Presentation

The patient is newborn male child of 3 kg birth weight delivered by full-term normal vaginal delivery at a hospital in Nepal after an uneventful pregnancy, with a good Apgar score, 3rd child with a gap of 4 years to previous birth. The child passed meconium within 24 h of birth. There is no family history of any congenital malformation in siblings or parents.

On day 11 of life, the mother noticed yellowish discolouration of the sclera of both eyes, for which she consulted a paediatrician. On DOL 13, the patient had 7–8 episodes of non-bilious vomiting containing breastmilk, along with excessive crying. Icterus was managed conservatively and the patient discharged on DOL 15. On DOL 18, the mother noticed a swelling over the right side of the abdomen, with associated vomiting and crying spells. The swelling was insidious in onset and was increasing gradually. There was no history of fever, weight loss, altered bladder or bowel habits.

Address for correspondence: Dr. Sudhir B. Chandna, Department of Paediatric Surgery, SVP Institute of Medical Sciences and Research, NHL Municipal Medical College, Ahmedabad - 380 006, Gujarat, India.
E-mail: sudhirchandna@gmail.com

Received: 12-12-2021 **Revised:** 21-01-2022 **Accepted:** 22-01-2022 **Available Online:** 08-06-2022

Access this article online

Quick Response Code:

Website: www.afrjpaedsurg.org

DOI: 10.4103/ajps.ajps_176_21

How to cite this article: Hakda RY, Makwana DV, Shukla R, Parikh U, Chandna SB. Isolated duodenal duplication cyst in a neonate. Afr J Paediatr Surg 2022;19:257-60.

On DOL 27, the patient was brought to our hospital for further evaluation and management. On physical examination, the neonate was a full term, appropriate for gestational age with normal growth, active with good cry, afebrile, no pallor, cyanosis or oedema with the presence of mild icterus, with a heart rate of 126/min, respiratory rate of 32/min and oxygen saturation of 99% on room air. There were no scars, sinuses or neuro-cutaneous markers present. No gross congenital anomalies were detected on examination. The abdomen was soft, with fullness present in the right lumbar region and approximately 6 cm × 6 cm sized single swelling present in the right lumbar region with well-defined margins, smooth surface, soft, cystic consistency, without any signs of inflammation and not attached to overlying skin. The rest of the systemic examination was normal.

Laboratory investigations showed a haemoglobin of 12.4 g/dL, total white blood cell 15,700/mm^3, platelet count of 619,000/mm^3, total bilirubin 7.21, direct bilirubin 0.8, indirect bilirubin 6.41.

An ultrasound of the abdomen was done which suggested a cystic lesion measuring 5.8 cm × 6.8 cm inferior to the liver and adjacent to pylorus whose origin could not be determined, with a possibility of a mesenteric cyst.

Multidetector computed tomography imaging of abdomen and pelvis [Figure 1] was performed with oral, per-rectal and IV contrast.

Computed tomography (CT) scan of the abdomen and pelvis demonstrated approximately 65 mm × 71 mm × 64 mm (AP × TR × CC) sized well-defined peripherally enhancing cystic lesion is noted in the subhepatic region extending up to the right lumbar region, the maximum wall thickness of cyst measures 2.5 mm. No evidence of any internal septa, calcification or soft-tissue component within the cyst. Anteriorly and right laterally it reaches up to the anterior abdominal wall. The lesion displaces a small bowel loop towards the left side. It displaces and compresses pylorus of the stomach, superiorly. Medially it compressed and displaces 1st and 2nd parts of the duodenum. Posteriorly it displaces and compresses hepatic flexure, ascending colon, right kidney and compresses inferior vena cava, however patent. Medially the lesion abuts the abdominal aorta, however patent. Minimal free fluid is noted in the subhepatic region.

Patient was kept NPO and nasogastric intubation done with IFT FG-8.

On DOL 30, the patient was taken for exploratory laparotomy under general anaesthesia. Upon reaching the peritoneal cavity, approx. 8 cm × 7 cm × 7 cm sized spherical cystic mass was identified [Figure 2a], which was attached to pylorus of stomach till third part of the duodenum, over the mesenteric border. The entire 1st and 2nd parts of the duodenum was not visualised separately from the mass. The cyst was dissected out from the mesenteric layer. Approximately 200 ml of seromucinous fluid was drained. The cyst was opened wide to check for any communication with pylorus or duodenum and luminal continuation was cross-checked by insufflation of 60 ml air via the IFT. Marsupialisation was done [Figure 2b], the cyst wall was excised up to 0.5 cm from the common wall and sent for histopathological examination. The remnant of the cyst mucosa was electrocauterised. The entire abdominal cavity was inspected, and no other abnormality detected.

Histopathological examination revealed mucosa lined by enterocytes along with Brunner's glands [Figure 3]. Outer smooth muscle layer was seen. There was no evidence of heterotopic mucosa or ectopic pancreatic mucosa and the findings were suggestive of duodenal duplication cyst.

Post-operative period was uneventful and the patient was discharged on post-op day 7.

In 1 week follow-up, the patient remained asymptomatic.

Discussion

Abdominal masses in infants have varied presentations, with 65% of them presenting in the flanks (55% of which are renal in origin), 20% intraperitoneal in nature and 15% being of pelvic origin.[7] The intraperitoneal masses could be of gastrointestinal (GI) (15%) or hepatobiliary (5%) in origin. GI masses are usually mesenteric cysts, enteric duplication cysts, omental cysts and meconium cysts or meconium ileus.[8] The clinical presentation is variable, depending on the size, location and mass effect. In the neonates, it often presents as an asymptomatic abdominal mass, with physical signs of abdominal distension or a palpable abdominal mass.[9]

One-third of mesenteric cysts present in children <15 years of age, with slight male preponderance. Clinical presentation is incidental as an asymptomatic abdominal mass, or acute abdomen due to the complications arising from the cyst such as infection, rupture of the cyst, haemorrhage, intestinal obstruction or volvulus.[8] Omental cysts present in children <10 years of age, with features of a palpable freely mobile mass and subacute intestinal obstruction. Meconium pseudocyst is an acute presentation of the newborn, in which foetal intestinal perforation along with leakage of meconium is contained in the form of a cyst, with features of peritonitis.[10]

Duodenal duplication cysts represent a minor fraction of all the GI duplications, the estimated prevalence of which is <1 in 100,000 live births.[11] The most most common of the duplications occur in jejunum and ileum.[11-14]

The earliest descriptions were given by Calder.[15] A duplication cyst is characterised by its location in or immediately adjacent to the wall of the GI tract, lying on the mesenteric side, sharing a common blood supply, containing a muscular wall with lining mucosa of any type, including ectopic gastric, pancreatic or respiratory tissue.[2,3] They are usually spherical, non-communicating cysts located along the first and second part of the duodenum. In our case, the mass was a spherical,

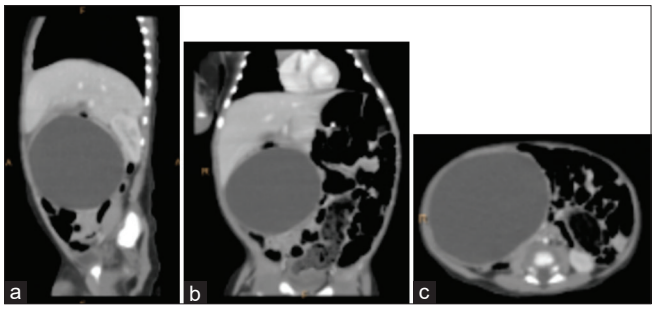

Figure 1: Multidetector computed tomography scan of whole abdomen showing sagittal (a) coronal (b) and transverse (c) sections, depicting fluid-filled cystic lesion pushing the bowel loops towards left side

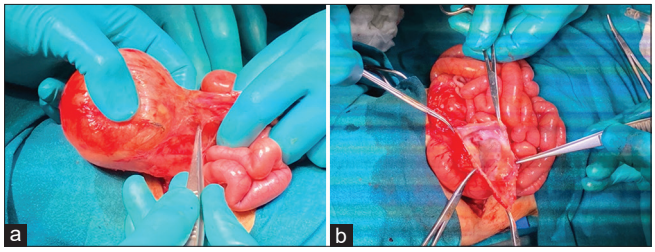

Figure 2: Intra-operative pictures of a non-communicating duodenal duplication cyst. (a) A cyst attached to the first and second part of duodenum. (b) Cyst wall opened showing no communication with the lumen of duodenum

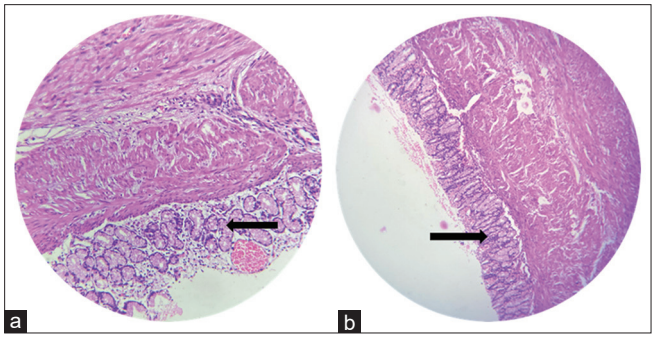

Figure 3: A 27-day old male child with a non-communicating duodenal duplication cyst. Photomicrographs of haematoxylin and eosin-stained tissue sections from the cyst wall showing the normal components of duodenal wall, with submucosal Brunner's glands (arrow in a and b). Magnifications: (a) ×40 and (b) ×10

non-communicating cyst lying over the mesenteric wall of the pylorus, first, second and third parts of the duodenum.

Several theories have been put forward to explain GI duplications, such as the abortive twinning theory, persistent embryologic diverticula theory, and the aberrant luminal recanalisation theory.[4,5]

The enteric duplications are most commonly present in infancy or early childhood, presenting as early as infancy till up to 52 years of age. These could be asymptomatic like that in infants,[6] or more commonly present as vague abdominal pain, nausea and vomiting or a palpable abdominal mass. Depending on the type of duplication present, the symptoms could also include GI bleed, intussusception, obstruction, jaundice or pancreatitis.[16-20] Rarely, the duplication cyst could undergo malignant degeneration in adults, however, no such cases were reported in children under the age of 16 years.[16] In the case we encountered, who was brought to the hospital at the age of 27 days, the presenting features included nausea and vomiting which was followed by the appearance of a palpable abdominal lump in the right lumbar region.

Various congenital anomalies could be present in association with the duplications, more commonly so in thoracic, midgut and hindgut duplications. Vertebral anomalies such as bifid or hemivertebrae or vertebral fusion may also be present; they are most often associated with thoracic and hindgut duplications. Other recorded associated congenital malformations include congenital cardiac disease, oesophageal atresia, congenital diaphragmatic hernia, congenital pulmonary malformations and myelomeningocele with foregut duplications; intestinal malrotation or (less commonly) intestinal atresia with midgut duplications; and genitourinary duplication, bladder exstrophy and imperforate anus with hindgut duplications.[16] In this case, however, no congenital anomaly was detected on clinical examination as well as imaging.

Microscopy usually reveals enteric mucosal lining, along with the presence of duodenal Brunner's gland in the submucosa, along with muscle layer and nerve plexus. Occasionally there is the presence of ectopic gastric or pancreatic epithelium, which predisposes to ulceration, bleeding and perforation.[16,21] In the specimen we sent for histopathological examination, the mucosa was that of the duodenum with evidence of Brunner's glands, smooth muscle layer, without any ectopic mucosa. There was also no evidence of malignant degeneration.

For diagnosis of such cysts, USG is usually done as first line investigation followed by CT scan to look for craniocaudal extension, related structures and delineate cyst anatomy. Other investigations may be required based on clinical presentation. If haemorrhage present, one can order technetium-99 m pertechnetate radionuclide scan to detect heterotopic gastric mucosa. in case of the duodenal cyst with suspicion of cystobiliary communication. Magnetic resonance cholangiography or, in older children, endoscopic retrograde cholangiopancreatography (ERCP) are useful.[16,22-24]

Complete excision of cyst is ideal and can be done for simple cysts but sometimes in complex conditions like cyst on the medial aspect of 2nd or 3rd part of duodenum with possible cystobiliary or luminal communication or it is sharing common muscular wall, it is wise to carry out marsupialisation or partial excision with mucosectomy.[16,19,25] Per-operatively, we found that the cyst was closely related to pylorus, 1st, 2nd and 3rd parts of the duodenum, therefore we did marsupialisation and excision of cyst up to 0.5 cm of common wall and cauterised the remaining mucosa to prevent future complications like recurrence, haemorrhage or infection.

In case of doubtful cystobiliary communication, one can aspirate the cyst and if aspiration yields bile, an on-table cholangiogram can be performed or cholecystectomy followed by passing of fine probe or catheter distally to look for any communication.[16]

Nowadays, minimally invasive treatment options are also growing which include endoscopic marsupialisation of duodenal duplication cyst.[26]

Conclusion

A rare entity of a duodenal duplication cyst manifesting as abdominal lump along with features of gastric outlet obstruction should be considered as a differential diagnosis in a newborn presenting with abdominal mass with vomiting. The diagnosis is confirmed radiologically by an experienced ultrasonologist or a CT scan of the abdomen, with advanced options such as MRCP or ERCP being the alternatives. The mainstay of the treatment is surgical excision of the cyst via open surgery, given the structures opening in the duodenum. Endoscopic approach is also performed by many surgeons, with a said difficulty of the procedure.

Duodenal duplication cysts are the rare varieties of GI tract duplications, presenting most commonly as a palpable abdominal mass and obstruction, occasionally associated with haemorrhage due to peptic perforation, jaundice due to biliary obstruction, or pancreatitis. Ultrasound, CT scan, Magnetic resonance imaging and ERCP are the diagnostic tools. Surgical modalities include simple cyst excision, marsupialisation, or Roux-en-Y cystjejunostomy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Mba, M. I. G. H. W., Md, P. M. J., & Peter, S. S. D., MD. (2019). Ashcraft's Pediatric Surgery: Expert Consult-Online+Print (7th ed.) [E-book]. Elsevier.
2. Macpherson RI. Gastrointestinal tract duplications: Clinical, pathologic, etiologic, and radiologic considerations. Radiographics 1993;13:1063-80.
3. Guarise A, Faccioli N, Ferrari M, Romano L, Parisi A, Falconi M. Duodenal duplication cyst causing severe pancreatitis: Imaging findings and pathological correlation. World J Gastroenterol 2006;12:1630-3.
4. Edwards H. Congenital diverticula of the intestine: With report of a case exhibiting heterotopia. Br J Surg 1929; 17:7-21.
5. Lewis FT, Thyng FW. The regular occurrence of intestinal diverticula in embryos of the pig, rabbit, and man. Am J Anat 1908; 7:505-519.
6. Ros PR, Olmsted WW, Moser Jr., RP *et al.* Mesenteric and omental cysts: Histologic classification with imaging correlation. Radiology. 1987;164:327-332. PMID: 3299483.
7. Standardized Toolbox of Education for Pediatric Surgery Abdominal Masses of Childhood. American Pediatric Surgical Association Committee of Education, 2012-13
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. Ranganath SH, Lee EY, Eisenberg RL. Focal cystic abdominal masses in pediatric patients. AJR Am J Roentgenol 2012;199:W1-16.
10. Cass DL. Fetal abdominal tumors and cysts. Transl Pediatr 2021;10:1530-41.
11. Chen JJ, Lee HC, Yeung CY, Chan WT, Jiang CB, Sheu JC. Meta-analysis: The clinical features of the duodenal duplication cyst. J Pediatr Surg 2010;45:1598-606.
12. Prasad TR, Tan CE. Duodenal duplication cyst communicating with an aberrant pancreatic duct. Pediatr Surg Int 2005;21:320-2.
13. Hata H, Hiraoka N, Ojima H, Shimada K, Kosuge T, Shimoda T. Carcinoid tumor arising in a duplication cyst of the duodenum. Pathol Int 2006;56:272-8.
14. You HS, Park SB, Kim JH, Lee HJ, Jang SP, Kim GH, *et al.* A case of duodenal duplication cyst manifested by duodenal polyp. Clin Endosc 2012;45:425-7.
15. Calder J. Medical essays and observations. Edinburgh: The Royal College of Physicians of Edinburgh; 1733. p. 205.
16. Puri, P., & Höllwarth, M. E. (2019). Pediatric Surgery. Springer Publishing.
17. Menon P, Rao KL, Thapa BR, Goyal R, Garge S, Rathore MK, *et al.* Duplicated gall bladder with duodenal duplication cyst. J Pediatr Surg 2013;48:e25-8.
18. Mirza B. Pyloroduodenal duplication cyst: The rarest alimentary tract duplication. APSP J Case Rep 2012;3:19.
19. Rai BK, Zaman S, Mirza B, Hanif G, Sheikh A. Duodenal Duplication Cyst having Ectopic Gastric and Pancreatic Tissues. APSP J Case Rep 2012;3:15.
20. Haliloglu M, Oto A, Karnak I, Tanyel FC, Eryilmaz M. Intrapancreatic duodenal duplication cyst with inversion of the superior mesenteric vessels: CT findings. Pediatr Radiol 2001;31:187-8.
21. GROSS RE, HOLCOMB GW Jr, FARBER S. Duplications of the alimentary tract. Pediatrics 1952;9:448-68.
22. Dipasquale V, Barraco P, Faraci S, Balassone V, De Angelis P, Di Matteo FM, *et al.* Duodenal Duplication Cysts in Children: Clinical Features and Current Treatment Choices. Biomed Hub 2020;5:152-64.
23. Holcomb GW 3rd, Gheissari A, O'Neill JA Jr, Shorter NA, Bishop HC. Surgical management of alimentary tract duplications. Ann Surg 1989;209:167-74.
24. Kawahara H, Takahashi T, Okada A. Characteristics of duodenal duplications causing pancreatitis in children and adolescents: A case report and review of the literature. J Pediatr Gastroenterol Nutr 2002;35:372-6.
25. Salazar E, Sin EI, Low Y, Khor CJL. Insulated-tip knife: An alternative method of marsupializing a symptomatic duodenal duplication cyst in a 3-year-old child. VideoGIE 2018;3:356-7.
26. Lecouffe P, Spyckerelle C, Venel H, Meuriot S, Marchandise X. Use of pertechnetate 99mTc for abdominal scanning in localising an ileal duplication cyst: Case report and review of the literature. Eur J Nucl Med 1992;19:65-7.