CASE REPORT

Duodenal duplication Cyst in a Pediatric patient: A case report

Maryam Riaz Afridi, Jamal Ahmad Shah, Abdul Haseeb, Sara Kalsoom, Zia Ullah

Abstract:
Duodenal duplication cyst is a rare congenital anomaly linked to defects in the gastric duct canal during embryonic development.

Case Report:
We report a rare case of a 12-year-old pediatric female patient with a duodenal duplication cyst who presented with complaints of vomiting and epigastric pain from the past three weeks.

A Computed Tomography (CT) scan of the abdomen revealed a well-defined 5 x 4 x 3 cm peripherally enhancing cystic lesion. On surgical exploration, a cystic mass was found on the mesenteric border of the second part of the duodenum. A roux en-y-duodeno-jejunostomy was performed. A duodenal duplication cyst was diagnosed on histopathological analysis without any heterotopic mucosa.

We concluded that when a cystic formation next to the duodenum is seen on radiography, duodenal duplication should considered in the differential diagnosis of ambiguous upper abdominal symptoms. Total excision is the ideal course of action when possible. When correctly managed, these lesions frequently show positive results.

Keywords: Dilated duodenum, mucosa, GI tract, nausea and vomiting, cysts

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Khyber Teaching Hospital, Peshawar
MR Afridi,
JA Shah,
S Kalsoom
Z Ullah

Hayatabad Medical Complex, Peshawar
A Haseeb

Correspondence:
Dr. Jamal Ahmad Shah
Department of General Surgery, Khyber Teaching Hospital, Peshawar, Pakistan.
Cell No:+92 305-9496210
email: jamalshah1414@gmail.com

Introduction:
Duodenal duplication cyst is a rare congenital anomaly linked to defects in the gastric duct canal during embryonic development. It usually appears during infancy and occurs seldom in adulthood. Although it may also manifest in conjunction with obstructive symptoms, the most common examination finding is abdominal mass.[1]

A duplication cyst is distinguished by its location in or close to the GI tract's wall, lying on the mesenteric side, sharing a shared blood supply, and having a muscular wall with lining mucosa of any kind, including ectopic gastric, pancreatic, or respiratory tissue. Several hypotheses have been proposed to explain GI duplications, including the abortive twinning theory, the persisting embryologic diverticula theory, and the aberrant luminal recanalization idea.[2]

A thorough understanding of clinical and surgical knowledge enables effective management and treatment of duplication cysts. Although duodenal duplication cysts are relatively common in the pediatric population as compared to adults, the diagnosis is still a challenging task. Our case report will help surgeons make a timely diagnosis and management of the patient.

Case presentation:
The patient is a 12-year-old female child. No siblings or parents have a history of any congenital abnormality. This patient had a 3-week

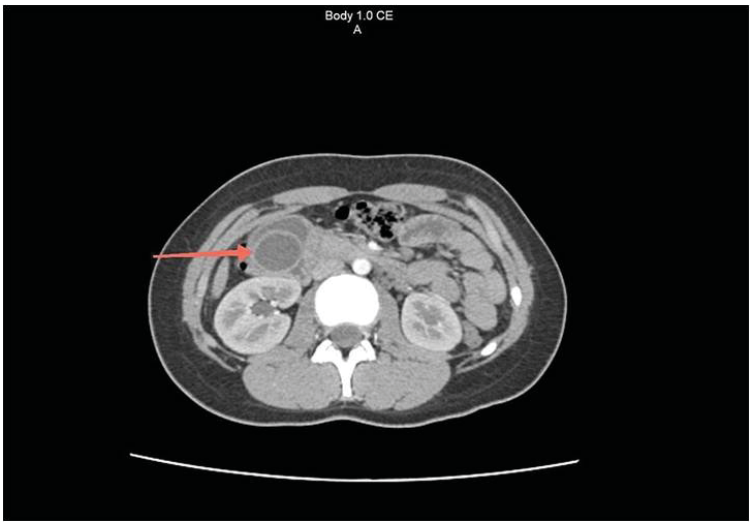

Figure 1: A CT scan showing an approximately 5 cm x 4 cm x 3 cm sized single cystic lesion with a double walled sign, pointed by Red arrow

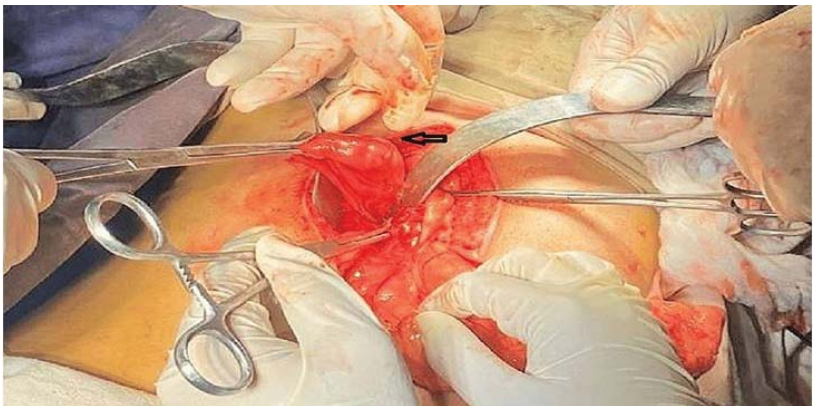

Figure 2: Intraoperative picture of a duodenal duplication cyst

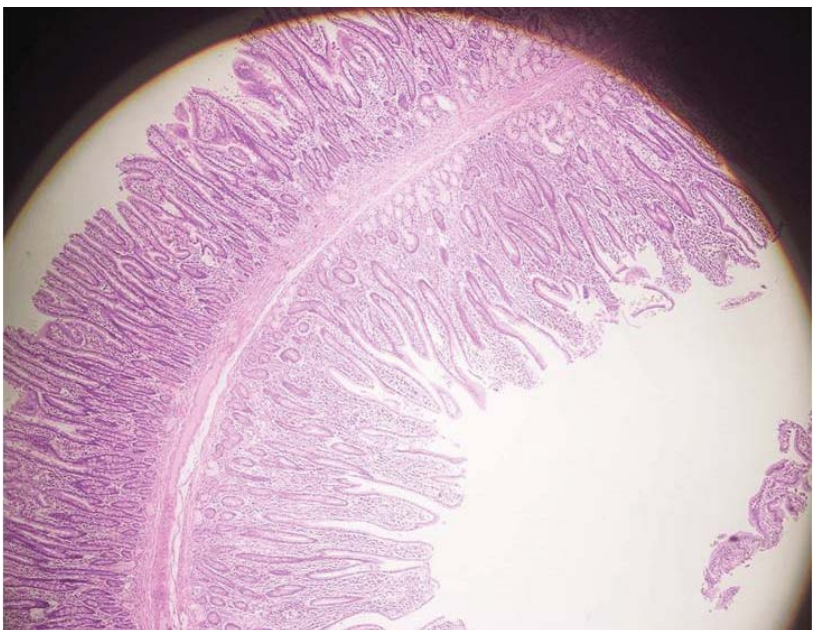

Figure 3: **Histopathology image reveals a cystic tissue fragment lined by unremarkable small bowel mucosa and underlying muscularis layer**

history of vomiting and pain epigastrium. She was initially managed in a local dispensary for symptomatic relief but her symptoms did not subside. She was brought to our hospital in the emergency department. On initial assessment, she was afebrile, with no pallor, cyanosis, or edema with the presence of mild dehydration, with a heart rate of 115 beats/min, a respiratory rate of 18 breaths/min, and oxygen saturation of 97% on room air.

On examination, no significant congenital abnormalities associated with duplication cysts such as bladder exstrophy, myelomeningocele, and imperforate anus were found. The abdomen was soft, with fullness present in the epigastric region with a palpable lump in the right hypochondrium. The lump was soft, fluctuant, mobile, non-pulsatile, non-tender, and roughly 5x3cm in size. The remainder of the systemic assessment was normal.

Laboratory investigations were normal with a hemoglobin of 13.2 g/dL and a total white blood cell count of 10,700/mm^3. An ultrasound of the abdomen showed a 6x3cm oblong cystic lesion in the right hypochondrium. Subsequently, a computed tomography (CT) imaging of the abdomen and pelvis was performed with intravenous contrast which showed an approximately 5cmx4cmx3cm sized well-circumscribed, round, fluid-filled, and unilocular with a thin enhancing wall at the level of 2nd part of the duodenum, having no communication with the pancreaticobiliary system as shown in figure-1.

A provisional diagnosis of a duodenal duplication cyst was made and the patient was optimized for surgery. After a detailed pre-operative workup including informed consent and counseling, the patient underwent exploratory laparotomy. Intra-operatively when the peritoneal cavity was opened, we noticed an approximately 4cm×3cm×3cm sized rounded cystic mass as shown in figure-2, which was attached to 2nd part of the duodenum and was communicating with the duodenal wall, over the mesenteric border. The cyst was intraluminal. The duodenum was opened and a large cystic mass of 5x4x3cm

was noted. Primary repair was not possible due to the removal of 4-5 cm of duodenal wall, so a roux-en-y duodenojejunostomy was performed. The specimen was sent for histo-pathological studies which revealed cystic tissue fragments lined by unremarkable small bowel mucosa and underlying muscular layer confirmatory of duodenal duplication cyst as shown in figure-3.

The post-operative recovery of the patient was uneventful. On day 5th she was discharged from the hospital on oral medications and subsequent follow-up visits up to three months, she was doing well.

Discussion:

The presence of epithelial mucosal lining, a thick layer of smooth muscle, and a close attachment to the natural gastrointestinal system characterize duplication cysts, which are an uncommon type of congenital anomaly with an incidence of 1:4500 live births that can arise anywhere along the alimentary tract. The distal ileum is where duplication cysts are most frequently found, followed by the esophagus, colon, and jejunum. Only 5 to 7% of all gastrointestinal tract duplication cysts are located in the duodenum.[3] The name of a duplication cyst is determined by the area of the gastrointestinal system in which it develops rather than the type of mucosa it contains. Although they are connected to the digestive tract, duplication cysts often do not exhibit luminal communication.[4]

47 duodenal duplication cyst cases were evaluated by Chen et al.[5] The clinical characteristics of duodenal duplication cysts range from asymptomatic cases to non-specific symptoms like nausea, vomiting, and abdominal pain. Small bowel obstruction or gastric outlet obstruction symptoms can appear in some people. Duodenal hemorrhage or peritonitis may result from an ulcer or perforation brought on by the presence of an ectopic mucosa.[6] Important pre-operative diagnosis methods include endoscopy, magnetic resonance cholangiopancreatography (MRCP), CT scan, upper gastrointestinal contrast study, and ultrasound.

We diagnosed our case with a CT scan which showed a cyst that was well-circumscribed, round, fluid-filled, and unilocular with a thin, enhancing wall. Peristalsis and the pathognomonic "double-wall" sign, which consists of an outer hypoechoic layer reflecting the muscularis propria and an inner hyperechoic rim associated with the mucosa-sub mucosa, are two ultrasonography (US) indications that are strongly diagnostic of intestinal duplication.[7,8] However, the ultrasound in our case was unremarkable regarding these signs and it just showed a cystic lesion in the right hypochondrium.

For most of the duodenal duplication cysts, surgical resection is the preferred course of action. The location, size, and proximity to the pancreaticobiliary duct or duodenal wall determine surgical management. The preferred treatment for small duodenal cysts that are not connected to the pancreatic, biliary, or duodenum is total excision. Pancreaticoduodenectomy (PD) may be required if the duodenal cysts are connected with the pancreaticobiliary tree.[3,8]

In our case, the duodenum was opened and it was seen that a large cystic mass of 5x4x3cm was communicating the duodenal wall. Primary repair was not possible due to the removal of 4-5cm of duodenal wall, so a roux-en-y duodenojejunostomy was performed. Uzun MA reported a case of a 38-year-old female with gastric outlet obstruction who was managed by an endoscopic approach what they call an intraduodenally performed internal derivation.[9]

Newer techniques such as Endoscopic submucosal dissection and endoscopic mucosal resection, offer a less invasive prospective resection alternative for low malignant potential lesions of the alimentary tract.[10] Additionally, they have been frequently used to treat submucosal lesions and have occasionally been successful in treating older children and adults who have duplication cysts.[11] In our case the cyst was intraluminal, large in size, communicating with the duodenal wall, and due to lack of expertise, the endoscopic excision was not feasible so we managed our patient surgically.

Conclusions:
In particular, when a cystic formation next to the duodenum is seen on radiography, duodenal duplication should be considered in the differential diagnosis of ambiguous upper abdominal symptoms. Total excision is the ideal course of action when possible. If not, internal derivation and/or subtotal excision may be used to treat the cyst. When properly cared for, this lesion often provides favorable outcomes

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Role and contribution of authors:
Maryam Riaz Afridi, collected the data, references and did the initial writeup.

Jamal Ahmad Shah, collected the data and helped in discussion writing.

Abdul Haseeb, collected the references and wrote the introduction and discussion.

Sara Kalsoom, collected the data, references and advised useful changes.

Zia Ullah, critically review the article and made final changes.

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