Isolated duodenal duplication cyst presenting as a complex solid and cystic mass in the upper abdomen

Salina D. Tsai[1*], Sabrina C. Sopha[2], Elliot K. Fishman[1]

1. The Russell H. Morgan Department of Radiology and Radiological Science, Johns Hopkins Medical Institutions, Baltimore, MD, USA

2. Department of Pathology, Johns Hopkins Hospital, Baltimore, MD, USA

* **Correspondence:** Salina D. Tsai, 601 North Caroline St. JHOC 3140F, Baltimore, MD 21287, USA (*stsai17@jhmi.edu*)

Radiology Case. 2013 Nov; 7(11):32-37 :: DOI: 10.3941/jrcr.v7i11.1785

ABSTRACT

Duodenal duplication cysts are a rare subtype of gastrointestinal duplications cysts. Approximately 5% of gastrointestinal duplication cysts occur in the duodenum. An 18-year-old woman presented with epigastric pain and a subjective abdominal bulge. A computed tomography scan was subsequently performed and showed a solid and cystic mass with wall calcifications in the lesser sac of the upper abdomen. A duodenal duplication cyst was found unexpectedly on histopathologic analysis. This was also an unusual case as there was no evidence of malignancy. Four years after surgery, the patient remains asymptomatic. We present a brief literature review on duodenal duplication cysts and discuss its differential diagnosis.

CASE REPORT

CASE REPORT

An 18-year-old woman presented to surgical clinic with a one-month history of a subjective waxing and waning bulge in the upper abdomen associated with transient sharp epigastric pain. The patient reported no other symptoms and had no relevant medical history. The mass was not palpable on physical examination. Serum CA 19-9 level was 98.9 U/ml (normal, 0.0-36.0 U/ml), and CEA level was 7.1 ng/ml (normal, 0.0-3.0 ng/ml), both elevated.

A dual-phase IV-contrast-enhanced computed tomography (CT) scan was subsequently performed in the arterial and venous phases. Sagittal and coronal multiplanar reformatted images and 3D volume rendering were also performed. CT scan of the abdomen demonstrated a heterogeneous, well-circumscribed 8.2 x 8.0 x 6.9 cm mass in the lesser sac (Fig. 1). The stomach was displaced slightly anteriorly and the duodenum laterally. The mass did not appear to arise from or connect with any neighboring organs. The mass was mostly cystic but demonstrated a thick soft tissue rim at the inferior aspect. Coarse calcifications were present in the inferior wall (Fig. 2). Fat was not present. The pancreatobiliary ductal system showed no mass effect. There was moderate extrinsic compression of the portal confluence. There was no evidence of metastatic disease or masses elsewhere in the abdomen.

Due to equivocal imaging findings and abdominal pain, the patient was referred for surgical removal. At laparotomy, a mass in the lesser sac was found, consistent with imaging findings. Gross pathologic examination demonstrated a 2.0 kg, 9.5 x 6.4 x 4.4 cm unilocular cystic mass with 200 mL tenacious mucus and a 0.2-1.0 cm thick wall. Microscopic examination showed a multilayered structure recapitulating small bowel, with focally ulcerated intestinal mucosa containing rare Paneth cells, muscularis mucosa, submucosa, and a double-layered muscularis propria with a myenteric neural plexus (Fig. 3). Foci of reactive histiocytes with calcifications were numerous. Four years after surgery, the patient remains well and asymptomatic.

DISCUSSION

Gastrointestinal duplication cysts are rare (1 per 100,000 births), occurring most commonly in the small intestine (ileum and jejunum) (47%), followed by the colon (20%), esophagus

(17%), stomach (8%), and duodenum (2-12%) [1-4]. Macpherson evaluated a series of 281 patients with gastrointestinal duplication cysts at all levels of the gastrointestinal tract and found that 5% of duplication cysts (14/281) occurred in the duodenum [5].

Calder gave the first description of a duodenal duplication. By definition, a duplication cyst is located in or immediately adjacent to the wall of the gastrointestinal tract on the mesenteric side, shares a common blood supply, contains a muscular wall, and contains lining mucosa of any type, including ectopic gastric, pancreatic, and respiratory tissue [1-9]. Duplication cysts are thus named according to the portion of the gastrointestinal tract in which they occur rather than by the type of mucosa they contain [5]. Duplication cysts are attached to the gastrointestinal tract but typically do not show luminal communication [5,6].

Duodenal duplication cysts are usually located posteromedial to the second and third duodenal segments and closely associated with the pancreatobiliary duct system [10]. Duodenal duplication cysts are usually spherical and noncommunicating, as seen in this patient [5]. Rarely, they may be tubular structures [5]. Duodenal duplication cysts have a well-developed smooth muscle coat and share a common wall with the native duodenum. Luminal communication is seen in 25% of cases [1].

Etiology and demographics:

The etiology of gastrointestinal duplication remains unknown although several theories have been proposed such as the abortive twinning theory (representing incomplete twinning), the persistent embryologic diverticula theory, and the aberrant luminal recanalization theory [5]. Duodenal duplication cysts are most commonly discovered in newborns or in early childhood but may present at a later age [5,11]. Males and females are approximately equally affected [4,8]. In a meta-analysis of 47 cases of duodenal duplication cysts, 44% of duodenal duplication cysts were 2.0 - 4.0 cm in size [4]. In the same series, approximately 40% of duodenal duplication cysts are discovered in the first decade of life, 21.3% in the second decade, and the remaining 38.3% are discovered after age 20, decreasing with increasing age [4].

Clinical and Imaging Findings:

The most commonly presenting symptoms are nonspecific abdominal pain, nausea and vomiting. Other presenting symptoms include gastrointestinal hemorrhage, intussusception, obstruction, jaundice, and pancreatitis [1,3,4,12]. In infants, they can be asymptomatic [13]. In the first decade, a common clinical presentation is nausea and vomiting; although in older age groups, they can present as a painful, palpable mass [4,13]. This patient was 18 years old, slightly older than most discovered gastrointestinal duplication cysts, but within an acceptable age range and with the expected symptomatology. There are case reports of duodenal duplication cysts communicating with the pancreatic duct [1,9]. Complications of duodenal duplication cysts include bowel obstruction, bile duct obstruction and pancreatitis, bleeding, intussusception, and malignancy [4,5].

Ectopic gastric and pancreatic mucosa are the only clinically significant mucosal lining [5]. Duodenal duplication cysts may be lined by ectopic gastric epithelium in 15% of cases, predisposing to ulceration, bleeding, and perforation [1]. Additionally, duodenal duplication cysts may harbor secondary malignancies [2,14,15]. Important clinical associations include intestinal or biliary atresia, malrotation, imperforate anus, double gallbladder, double uterus, partial gastric diverticulum, complete large bowel duplication, situs inversus, and intraspinal neuroenteric cyst [4,8].

Duplication cysts have an enteric mucosal lining with a double muscle layer of inner circular and outer longitudinal smooth muscle and neural plexus. Accordingly, on ultrasound characteristic gut wall signature and mural layers may be seen manifested as an echogenic mucosal layer and hypoechoic muscle layer. Therefore, ultrasound may be used to demonstrate bowel wall layers in order to distinguish duplication cysts from other cystic lesions. Peristaltic activity may also be seen favoring a duplication cyst. Duplication cysts are usually cystic and may contain debris due to hemorrhage or inspissated material or may be simple-appearing [4,6,16]. On CT, duplication cysts are usually well-circumscribed, round, fluid-filled and unilocular with a thin, enhancing wall [5]. Mural nodularity or a soft tissue component raises suspicion of malignancy [10]. Case reports of carcinoid tumor and adenocarcinoma arising in a duodenal duplication cyst have been described [2,14,15]. Wall calcifications have been reported in duodenal duplication cysts [16,17]. This case is an atypical example of a duodenal duplication cyst due to the nodular and diffusely thickened wall as well as the prominent soft tissue component in the inferior aspect of the mass without harboring malignancy.

Treatment and prognosis:

Surgical excision either open or laparoscopic is required for diagnosis, symptomatic relief, and to exclude malignancy. Prognosis is excellent if the lesion is surgically excised and metastatic disease is not present [1,9,12].

Differential diagnosis:

Other cystic lesions that may be considered in the differential diagnosis based on clinical and radiologic findings are gastrointestinal stromal tumors, gastrointestinal teratomas, neurogenic tumors, and mesenteric cysts.

A gastrointestinal stromal tumor (GIST) was included in the differential diagnosis due to the prominent cystic component and location adjacent to the bowel. A soft tissue mass may be suggested on radiographs which may cause small bowel dilation or an irregular gas collection may be seen. GISTs may become cystic due to necrosis, cavitation, or cystic degeneration and rarely contain calcifications. GISTs are also usually round and exophytic, as was seen in our case, and due to their origin from the muscularis propria, they tend to grow exophytically. GISTs demonstrate variable signal intensity on magnetic resonance imaging (MRI) depending on the degree of hemorrhage or necrosis [18-20].

Gastrointestinal teratomas, which may also be cystic, are rare tumors usually found in infants and children, and are generally benign. On CT or MRI, they are usually solid and cystic masses, contain calcifications, and usually contain a fatty component. They are generally similar in appearance on CT to ovarian teratomas. Fat however was not seen in the mass in our patient. Shadowing from calcifications can be seen with ultrasound [21].

A neurogenic tumor of the abdomen was also considered. These tumors may arise from ganglion cells (such as ganglioneuromas), from the paraganglionic system (such as paragangliomas), or from nerve sheaths in the abdomen (such as schwannomas). Occasionally, these tumors arise in the abdomen from the bowel wall or from the abdominal wall. Ganglioneuromas of ganglion cell origin are usually found in children and young adults. In the abdomen, they usually arise in the retroperitoneum or along the paravertebral sympathetic plexus. They are usually well-defined masses with variable enhancement and may contain punctate calcifications. Paragangliomas usually are avidly enhancing and show necrosis and hemorrhage. Schwannomas also are usually well-defined, homogeneous or heterogeneous with solid and cystic components, and smooth with variable enhancement. In adults, neurogenic tumors are usually benign [22-24].

Mesenteric cysts such as enteric or mesothelial cysts could also be considered. However, a thin wall and no calcifications are usually seen, in contrast to our case. High signal intensity on T2-weighted MRI images is also seen due to fluid components. A lymphangioma was similarly not considered as it usually has a thin wall, can be multi-loculated, and may contain debris. Lymphangiomas usually do not contain calcifications and may cross retroperitoneal compartments [25].

TEACHING POINT

Duodenal duplication cysts are usually well-defined, thin-walled structures filled with fluid on CT. Duodenal duplication cysts which contain a solid component are concerning for malignancy and should be surgically removed.

REFERENCES

1.Prasad TR, Tan CE. Duodenal duplication cyst communicating with an aberrant pancreatic duct. Pediatr Surg Int. 2005;21:320-322. PMID: 15723222

2. Hata H, Hiraoka N, Ojima H et al. Carcinoid tumor arising in a duplication cyst of the duodenum. Pathol Int. 2006;56:272-278. PMID: 16669876

3. You HS, Park SB, Kim JH et al. A case of duodenal duplication cyst manifested by duodenal polyp. Clin Endosc. 2012;45:425-427. PMID: 23251893

4. Chen JJ, Lee HC, Yeung CY et al. Meta-analysis: the clinical features of the duodenal duplication cyst. J Pediatr Surg. 2010:45:1598-1606. PMID: 20713206

5. Macpherson RI. Gastrointestinal tract duplications: clinical, pathologic, etiologic, and radiologic considerations. Radiographics. 1993;13(5):1063-1080. PMID: 8210590.

6. Hsu H, Gueng MK, Tseng YH et al. Adenocarcinoma arising from colonic duplication cyst with metastasis to omentum: a case report. J Clin Ultrasound. 2011;39(1):41-43. PMID: 20812340

7. Emoto K, Mukai M, Kawachi S et al. Duplication of the digestive organs in the retroperitoneum: a case report with reference to the importance of a standardized nomenclature and definition. Pathol Int. 2011;61:430-434. PMID: 21707847.

8. Gross RE, Holcomb Jr. GW, Farber S. Duplications of the alimentary tract. Pediatrics. 1952;9:449-468. PMID: 14920118.

9. Koffie RM, Lee S, Perez-Atayde A, Mooney DP. Periampullary duodenal duplication cyst masquerading as a choledochocele. Pediatr Surg Int. 2012;28:1035-1039. PMID: 22752148.

10. Jayaraman MV, Mayo-Smith WW, Movson JS, Dupuy DE, Wallach MT. CT of the duodenum: an overlooked segment gets its due. Radiographics. 2001;21:S147-160. PMID: 11598254.

11. Berrocal T, Torres I, Gutiérrez J, et al. Congenital anomalies of the upper gastrointestinal tract. Radiographics. 1999;19:855-872. PMID: 10464795.

12. Meier AH, Mellinger JD. Endoscopic management of a duodenal duplication cyst. J Pediatr Surg. 2012;47:E33-E35. PMID: 23164028

13. 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.

14. Falk GL, Young CY, Parer J. Adenocarcinoma arising in a duodenal duplication cyst: a case report. Aust N Z J Surg. 1991;61:551-553. PMID: 1859319.

15. Inoue M, Nishimura O, Andachi H, Koga S. Early cancer of duodenal duplication a case report. Gastroenterol Jpn. 1979;14(3):233-237. PMID: 447002.

16. Kim SK, Kim HK, Lee SJ, Park CK. Completely isolated enteric duplication cyst: case report. Abdom Imaging. 2003;28:12-14. PMID: 12483377.

17. Alford BA, Armstrong P, Franken EA, Shaw A. Calcification associated with duodenal duplications in children. Radiology. 1980;134: 647-648. PMID: 7355213.

18. Kim HC, Lee JM, Son KR et al. Gastrointestinal stromal tumors of the duodenum: CT and barium study findings. Am J Roentgenol. 2004;183:415-419. PMID: 15269035.

19. Levy AD, Remotti HE, Thompson WM, Sobin LH, Miettinen M. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. Radiographics. 2003;23(2): 283-304. PMID: 12640147.

20. Burkill GJ, Badran M, Al-Muderis O et al. Malignant gastrointestinal stromal tumor: distribution, imaging features, and pattern of metastatic spread. Radiology. 2003;226:527-32. PMID: 12563150.

21. Bowen B, Ros PR, McCarthy MJ, Olmsted WW, Hjerstad BM. Gastrointestinal teratomas: CT and US appearance with pathologic correlation. Radiology. 1987;162(2):431-433. PMID: 3541031.

22. Rha SE, Byun JY, Jung SE et al. Neurogenic tumors in the abdomen: tumor types and imaging characteristics. Radiographics. 2003;23(1):29-43. PMID: 12533638.

23. Choi H, Whitman G, Ro J, Charnsangavej C. Benign schwannoma in the porta hepatis. AJR Am J Roentgenol. 2001;177: 652. PMID: 11517064.

24. Levy AD, Quiles AM, Miettinen M, Sobin LH. Gastrointestinal schwannomas: CT features with clinicopathologic correlation. AJR Am J Roentgenol. 2005;184:797-802. PMID: 15728600

25. Yang DM, Jung DH, Kim H et al. Retroperitoneal cystic masses: CT, clinical, and pathologic findings and literature review. Radiographics. 2004;24(5):1353-1365. PMID: 15371613.

FIGURES

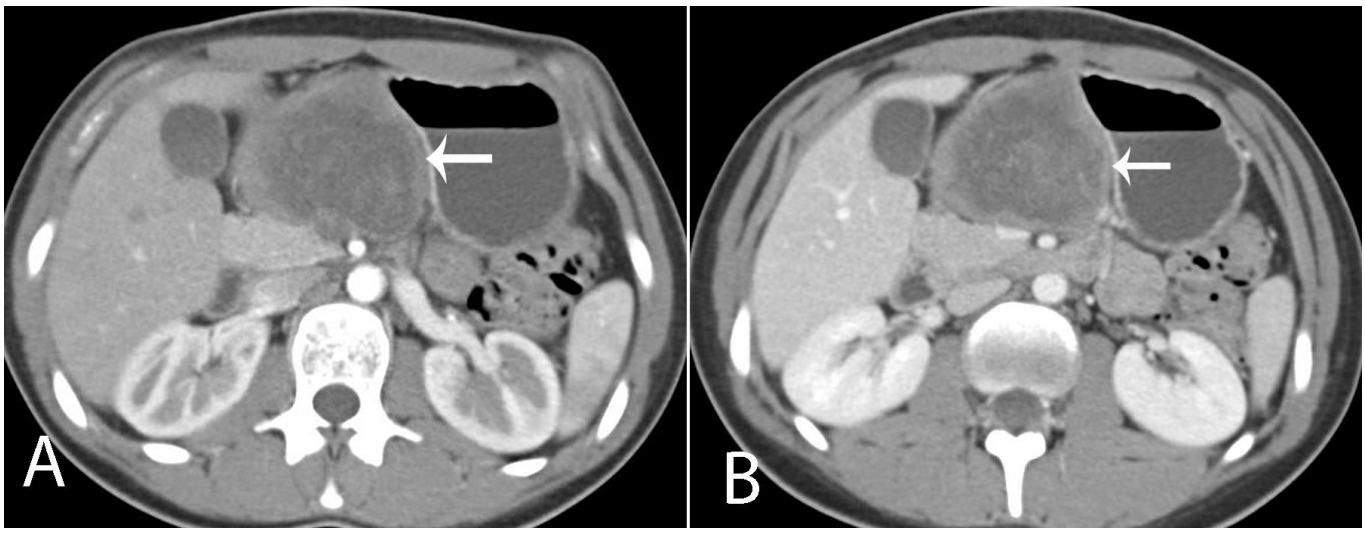

Figure 1: 18-year-old woman with a duodenal duplication cyst presenting with epigastric pain and abdominal bulge. Axial CT images in the arterial phase (A) and venous phase (B) of the upper abdomen show a 8.2 x 8.0 x 6.9 cm smoothly-marginated mass (arrow) in the lesser sac posterior to the stomach and medial to the first portion of the duodenum. (Protocol: Siemens CT scanner, 225 mA, 120kV, 5mm slice thickness, 100 cc IV Omnipaque 350).

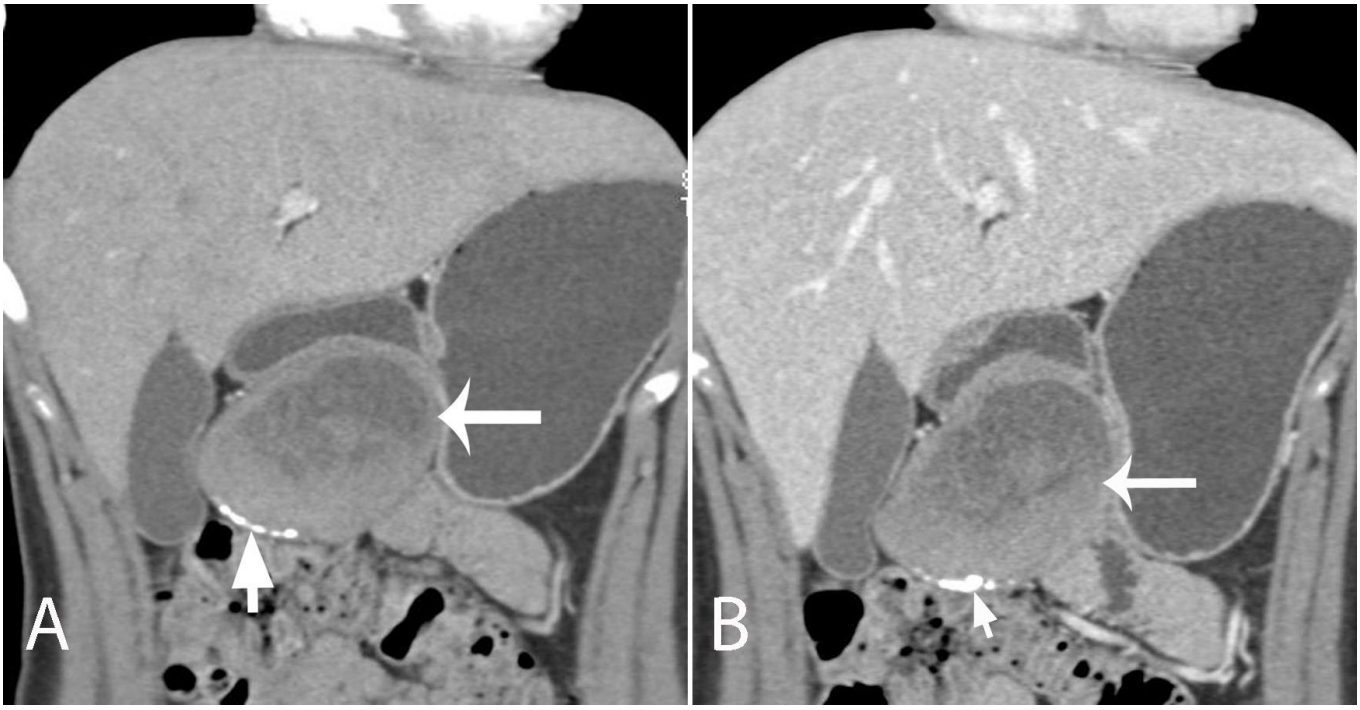

Figure 2: 18-year-old-woman with a duodenal duplication cyst presenting with epigastric pain and abdominal bulge. Coronal reformatted CT images of the upper abdomen in the arterial phase (A) and venous phase (B) show a 8.2 x 8.0 x 6.9 cm well-circumscribed mass (large arrow) that is mostly cystic superiorly but with some solid component in the inferior aspect. Coarse calcifications are present in the inferior wall of the mass (small arrow). (Protocol: Siemens CT scanner, 225 mA, 120kV, 5mm slice thickness, 100 cc IV Omnipaque 350).

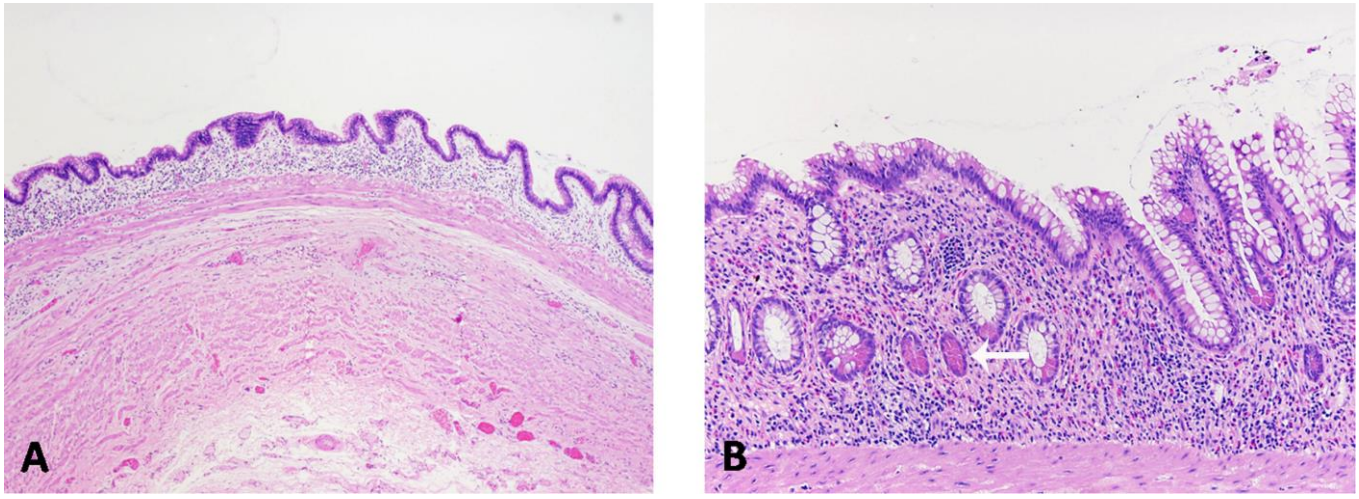

Figure 3: 18-year-old-woman with a duodenal duplication cyst. Photomicrographs of hematoxylin and eosin-stained tissue sections showing the normal complement of gastrointestinal wall layers (a, 40X magnification), with foci of Paneth cells (arrow in b, 100X magnification) supporting small bowel origin.

Etiology	Unknown
Incidence	1 in 100,000 births
Gender ratio	Approximately equal among males and females
Age predilection	Most common in newborns and early childhood
Treatment	Surgical resection especially if symptomatic
Prognosis	Excellent without evidence of malignancy or metastatic disease

Table 1: Summary table for duodenal duplication cyst

Differential	Plain X-ray	Ultrasound	CT	MRI
Gastrointestinal stromal tumor	Small bowel dilation or a soft tissue mass may be seen, irregular gas collection if cavitary	Hypoechoic, submucosal	Prominent cystic component, round, usually exophytic located adjacent to bowel, can be intraluminal	Variable signal intensity depending on degree of hemorrhage or necrosis. Gadolinium enhancement can be seen in solid components
Teratoma	Large soft tissue mass, can contain coarse calcifications	Predominantly cystic mass to predominantly solid mass with multiple cysts, can see shadowing from calcifications	Solid and cystic mass containing calcifications and fat	Fatty component high signal intensity on T1-weighted images, variable signal intensity on T2-weighted images
Schwannoma	May show subtle soft tissue mass or calcifications	Solid and cystic mass	Well-defined, homogeneous or heterogeneous, smooth, variable enhancement	Low signal intensity on T1-weighted images. Heterogeneous high signal intensity on T2-weighted images, enhancing solid components
Mesenteric cyst	Noncalcified mass displacing bowel may be seen	Unilocular cyst, thin walls, may contain few septations	Thin wall, no calcifications	High signal intensity on T2-weighted images due to fluid components.
Lymphangioma	Noncalcified mass displacing bowel may be seen	Predominantly cystic with multiple thin septations. No solid component.	Thin wall, no calcifications, may cross compartments	High signal intensity on T2-weighted images due to fluid components

Table 2: Differential diagnosis table for duodenal duplication cyst

ABBREVIATIONS

CA 19-9 = cancer antigen 19-9
CEA = carcinoembryonic antigen
CT = computed tomography
GIST = gastrointestinal stromal tumor
MRI = magnetic resonance imaging

KEYWORDS

Duodenum; Duplication cyst; Gastrointestinal stromal tumor; Computed tomography; Lesser sac

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/1785

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad

www.EduRad.org