Isolated Duodenal Duplication Cyst in a Neonate

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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. 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. 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. 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, 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 discoloration 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.

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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/mm³, platelet count of 619,000/mm³, 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. 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. 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.

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. 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.

Duodenal duplication cysts represent a minor fraction of all the GI duplications, the estimated prevalence of which is <1 in 100,000 live births. The most common of the duplications occur in jejunum and ileum. The earliest descriptions were given by Calder. 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. 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,
Duodenal duplication cyst: A rare case entity in a neonate

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,[10] 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 extrophy 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 cranio-caudal 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.[16,19,23] 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.\textsuperscript{[16]}

Nowadays, minimally invasive treatment options are also growing which include endoscopic marsupialisation of duodenal duplication cyst.\textsuperscript{[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 variants 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 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.

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