

# Sarcomatoid Carcinoma Arising in a Gastric Duplication Cyst

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## ABSTRACT

Malignancy arising within a gastric duplication cyst (GDC) is extremely rare; only 15 cases have been reported in the literature. We present a 70-year-old woman who was referred with a history of vague postprandial abdominal discomfort. Subsequent imaging identified a gastric cystic mass. A laparoscopic sleeve gastrectomy of a 90 × 60 × 60-mm cystic mass was performed. Histopathological examination showed the presence of a sarcomatoid carcinoma arising within a GDC. The patient, unfortunately, died 5 months after surgery with metastatic disease. To the best of our knowledge, this is the first case of sarcomatoid carcinoma arising within a GDC.

## INTRODUCTION

Duplication cysts are rare congenital anomalies that typically occur in children in the first year of life, and 150 cases have been reported in the medical literature since 1911.<sup>1-5</sup> This diagnosis is incidental in most adults.<sup>1</sup> Malignancy arising within a gastric duplication cyst (GDC) is extremely rare, with only 15 cases reported in the literature and predominantly adenocarcinoma.<sup>6</sup> We report a case of sarcomatoid carcinoma arising within a GDC.

## CASE REPORT

A 69-year-old woman was referred with a history of vague postprandial abdominal discomfort for a 1-year duration. She denied any other symptoms. Medical history revealed hypertensive obstructive cardiomyopathy and irritable bowel syndrome. On examination, she was overweight but not anemic, and there was no clinical evidence of jaundice. A small hernia was seen in the epigastric region. Investigations revealed a hemoglobin of 12 g/dL; liver function tests were normal. Endoscopic examination revealed a hypochoic cystic lesion with a heterogenous internal echotexture and regular margins.

Computed tomography (CT) scan confirmed the hernia and incidentally showed a cyst, 75 mm in diameter arising from the greater curvature of the stomach abutting the pancreas (Figure 1). The differential diagnoses included either a GDC or a gastrointestinal stromal tumor (GIST) with significant cystic degeneration. The liver was normal in size in an ultrasound scan. Laparoscopic sleeve gastrectomy with en bloc resection of the adherent transverse colonic mesentery was performed. The immediate postoperative period was uneventful. On macroscopic examination, this was a 90 × 60 × 60-mm cystic mass with intact serosa, and an overlying slightly raised intact gastric mucosa was noted. On opening, the cyst was filled with brown muddy material along with intramural solid nodular areas (Figure 2). The histological examination revealed a cyst wall partially lined by mucinous epithelium and partly by primitive squamous epithelium merging with a columnar epithelium reminiscent of gastric antral mucosa, in keeping with a GDC (Figure 2). Lymphoid follicles were noted in the lamina propria. The cyst wall in the region of the mural nodule was infiltrated by a



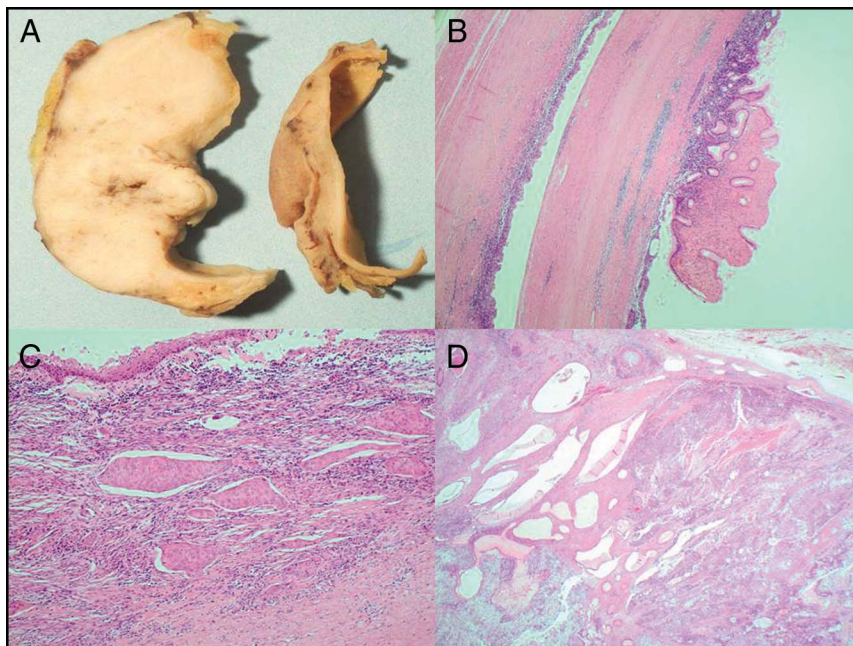
**Figure 1.** Axial view of the computed tomography scan showing large cystic lesion in the supraumbilical region, likely arising from the stomach, and abutting the pancreas.

tumor composed of islands of atypical squamous epithelial cells with keratin pearls admixed with areas of glandular differentiation on a background of malignant stroma comprising highly pleomorphic discohesive cells with numerous mitotic figures (Figure 2).

The epithelial component was positive for AE1/AE3, CAM 5.2, and CK 7. A patchy positivity for AE1/AE3 was also present in the stromal component. There was no expression of desmin, CK20, DOG-1, S-100, ER, myogenin, CDX2, PAX8, and TTF-1 within the tumor. Overall, the morphological appearances were those of a sarcomatoid carcinoma arising within a GDC. There was focal infiltration into the adherent mesocolic fat; however, the serosa of the cyst wall was intact. The overlying gastric mucosa was free of atrophy, intestinal metaplasia, dysplasia, or malignancy. Three months postoperatively, multiple liver metastases were identified on the CT scan. The patient received a single cycle of palliative chemotherapy including an anthracycline, epirubicin, carboplatin, and capecitabine but clinically deteriorated rapidly with disease-related symptoms and worsening liver function tests secondary to disease progression precluding further treatment. The patient died of disease 5 months after the surgery.

## DISCUSSION

Gastrointestinal duplication cyst is a rare congenital abnormality, and GDC is even rarer, comprising only 2%–8% of all gastrointestinal duplications.<sup>6–8</sup> The pathogenesis remains poorly defined; the most widely accepted theory is abnormal recanalization after the solid epithelial stage of embryonic bowel development.<sup>2</sup> The duplication cyst is surrounded by at least 1 coat of smooth muscle, usually fusing the muscularis propria of the cyst and the stomach.<sup>2,6</sup> The cyst is lined by typical gastric mucosa, often accompanied by patches of ectopic



**Figure 2.** (A) Macroscopic examination of the gastric duplication cyst wall showing thick intramural solid tumor nodule, (B) photomicrograph of gastric duplication cyst showing a columnar epithelium reminiscent of gastric antral mucosa with dense lymphoid infiltrate in the lamina propria (hematoxylin and eosin stain, 40× magnification), (C) the cyst wall was infiltrated by islands of atypical squamous epithelial cells (hematoxylin and eosin stain, 100× magnification), and (D) the cyst wall showing areas of tumor with glandular differentiation and malignant stroma (hematoxylin and eosin stain, 20× magnification).

intestinal, colonic, respiratory epithelium, or pancreatic tissue.<sup>8</sup> Patients usually present with epigastric pain, vomiting and anemia, and a palpable abdominal mass on physical examination. A small subset of patients remains asymptomatic. Hemorrhage, perforation, complete gastric outlet obstruction, ulceration, and fistula formation are known complications based on location or type of ectopic tissue present.<sup>3,4,8</sup>

Malignancies within GDCs are extremely rare and generally occur in middle-aged adults (age range: 25–71 years; mean: 54 years).<sup>9</sup> The main symptoms are abdominal pain, weight loss, and vomiting. The reported size of duplication cysts ranges from 32 to 170 mm (mean: 89 mm).<sup>9</sup> Similar findings were noted in the current case study. Preoperative diagnosis is extremely difficult, and the differential diagnoses based on radiologic or endoscopic findings are usually GIST, neuroendocrine tumor, neurogenic tumor, or heterotopic pancreas.<sup>8</sup> At present, CT scan remains the best imaging modality to diagnose or at least suggest GDC with malignancy.<sup>1</sup> In this case study, the differentials included GIST and GDC.

Malignant transformation of GDC into adenocarcinoma, squamous cell carcinoma, mixed adenocarcinoma and squamous cell carcinoma, GIST, and neuroendocrine carcinoma has been reported in the literature.<sup>6,8–10</sup> Adenocarcinoma is the most common histologic type of malignant change associated with GDC.<sup>2</sup> Therefore, it is important to differentiate between adenocarcinoma arising from the gastric mucosa and those arising from a GDC. In our case, the overlying gastric mucosa did not show evidence of in situ malignancy, dysplasia, atrophic gastritis, or intestinal metaplasia. However, the GDC showed foci of squamous lining epithelium with underlying islands of infiltrating atypical squamous epithelium and background sarcomatoid transformation. These findings suggest that the carcinoma arose from the GDC and invaded the gastric wall. Sarcomatoid transformation has not been described previously in the literature.

The prognosis of cases with malignant complications of GDC is poor, according to previous reports.<sup>1,9</sup> Metastasis may be seen either at presentation or later on follow-up imaging.<sup>1</sup> The first choice of therapy has always been surgical resection for GDC with malignancies.<sup>1,10</sup> In this case, liver metastases were identified on imaging 4 months after surgery. Although sarcomatoid carcinoma is relatively insensitive to chemotherapeutic drugs, certain patients in various previous studies have demonstrated benefit from chemotherapy.<sup>11,12</sup> However, whether or not to treat gastric sarcomatoid carcinoma with chemotherapy remains a controversial issue in the absence of compelling data regarding efficacy.<sup>11</sup> Our patient commenced platinum-based chemotherapy including an anthracycline, epirubicin, carboplatin, and capecitabine, which was stopped prematurely due to futility. The patient died of disease 5 months postoperatively. In

summary, malignant transformation within GDC is extremely rare, difficult to diagnose preoperatively, and the overall prognosis is usually dismal. Surgical resection still remains the primary modality of treatment. To the best of our knowledge, this is the first case of sarcomatoid carcinoma arising from a GDC.

## DISCLOSURES

Author contributions: MAH Ahmed and KS Liyanaarachchi wrote the manuscript and approved the final manuscript. SR Preston and M. Hewish revised the manuscript for intellectual content and approved the final manuscript. IN Bagwan approved the final manuscript and is the article guarantor.

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Informed consent was obtained for this case report.

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## REFERENCES

- Sethi S, Godhi S, Puri SK. Papillary adenocarcinoma in a gastric duplication cyst. *Indian J Surg Oncol* 2018;9(1):79–82.
- Chan BPH, Hycza M, Ramsay J, et al. Adenocarcinoma arising from a gastric duplication cyst. *ACG Case Rep J* 2018;5:e42.
- Doepker MP, Ahmad SA. Gastric duplication cyst: A rare entity. *J Surg Case Rep* 2016;5:1–3.
- Scatizzi M, Calistri M, Feroci F, et al. Gastric duplication cyst in an adult: Case report. *In Vivo* 2005;19(6):975–8.
- Perek A, Perek S, Kapan M, et al. Gastric duplication cyst. *Dig Surg* 2000;17:634–6.
- Lee S, Uno K, Fujishima F, et al. Gastric duplication cyst with occult GIST component. *ACG Case Rep J* 2019;7:e00260.
- Kuraoka K, Nakayama H, Kagawa T, et al. Adenocarcinoma arising from a gastric duplication cyst with invasion to the stomach: A case report with literature review. *J Clin Pathol* 2004;57(4):428–31.
- Kang HJ, Jang SJ, Park YS. Adenocarcinoma arising in gastric duplication cyst. *Korean J Pathol* 2014;48(2):159–61.
- Zheng J, Jing H. Adenocarcinoma arising from a gastric duplication cyst. *Surg Oncol* 2012;21(2):e97–101.
- Barussaud ML, Meurette G, Cassagnau E. Mixed adenocarcinoma and squamous cell carcinoma arising in a gastric duplication cyst. *Gastroenterol Clin Biol* 2008;32(2):188–91.
- Lee SE, Park SY. Sarcomatoid carcinoma of the small intestine: A rare and highly aggressive tumor. *J Korean Surg Soc* 2012;83(5):321–4.
- Zhu CC, Li MR, Lin TL, et al. Sarcomatoid carcinoma of the stomach: A case report and literature review. *Oncol Lett* 2015;10(3):1385–9.

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