

Case Report

Gastric bronchogenic cyst presenting as a gastrointestinal stromal tumor

Longhao Sun, Li Lu, Weihua Fu, Weidong Li, Tong Liu

Department of General Surgery, Tianjin Medical University General Hospital, Tianjin 300052, China

Received May 3, 2015; Accepted June 22, 2015; Epub October 1, 2015; Published October 15, 2015

Abstract: Bronchogenic cyst (BC) is congenital abnormality of the tracheobronchial bud derived from the primitive foregut which is predominantly found in the mediastinum. Surgery remains the most common treatment when malignancy is suspected, or when there are presenting. Only infrequently, is BC located at an extrathoracic site. Although rarely located in the stomach, BC should be considered as a differential diagnosis of gastric neoplasm. For these cases, surgery remains a common choice. Minimally invasive procedures such as endoscopic ultrasonography-guided fine needle biopsy aspiration (EUS-FNA) and endoscopic submucosal dissection (ESD) should also be considered when the diagnosis of BC is suspected. They are valuable diagnostic methods that can assess and identify the location of the lesion, and facilitate histological examination of the cyst. In some cases of more superficial lesions ESD can take the place of surgery as it avoids unnecessary complications of a more invasive procedure. Here we present a case of gastric BC located in the fundus of the stomach that resembled a gastrointestinal stromal tumor (GIST). We discuss its embryology, pathogenesis, radiological, clinical and treatment modalities. We also provide a thorough review of 14 cases (including our own case), which completely meet pathological criteria has been undertaken focusing on symptom, location, treatment, and histological features.

Keywords: Bronchogenic cyst, gastrointestinal stromal tumor (GIST), stomach

Introduction

Bronchogenic cyst (BC) is a congenital abnormality of the tracheobronchial bud arising from the outward protruding pouches of the primitive foregut, between the 3 and 7 wells prenatally. Consequently, the cysts are usually detected in newborn infants or children [1]. Most frequently, the lesions are located in the mediastinum, pericardial areas, or pulmonary parenchyma, depending on the level of abnormal budding that occurred during development.

The thoracic and abdomen cavity are linked by the pericardioperitoneal canal in the early embryonic stage. This canal is divided into two cavities by fusion of the pleuroperitoneal membranes, which form the components of the diaphragm. However, if there is complete separation and migration through the canal, the abnormal buds of the tracheobronchial tree are pinched off and migrate into the abdomen, retroperitoneum and diaphragm, resulting in extrathoracic BC.

Gastric BC is extremely rare among cases of extrathoracic BC [2]. Here we present a case of gastric BC located in the fundus of the stomach that resembled a gastrointestinal stromal tumor (GIST). We discuss its embryology, pathogenesis, radiological, clinical and treatment modalities. We also provide an overview of literature cases of gastric BCs.

Case report

A 67-year-old male was admitted with a 2 year history of dull epigastric pain. There was no dysphagia, vomiting, change in bowel habit or other significant symptoms. The patient had a duodenal peptic ulcer perforation 3 years ago which was treated without surgical intervention at a local hospital.

Physical examinations were unremarkable. Most blood tests, including tumors markers (carcinoembryonic antigen, carbohydrate antigen 50 and carbohydrate antigen 125), were normal. But serum carbohydrate antigen 19-9 (CA19-9)

Gastric bronchogenic cyst presenting as GIST

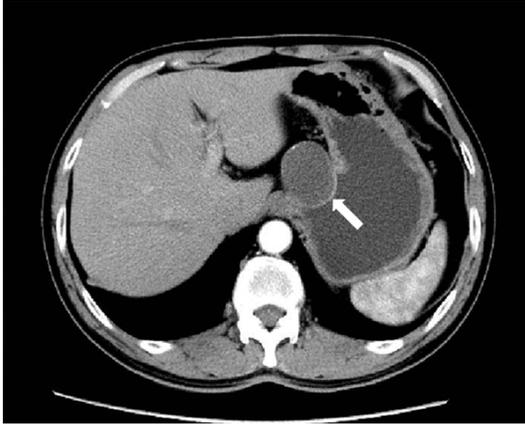


Figure 1. CT scan of the abdomen. CT scan of the abdomen demonstrated a well-defined, 42×32 mm low-density, ovate, cystic lesion located in the fundus of stomach with cyst wall enhancement and punctate calcifications (white arrow).

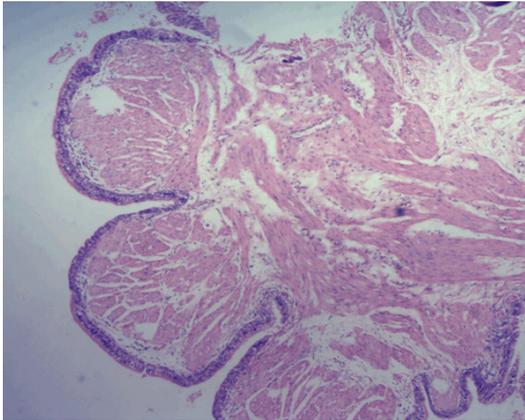


Figure 2. Histopathology. Histopathology showed pseudostratified ciliated columnar epithelium, cartilage, seromucous gland and smooth muscle (hematoxylin and eosin stain, ×40).

was elevated to 62.04 U/mL (normal range: 0-37 U/mL).

Abdominal contrast-enhanced computed tomography demonstrated a well-defined, 42×32 mm ovate, low density, cystic lesion located in the fundus of stomach, with cyst wall enhancement and punctate calcification (**Figure 1**). Upper GI endoscopy showed mild oesophagitis and a normal gastric mucosa. Endoscopic ultrasound (EUS) confirmed that a 41×32 mm lesion was located in the muscularis propria of stomach. It also identified a unilocular cyst containing echogenic spots suggestive of mucus. In view of the presence of intra-abdominal adhesions

which were a legacy of previous duodenum perforation, it was not possible to undertake endoscopic ultrasonography-guided fine needle biopsy aspiration (EUS-FNA). Therefore a presumptive diagnosis of GIST with cystic degeneration was made based on the radiological findings. Laparoscopic procedures and other minimally invasive techniques were also excluded due to the potential surgical risk caused by intra-abdominal adhesions.

During laparotomy, a 50 mm ovate cyst was found in the fundus of the stomach close to the esophagogastric junction (EGJ). The cyst was entirely located in the stomach wall with no connections to the digestive lumen. Based on these findings proximal gastrectomy and esophagogastric anastomosis was performed and the lesion was removed completely because of the possibility of malignancy.

Histopathologic examination indicated a cystic structure filled with thick white mucus. Microscopically the cyst was partially lined with pseudostratified ciliated columnar epithelium (**Figure 2**). Islands of cartilage, serous glands and smooth muscle fibers were also present. The adjacent gastric mucosa appeared normal. Based on these findings the lesion was histologically diagnosed as a bronchogenic cyst.

The postoperative course was uneventful and the patient was discharged 8 days postoperatively. He had remained asymptomatic throughout biweekly follow-ups for 3 months.

Discussion

BC is a rare congenital bronchopulmonary lesion with prevalence between 1 in 68,000 and 1 in 42,000 [3]. Only sporadic cases of gastric BC have been reported in the literature. A thorough review of 14 cases (including our own case) summarized in **Table 1**, which completely meet pathological criteria has been undertaken focusing on symptom, location, treatment, and histological features. The age of the patients ranged widely, from 25 to 81 years; the ratio of males to females was 1 to 2.5. Eight of the 14 cases were asymptomatic and discovered incidentally. Symptoms in the other six patients were generally nonspecific, including intermittent nausea, vomiting, epigastric discomfort, abdominal pain, or dysphagia. The symptoms may have resulted from cyst expansion.

Gastric bronchogenic cyst presenting as GIST

Table 1. Bronchogenic cysts of stomach

Case No.	Reference	Year	Age/Gender	Symptoms	Location	Size (mm)	CT	MRI	B-US	EUS	Preoperative diagnosis	Treatment	Contents	Pathological findings			
														pseudostratified ciliated columnar epithelium	cartilage	smooth muscle	sero-mucous glands
1	Braffman [12]	1988	64/F	nausea, vomiting	posterior wall of the fundus	150×80	thin-walled, loculated lesion	-	-	-	gastric duplication, gastric diverticulum, pseudodiverticulum, pancreatic pseudocyst	laparotomy	not described	√	√	√	√
2	Song [13]	2005	62/F	-	lesser curvature	17×17	solid, homogeneous, low-density nodule	-	solid, homogeneous, low-density nodule	-	GIST	laparotomy	gelatinous mucoid	√	×	√	√
3	Melo [14]	2005	39/F	rib pain	posterior wall of the fundus	60×60	mass	-	-	-	GIST	laparoscopy	viscous mucoid cream-colored	√	×	√	×
4	Lee [15]	2006	38/F	-	cardia	70×70	solid nodule	-	-	-	GIST	endoscopic mucosal resection	serious yellow	√	×	√	√
5	Wakabayashi [16]	2007	37/M	upper abdominal dull pain, dysphasia	lesser curvature	50×50	cyst	-	cyst	cystic lesion	duplication cyst	laparoscopy	yellow-greenish turbid	√	×	√	×
6	Shibahara [17]	2008	43/M	epigastric pain	lesser curvature of the cardia	90×40	cyst	cyst	hepatic cyst	-	hepatic cyst	laparotomy	greenish gelatinous	√	×	√	√
7	Sato [18]	2008	60/F	-	lesser curvature of the cardia	30×30	low-density, rounded lesion	-	-	cystic lesion	cystic neoplasm	EUS-FNA biopsy	yellowish mucous	√	×	×	√
8	Jiang [19]	2010	25/F	epigastric pain	fundus	30×30	-	-	-	-	cystic neoplasm	laparotomy	yellow mucous	√	×	√	√
9	Tan [6]	2010	30/F	-	posterior wall	60×30	cyst	-	cyst	cystic lesion	pancreatic cyst	laparoscopy	gelatinous	√	×	√	√
10	Hosomura [20]	2011	44/F	-	posterior wall	75×65	homogeneous low-density mass	T1: low signal intensity, T2: high signal intensity	-	-	pancreatic cyst, GIST	laparotomy	not described	√	×	√	×
11	Ubukata [21]	2011	81/F	-	lesser curvature	26×26	homogeneous cyst	T1: low signal intensity, T2: high signal intensity	-	-	congenital cyst	laparotomy	clear and whitish mucous	√	×	√	√

Gastric bronchogenic cyst presenting as GIST

12	Yang [22]	2013	50/M	-	fundus and cardia	80×60	homogeneous lesion	-	cyst	-	retroperitoneal mass	laparotomy	white mucus gelatinous	√	√	√	√
13	Yang [22]	2013	37/F	-	posterior wall	100×60	homogeneous low-density cystic lesion	-	-	-	GIST, lymphangioma	laparotomy	yellowish gelatinous, mucoid	√	√	√	×
14	present case	2013	67/M	epigastric pain	fundus	50×50	low-density lesion	-	-	cystic lesion	GIST	laparotomy	white mucous	√	√	√	√

CT computerized tomography, MRI magnetic resonance imaging, EUS endoscopic ultrasonography, B-US b ultrasonography, FNA fine needle aspiration.

Gastric bronchogenic cyst presenting as GIST

sion with compression of adjacent viscera, rupture, or secondary infection [4]. Twelve patients underwent laparotomy or laparoscopy for potential malignancy. Two patients underwent minimally invasive procedures, such as endoscopic mucosal resection and EUS-FNA, which prevented the need for gastrectomy.

In general blood tests showed no characteristic changes. However, one unusual feature of these cases was elevated CA 19-9 level. CA19-9 is known to be elevated in pancreatic, colorectal and hepatocellular carcinoma. Elevated levels may also be found in patients with pancreatitis, cirrhosis, and obstructive disease of the bile duct. In the context of MC, it is likely that the epithelial cells secrete CA 19-9 due to the foregut origin of these lesions. The high serum levels of CA 19-9 are possibly caused by a gradual accumulation in the cyst and absorption into the serum. Several published case reports have shown that BC is associated with elevated serum CA 19-9 [5]. Most of these cases were intrathoracic: only one was gastric [6].

CT and magnetic resonance imaging (MRI) are frequently used in the assessment of gastric lesions. However, it is difficult to distinguish BCs from other cysts solely on the basis of imaging. On CT examination, BCs typically appear as unilocular, ovate lesions with well circumscribed smooth or lobulated borders. They are often described as homogenous hypoattenuating lesions without enhancement. Calcification is sometimes present in the cyst wall. These lesions can be misdiagnosed as solid masses such as GIST because they appear to be hyperattenuating due to the presence of hemorrhagic, thick mucinous, or proteinaceous contents [2]. These cystic lesions can also resemble solid masses with cystic degeneration. Degeneration seldom happens in the masses with the diameter less than 6 cm, but often appears in multilocular lesions or unilocular lesions with membranous septa.

Cystic degeneration is always patchy and causes an irregular thick wall with enhancement which is in clear contrast to the regular thin wall of BC. MRI provides far greater clarity than CT and is able to distinguish different contents by signal intensity. Due to the presence of methemoglobin, mucin, or protein the cysts usually show intermediate to high signal intensity on T1-weighted images and markedly high

signal intensity on T2-weighted images. Additionally, the signal is not inverted in the T1 weighted fat-suppressed image [7]. These image features are shared with other solid tumors with or without cystic degeneration, such as GIST and adrenal adenoma. In these cases, BC may masquerade a solid mass on CT and T1-weighted MRI. Consequently, imaging findings cannot be solely used to diagnose BC; instead histopathological data is needed for a definitive or confirmatory diagnosis.

Anatomical and pathological criteria for diagnosis include the presence of pseudostratified ciliated columnar epithelium together with at least one of the following: cartilage, smooth muscle or seromucous glands [8]. These features distinguish BC from esophageal duplication cysts which have two smooth muscle layers, as does the esophagus itself.

The bronchopulmonary sequestration is also lined by pseudostratified ciliated columnar epithelium, but there is mature and well-organized lung and bronchial tissue covered with pleural tissue. Cysts with ciliated epithelium lacking other distinguishing features, therefore, represent undifferentiated foregut cysts. In the past, confusion of anatomical and pathological criteria resulted in misclassification of BCs and lack of consensus toward suspected cases in the literature.

Surgical management and pathological examination are common choices. However, the indications for surgical intervention remain controversial. In general terms surgery is indicated when malignancy is suspected, or when there are symptoms and complications, such as infection, rupture, or compression of an adjacent organ. Although the exact malignant potential of these lesions is controversial, minimal but complete excision of the lesion is regarded as the proper surgical approach. Once surgical management has been decided, laparoscopy seems to be the standard technique undertaken by expert laparoscopic surgeons [9].

For asymptomatic lesions located near the EGJ, proximal subtotal gastrectomy performed only for the purpose of securing a pathological diagnosis is not a valid option. For cysts < 6 cm in diameter, BC should be considered in the differential diagnosis of gastric neoplasm, especially if the masses are located near the EGJ,

Gastric bronchogenic cyst presenting as GIST

and a considered therapeutic choice should be deliberated. CT and MRI should be performed to assess the morphological characters of the cyst and the signal characters of its contents.

EUS is often useful in identifying the location of lesions in the different layers of gastric wall. For the highly speculative lesions, EUS-FNA should be performed as an important supplementary method to secure a definitive diagnosis in some cases. The technique has a sensitivity of 93 to 95%, a diagnostic rate of 82 to 86% [10] and a low complication rate of 1 to 3% [11]. Complications include recurrence, ulceration, infection, and hemorrhage.

When assessing the location by EUS, endoscopic submucosal dissection (ESD) can be considered for submucosal cysts or for those located in more superficial layers. This avoids the unnecessary complications of surgery. However, ESD should not be considered for deeper, asymptomatic cases of BC, especially if the diameter is > 6 cm, because of the high risks of complications. Dynamic observation can also be a valid option for selected cases.

Although rarely located in the stomach, BC should be considered as a differential diagnosis of gastric neoplasm. For these cases, surgery remains a common choice. Minimally invasive procedures such as EUS-FNA and ESD should also be considered when the diagnosis of BC is suspected. They are valuable diagnostic methods that can assess and identify the location of the lesion, and facilitate histological examination of the cyst. In some cases of more superficial lesions ESD can take the place of surgery as it avoids unnecessary complications of a more invasive procedure.

Disclosure of conflict of interest

None.

Address correspondence to: Dr. Tong Liu, Department of General Surgery, Tianjin Medical University General Hospital, Tianjin 300052, China. Tel: +86-13102117641; Fax: +86-22-60362365; E-mail: doctorliutong@163.com

References

[1] Cuypers P, De Leyn P, Cappelle L, Veroustraete L, Demedts M, Deneffe G. Bronchogenic cysts: a review of 20 cases. *Eur J Cardiothorac Surg* 1996; 10: 393-6.

- [2] McAdams HP, Kirejczyk WM, Rosado-de-Christenson ML, Matsumoto S. Bronchogenic cyst: imaging features with clinical and histopathologic correlation. *Radiology* 2000; 217: 441-6.
- [3] Sanli A, Onen A, Ceylan E, Yilmaz E, Silistreli E, Acikel U. A case of a bronchogenic cyst in a rare location. *Ann Thorac Surg* 2004; 77: 1093-4.
- [4] Wang SE, Tsai YF, Su CH, Shyr YM, Lee RC, Tsai WC, Li FY, Chen TH, Wu CW, Lui WY. Retroperitoneal bronchogenic cyst mimicking pancreatic cystic lesion. *J Chin Med Assoc* 2006; 69: 538-42.
- [5] Okubo A, Sone S, Ogushi F, Ogura T, Uyama T, Monden Y, Li K. A case of bronchogenic cyst with high production of antigen CA 19-9. *Cancer* 1989; 63: 1994-7.
- [6] Tan KK, Nandini CL, Ho CK. A case of gastric bronchogenic cyst in Singapore with multiple intrigues. *ANZ J Surg* 2010; 80: 286-7.
- [7] Murakami R, Machida M, Kobayashi Y, Ogura J, Ichikawa T, Kumazaki T. Retroperitoneal bronchogenic cyst: CT and MR imaging. *Abdom Imaging* 2000; 25: 444-7.
- [8] Liang MK, Marks JL. Congenital bronchogenic cyst in the gastric mucosa. *J Clin Pathol* 2005; 58: 1344.
- [9] Ishizuka O, Misawa K, Nakazawa M, Nishizawa O. A retroperitoneal bronchogenic cyst: laparoscopic treatment. *Urol Int* 2004; 72: 269-70.
- [10] Matsui M, Goto H, Niwa Y, Arisawa T, Hirooka Y, Hayakawa T. Preliminary results of fine needle aspiration biopsy histology in upper gastrointestinal submucosal tumors. *Endoscopy* 1998; 30: 750-5.
- [11] O'Toole D, Palazzo L, Arotcarena R, Dancour A, Aubert A, Hammel P, Amaris J, Ruszniewski P. Assessment of complications of EUS-guided fine-needle aspiration. *Gastrointest Endosc* 2001; 53: 470-4.
- [12] Braffman B, Keller R, Gendal ES, Finkel SI. Subdiaphragmatic bronchogenic cyst with gastric communication. *Gastrointest Radiol* 1988; 13: 309-11.
- [13] Song SY, Noh JH, Lee SJ, Son HJ. Bronchogenic cyst of the stomach masquerading as benign stromal tumor. *Pathol Int* 2005; 55: 87-91.
- [14] Melo N, Pitman MB, Rattner DW. Bronchogenic cyst of the gastric fundus presenting as a gastrointestinal stromal tumor. *J Laparoendosc Adv Surg Tech A* 2005; 15: 163-5.
- [15] Lee SH, Park DH, Park JH, Kim HS, Park SH, Kim SJ, Oh MH. Endoscopic mucosal resection of a gastric bronchogenic cyst that was mimicking a solid tumor. *Endoscopy* 2006; 38 Suppl 2: E12-3.
- [16] Wakabayashi H, Okano K, Yamamoto N, Suzuki Y, Inoue H, Kadota K, Haba R. Laparoscopically resected foregut duplication cyst (bronchogen-

Gastric bronchogenic cyst presenting as GIST

- ic) of the stomach. *Dig Dis Sci* 2007; 52: 1767-70.
- [17] Shibahara H, Arai T, Yokoi S, Hayakawa S. Bronchogenic cyst of the stomach involved with gastric adenocarcinoma. *Clinical Journal of Gastroenterology* 2009; 2: 80-4.
- [18] Sato M, Irisawa A, Bhutani MS, Schnadig V, Takagi T, Shibukawa G, Wakatsuki T, Imamura H, Takahashi Y, Sato A, Hikichi T, Obara K, Hashimoto Y, Watanabe K, Ohira H. Gastric bronchogenic cyst diagnosed by endosonographically guided fine needle aspiration biopsy. *J Clin Ultrasound* 2008; 36: 237-9.
- [19] Jiang L, Jiang L, Cheng N, Yan L. Bronchogenic cyst of the gastric fundus in a young woman. *Dig Liver Dis* 2010; 42: 826.
- [20] Hosomura N, Kono H, Kawaida H, Amemiya H, Itakura J, Fujii H. A case of foregut gastric duplication cyst with pseudostratified columnar ciliated epithelium. *Clin J Gastroenterol* 2012; 5: 82-7.
- [21] Ubukata H, Satani T, Motohashi G, Konishi S, Goto Y, Watanabe Y, Nakada I, Tabuchi T. Intra-abdominal bronchogenic cyst with gastric attachment: report of a case. *Surg Today* 2011; 41: 1095-100.
- [22] Yang X, Guo K. Bronchogenic cyst of stomach: two cases report and review of the English literature. *Wien Klin Wochenschr* 2013; 125: 283-7.