

Adenocarcinoma arising from a gastric duplication cyst: a case report and literature review

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Introduction: Alimentary tract cystic duplication is a rare congenital anomaly predominantly affecting females, and diagnosed mostly in the early years of life.

Case report: We present here a case of a 51-year-old man presenting with a 2-day history of melena. Gastroscopy showed fresh blood, as well as a mass lesion and cavity at the fundus. Biopsies of the mass edge reported the occurrence of moderately differentiated adenocarcinoma and mild chronic gastritis. Computed tomography imaging showed a 5.1×6.5 cm cystic mass in the gastric greater curvature. The patient received six cycles of epirubicin, capecitabine, and oxaliplatin, followed by uncomplicated total gastrectomy.

Literature review: Data from all 11 reports of similar cases were reviewed and pooled. The result shows male predominance and variable symptoms, as well as a wide age range (25–76 years) at presentation. The cysts are commonly located along the greater curvature and are unilocular. Surgery was the treatment in most cases.

Conclusion: Alimentary tract cystic duplication is rare and may predispose to malignancy. Early diagnosis and prompt surgical intervention is important for the best outcome.

Keywords: gastric duplication cyst, gastric mass, adenocarcinoma, cystic duplication, alimentary tract cystic duplication

Introduction

Duplication of the alimentary tract is a rare congenital anomaly, with an incidence of 1 in 4,500 live births.¹ Two forms of duplication have been reported, namely, tubular and cystic. The tubular form communicates with the normal intestinal tract, while the cystic form is separate from it.^{1,2} Furthermore, most enteric duplication cysts are diagnosed during the first year of life, and about 70% of cases are identified by the age of 12 years.³ In 1961, Potter reported 2 cases in >9,000 fetal and neonatal autopsies.⁴ They may be slightly more common in males.⁵ The most common location is the ileum (35%), while the least common site is the stomach (2%–9%). Although very rare, carcinoma can arise from a gastric duplication cyst (GDC). Adenocarcinoma rarely arises from a GDC and, to our knowledge, only 11 cases (in the English language literature) have been reported to date (Table 1).^{6–16} Numerous other cases were reported in languages other than English.^{17–19} Nonetheless, there can be no argument that carcinomas arising from a GDC are especially rare. We are reporting, herein, a case of a 51-year-old man who, along with a duplication of the alimentary tract affecting the stomach, presented with melena, and upon further investigation, a well-differentiated adenocarcinoma was detected. In addition, we review the diagnosis and treatment of similar patients reported in the literature.

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Table 1 Summary of case studies reporting adenocarcinoma arising in GDC

References	Age (years), sex	Symptoms	Location	Size	Note
Yamasaki et al, 2016 ⁶	42, female	No	Adjacent to the greater curvature of stomach	10.0×9.5×8.0 cm unilocular cystic mass	Despite complete resection, the patient developed metastasis, underwent oral chemotherapy (TS-I), and died 2 months later.
Liu et al, 2014 ⁸	28, male	No	Adhered to gastric corpus	10×10 cm	7 months after surgery (in which the cyst ruptured), metastatic adenocarcinoma with peritoneal carcinomatosis was diagnosed.
Kang et al, 2014 ⁷	56, male	No	Along the greater curvature of the stomach	5.5×3.5×2.5 cm unilocular cyst	Patient underwent a wedge resection to remove the lesion. Adenocarcinoma showing invasion to the proper muscle layer of the cyst was observed.
Shin et al, 2014 ⁹	52, male	Abdominal pain	Peritoneum	4×3×3 cm unilocular cyst	Entire cyst was excised. Findings were consistent with an adenocarcinoma that had arisen from the intestinal duplication cyst.
Blank et al, 2012 ¹⁰	51, male	No	Ileal mesentery	4×10 cm	An en bloc resection was done. H–E staining revealed poorly differentiated invasive adenocarcinoma (intestinal type).
Zheng and Jing, 2012 ¹¹	25, male	No	The greater curvature of the stomach	Not found	Resection with total gastrectomy and regional lymphadenectomy were done. This is the youngest patient reported.
Jiang et al, 2011 ¹²	76, male	Incidentally found perigastric mass	Below the gastroesophageal junction	4×4 cm	Preoperative diagnosis was a GIST, and surgical excision was performed. The patient had an uneventful recovery.
Fukamoto et al, 2008 ¹³	50, male	Persistent vomiting due to pyloric stenosis	Mass on greater curvature of upper gastric corpus and mass adjacent to the bulbous	Masses of sizes 2 cm and 3 cm	Histological findings showed adenocarcinoma in the anal duplication cyst wall. A pancreatoduodenectomy was performed 9 days after initial surgery. Patient died 14 months later of local recurrence and multiple liver metastases.
Kuraoka et al, 2004 ¹⁴	40, male	Fever and back pain	Between stomach and spleen	7 cm in diameter and unilocular	Tubular adenocarcinoma was discovered, wherein the cyst had adhered 1 month after cystectomy.
Coit and Mies, 1992 ¹⁵	72, female	No	Adjacent to greater curvature of stomach	3.2 cm in diameter	Adenocarcinoma believed to have arisen in the GDC was identified, and the patient underwent a radical distal subtotal gastrectomy with Billroth I reconstruction.
Mayo et al, 1955 ¹⁶	64, female	Weakness, anorexia, and malaise	Distal end of antrum of the stomach	6 cm in diameter	Carcinoma had developed, but patient recovered after radical subtotal gastric resection.

Abbreviations: GDC, gastric duplication cyst; GIST, gastrointestinal stromal tumor; H–E, hematoxylin and eosin; TS, titanium silicate; yrs, years.

Case report

A 51-year-old man presented to the Accident and Emergency Department of Salmaniya Medical Complex with a 2-day history of melena. There was no past medical history of note, no alcohol intake, and he was not on any medications. The patient stopped smoking 10 years prior to admission. There was a first-degree family history of colorectal cancer.

Upon investigation, a gastroscopy revealed fresh blood and clots in the fundus of the stomach. A mass lesion and cavity were seen along the greater curvature of the stomach

during retroflexed examination of the fundus. The cavity contained an ulcerated area with clots (Figure 1). Biopsies of the mass in the cavity showed normal gastric mucosa with moderately differentiated adenocarcinoma and mild chronic gastritis (Figure 2). Computed tomography (CT) with contrast of the chest, abdomen, and pelvis demonstrated a 5.1×6.5 cm cystic mass in the greater curvature of the stomach (Figure 3). The cyst had a 6 mm thickened wall, with a septum dividing it into 2 parts. Hematoxylin and eosin (H–E) staining revealed a cyst lined by benign epithelial lining (Figure 4).

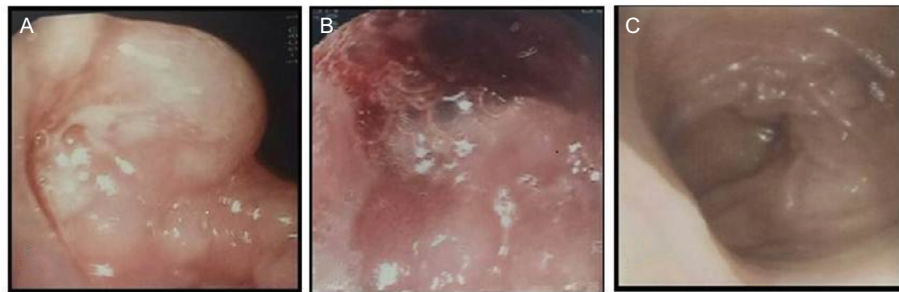


Figure 1 Representative endoscopic images.

Notes: (A) Image depicts a mass lesion with cavity opening observed in the fundus. (B) The cavity contained marginal ulcers with fresh and clotted blood. (C) An image of normal antrum.

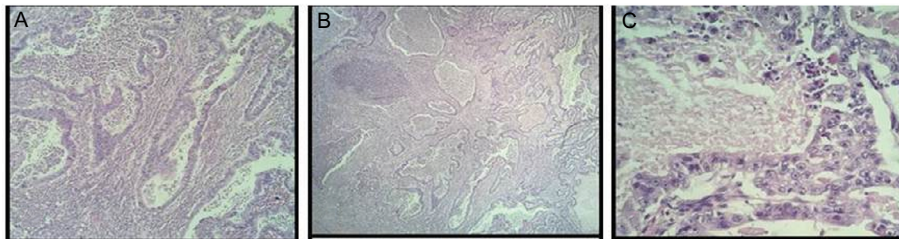


Figure 2 Histology images of moderately differentiated adenocarcinoma.

Notes: H-E staining depicts invasive atypical glands with central necrosis in (A) (original magnification $\times 100$) and (B) (original magnification $\times 40$). (C) Invasive malignant glands lined by atypical pleomorphic cells with prominent nucleoli and central necrosis can be seen (original magnification $\times 400$).

Abbreviation: H-E, hematoxylin and eosin.

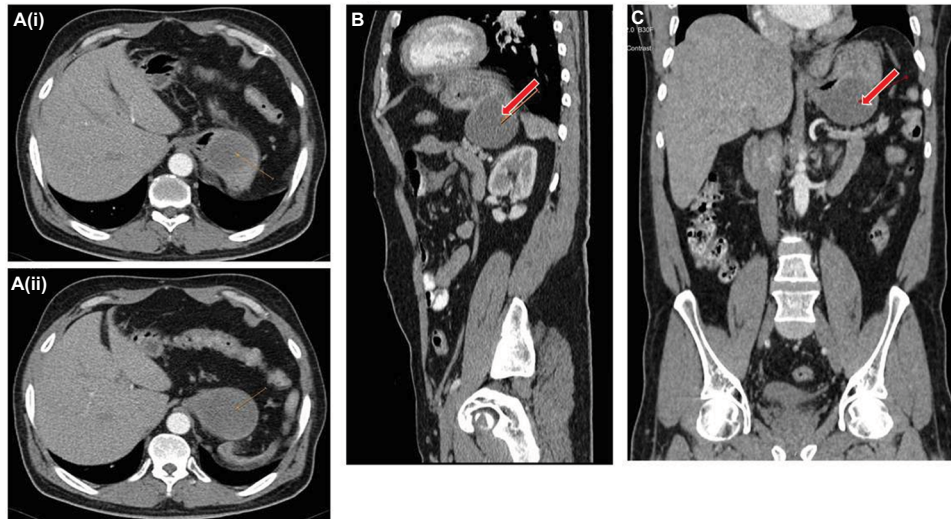


Figure 3 CT scan of the duplication cyst.

Notes: (A(i) and A(ii)) Axial, (B) sagittal, and (C) coronal scans of the chest, abdomen, and pelvis demonstrated a well-circumscribed, round cystic mass of homogeneous density along the greater curvature of the stomach (arrows), consistent with a duplication cyst. The mass measured 5×4 cm.

Abbreviation: CT, computed tomography.

These findings fulfilled the diagnostic criteria of GDC. No metastases or other abnormalities were seen (Figure 3).

The patient declined immediate surgical resection but agreed to neoadjuvant chemotherapy followed by surgery. Six cycles of epirubicin, capecitabine, and oxaliplatin were

administered over 6 months without complication. The patient achieved a partial response to chemotherapy and subsequently underwent an uncomplicated total gastrectomy. The specimen contained the full stomach with omentum and measured $18.5 \times 5 \times 4$ cm. A fungating mass measuring

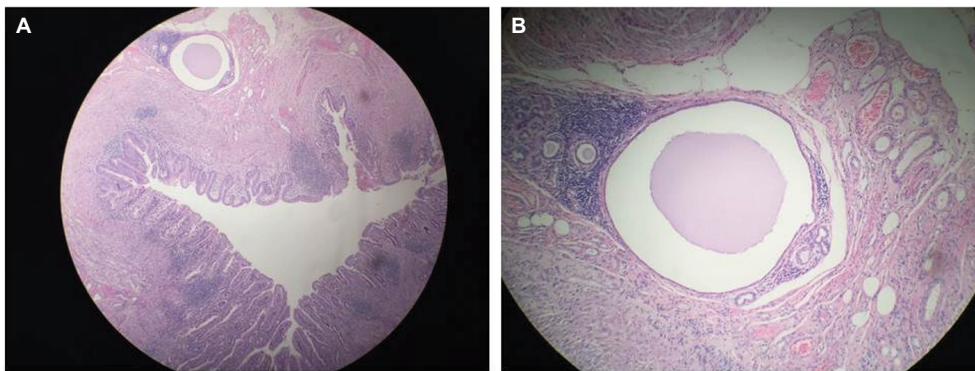


Figure 4 Histology of cyst lined by benign epithelial lining.

Note: H–E staining shows cyst lined by benign epithelial lining, surrounded by smooth muscle fibers and chronic inflammatory cells: **(A)** (original magnification $\times 10$); and **(B)** higher magnification (original magnification $\times 100$).

Abbreviation: H–E, hematoxylin and eosin.

5 \times 2.5 cm was present in the fundus. The resection margins ranged from 2 to 13 cm. Multiple lymph nodes of <1 cm were present in the perigastric fat. Microscopic examination showed a well-differentiated adenocarcinoma with ypT2 invasion of the muscularis propria. The margins were clear of cancer. The 7th edition of the American Joint Committee on Cancer (AJCC) *Tumor-Node-Metastasis (TNM) Staging Manual* for the stomach was used, and the final TNM stage was ypT2 N0 Mx. Written informed consent was provided by the patient to have the case details and any accompanying images published.

Literature review

As shown in Table 1, data from all the 11 reports of similar cases collected from the English language literature were reviewed and pooled. It shows male predominance (8 of 11 cases were males), variable symptoms (from incidental finding to no symptoms, to some more remarkable symptoms), and a wide age range (from 25 to 76 years) at presentation. The cysts are of variable sizes (from 2.5 cm to 10 cm in diameter), commonly located along the greater curvature of the stomach, and unilocular. Surgery was the rule in most cases.

Discussion

GDCs comprise about 2%–9% of all alimentary tract duplications, and cystic types with no connection to the gastric lumen comprise up to 80% of these lesions.³ There is a higher male incidence of GDCs complicated with malignancy, as in our reported case and that of others (Table 1), which shows only 3 females in the pooled 11 cases.^{6–16} This goes with the higher male predominance of gastric cancer in most literature. Most gastric duplications are single in nature, although multiple duplications have been reported.⁶ GDCs are true cysts. They typically have a mucosal lining, consisting of gastric

epithelium, which is surrounded by a muscularis propria. Mucosal linings consisting of ectopic pancreatic tissue or respiratory epithelium are less commonly seen.²⁰ GDCs are typically found in the greater curvature of the stomach.

The pathogenesis for alimentary duplication cysts is a matter of controversy. It is, however, regarded as a congenital condition, and the most plausible theory consists of an aberration in the recanalization and fusion of longitudinal folds. Kim et al proposed that duplication cysts originated from the fusion of longitudinal folds, allowing the passage of a bridge of submucosa and muscle at the second and third months of intrauterine life.²¹ Other theories include the development of traction diverticulum, which leads to duplication cyst formation as a result of notochord adhesion and therefore a lag of the elongation process of the embryonic endoderm relative to its surrounding structures. Other potential mechanisms proposed include abortive twinning, anoxia, and persistent embryological diverticula.²¹

Pathologic conditions associated with alimentary tract duplication cysts include pulmonary sequestration, multicystic dysplastic kidney, and distant neoplasia.²² Malignancy arising due to GDC is also an important differential diagnosis. Adenocarcinoma is currently considered the most common histologic type of malignancy arising in GDC cases; however, neuroendocrine carcinoma and squamous cell carcinoma have also been reported.⁷ Table 1 represents an overview of case studies reporting adenocarcinoma arising in GDC. The risk of malignant transformation and associated complications has led to surgical excision being recommended as the standard of care for this condition.²³ Open and laparoscopic excisions have been reported. Some patients have a duplication cyst that has a wall that is shared with the functional bowel. Treatment of these patients generally includes excision of only the duplicated area.²

Table 2 Common clinical findings in patients with duplication cysts

Adults	Children
Asymptomatic; vague or acute abdominal pain	Asymptomatic; an intra-abdominal mass*
An intra-abdominal mass*	Frank or occult bleeding
Chronic abdominal pain	Abdominal distention
Symptoms of acute diverticulitis	Constipation
Frank or occult bleeding	Vomiting
At autopsy	Failure to thrive
During laparotomy	At autopsy
Symptoms of acute pancreatitis or a pancreatic pseudocyst mimic**	During laparotomy

Notes: These data have been collected from literature. *Due to compression of adjacent organs, such as pancreas, kidney, spleen, and adrenal gland. **As up to 10% of GDCs may contain ectopic pancreatic tissue.

Abbreviation: GDC, gastric duplication cyst.

Table 3 The essential criteria for diagnosis of a gastric duplication cyst

The wall of the cyst is contiguous with the stomach wall
The cyst is surrounded by smooth muscle, which is continuous with the muscle of the stomach wall
The cyst wall is lined by epithelium consisting of gastric or any other type of gut mucosa

Note: Data collected from literature, especially from studies Singh et al,¹ Hsu et al,³ and Kuraoka et al.¹⁴

GDCs can grow in size, leading to compression of adjacent organs, including the pancreas, kidney, spleen, or adrenal gland.²⁴ Alimentary tract cysts are at risk for obstruction, infection, torsion, perforation, hemorrhage, and malignancy.¹ Indeed, this case presented with melena (overt bleeding), which is secondary to ulcerated cancer. The possibility that this case is that of a GDC and gastric cancer arising from the gastric lumen, and not from the epithelium of the GDC, exists, and only a macroscopic image of the GDC and cancer is strongly required to prove that. Unfortunately, this image was missed in this case. Patients' symptoms at presentation are related to the pathology involved in the duplication and the age at presentation (Table 2).

The diagnosis of GDC is based on established criteria (Table 3). The cyst has to be intimately connected to the gastric wall and surrounded by one or more muscle layers, which are adherent to the muscle coats of the stomach. As part of the criteria, the epithelium lining is of gastric mucosa, though at times, other gut mucosa may also be found such as heterotopic intestinal or pancreatic tissues.²²

Radiologically, GDCs appear classically as thick-walled cystic lesions with inner lining enhancement and occasional calcifications on both CT and magnetic resonance imaging (MRI).²³ The MRI can further differentiate the type of the cyst and characterize the cystic contents.²² The nature of the fluid

in the cyst can vary with the presence of bleeding, chronic inflammation, or infection.

Endoscopic examination commonly shows a gastric mass. The overlying mucosa is often intact but, not uncommonly, can appear erythematous with or without ulceration.²⁴ Biopsy of the lesion is essential, and the typical changes include the following: 1) mucous membrane with a well-organized layer of smooth muscle, 2) granulomatous inflammation with denuded mucosa, and 3) foreign body giant cells.³ Endoscopic ultrasonography (EUS) can be a useful tool for diagnosing GDCs. The classic appearance is of an echogenic internal mucosal layer and a hypoechoic intermediate muscular layer. However, the yield of EUS-guided fine needle aspiration (FNA) remains uncertain as the findings are frequently indistinguishable from those of mucinous pancreatic neoplasms.²³

Alimentary tract duplications are best managed by surgical resection. The conservative approach may be considered for patients with communicating GDCs and in whom both gastric lumens were patent. The use of drainage and marsupialization of the cyst has been discouraged as the unprotected mucosa of the cyst will be exposed to a greater volume of gastric contents, placing it at higher risk of ulceration. Complications such as anastomotic strictures and blind loop syndrome have been frequently reported following drainage procedures such as cystojejunostomy.⁸

Conclusion

GDC is a rare condition that may pose a diagnostic challenge. GDC may predispose to complications, including malignancy, as in our case. GDC should be considered when a cystic lesion is found adjoining the gastric wall. Early diagnosis and prompt surgical intervention is important for the best outcome.

Disclosure

The authors report no conflicts of interest in this work.

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