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CASE REPORT | STOMACH

Gastrointestinal Stromal Tumor Arising From a Gastric Duplication Cyst

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Abstract

Gastric duplication cysts (GDC) are rarely diagnosed in adults, but previous cases have been associated with malignancy. We present a case of gastrointestinal stromal tumor (GIST) arising from a GDC in a 71-year-old woman who presented with 3 years of early satiety, anorexia, abdominal distention, and weight loss. Abdominal CT showed a 9.3 x 5.2 x 9.5-cm well-circumscribed cystic mass arising 3 cm above the gastroduodenal junction. The cyst was resected, and histopathology was consistent with GDC. Future studies are needed to clarify the malignant potential of GDC and the molecular pathways for its development.

Introduction

Gastric duplication cysts (GDC) belong to a group of congenital abnormalities called gastrointestinal duplication cysts, which can be found at any portion of the alimentary tract. GDCs are located in or adjacent to the wall of the gastrointestinal tract, have smooth muscle components, and are lined by alimentary tract mucosa. They are rarely diagnosed in adults. Malignant transformation of GDC is rare, and only 9 previous cases have been described in the English literature, with adenocarcinoma being the most common.[1] Gastrointestinal stromal tumors (GIST) are the most frequent mesenchymal tumors found in the gastrointestinal tract, but have not been previously associated with GDC.[2,3]

Case Report

A 71-year-old woman presented with a 3-year history of early satiety, anorexia, and abdominal distention, as well as progressive weight loss of 10 kg. Physical examination revealed a non-tender, 5-cm epigastric mass, but was otherwise normal, as was her past medical history. Laboratory tests including blood cell counts, biochemistry, hepatobiliary tests, and pancreatic enzymes were all within normal range, except for mild hypoalbuminemia (3.2 g/dL). Tumor markers, including CA 19-9 and CEA, were negative.

Abdominal computed tomography (CT) revealed a 9.3 x 5.2 x 9.5-cm, well-circumscribed cystic mass with thickened walls arising 3 cm above the gastroduodenal junction (Figure 1). No enlarged lymph nodes were observed. Upper endoscopy revealed a 5-cm mass protruding into the gastric antrum covered by intact mucosa and draining whitish fluid. A biopsy from the mucosa revealed chronic gastritis, and stains for *Helicobacter pylori* were negative.

The patient underwent exploratory laparotomy. A saccular mass with a nodular external layer was found attached to the anterior wall of the stomach, 3.5 cm above the pylorus and shared vascular supply with the greater gastric curvature. The mass was resected with a margin of macroscopically normal stomach. Gross examination revealed

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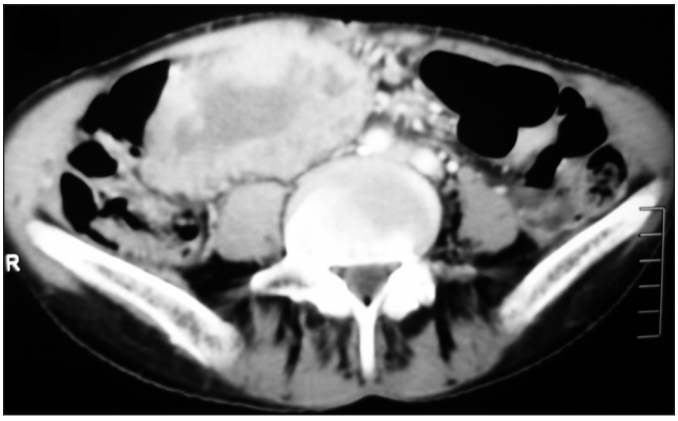

Figure 1. Axial abdominal CT showing a cystic mass with thickened, heterogenous walls.

a 12 x 10 x 4-cm cyst with nodular, thick (2 cm), congestive walls coalesced with normal stomach. Histopathology showed gastric inner mucosal lining, a submucosal layer, a thickened muscularis propia, and an outer smooth muscle layer (Figure 2). A coat of shared smooth muscle surrounded the cyst. There was an area of uniform, densely packed, diffuse sheets of atypical spindle-shaped cells with diffusely cytoplasmatic reaction to CD 117 (Figure 3). All features were consistent with GDC. Immunohistochemistry was negative for desmin and S-100. These histopathologic features were consistent with GIST arising from a GDC. The patient remained asymptomatic 2 years after surgery, without additional therapy or disease recurrence.

Discussion

Gastrointestinal duplication is a rare congenital abnormality that can present in any portion of the gastrointestinal tract.[4,5] The most commonly affected structures are ileum (35%), esophagus (19%), jejunum (10%), stomach (9%), and colon (7%).[1] About 150 cases of GDC have been reported in the medical literature since 1911.[6] It is typically found in children, and only 20% of patients are diagnosed after 12 years of age.[5] Associated congenital anomalies such as concurrent duplications are found in 50% of cases.[7] Among the patients with GDC, 95% arise from the greater curvature.[8,9]

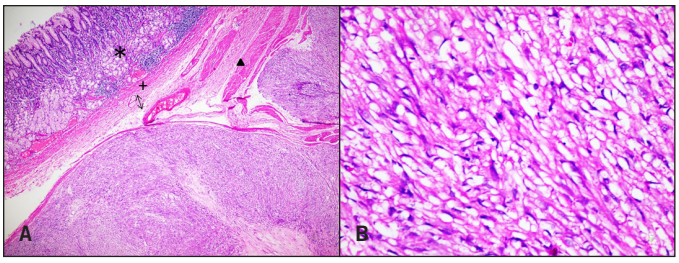

Figure 2. Histopathologic findings of GDC biopsy. (A) The cyst covering was comprised of gastric mucosa (*), muscularis mucosa (+), submucosa (arrow), and muscularis externa (triangle; H&E stain, x40) (B) Diffuse spindle shaped cells (H&E stain, x100).

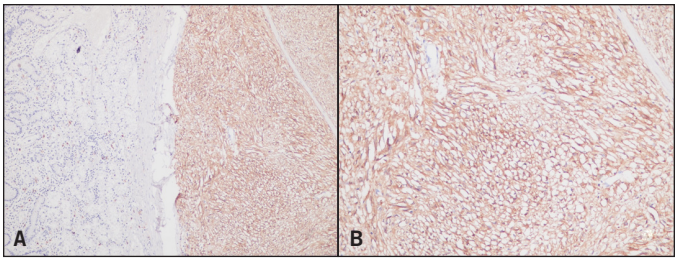

Figure 3. (A) Immunohistochemical staining showing positive reaction for CD117, sparing the gastric mucosa (x40) (B) Immunohistochemical staining shows positive reaction for CD117 in the cytoplasmic membrane (x100).

The essential features of GDC were described in 1959, and include: 1) continuity of the cyst wall within the stomach, 2) a coat of shared smooth muscle surrounding the cyst, 3) a common blood supply between the cyst and the stomach, and 4) alimentary tract epithelium lining.[10] Common symptoms of GDC include abdominal pain, nausea, vomiting, weight loss, bloating, fever, or epigastric mass.[11,12] Complications such as obstruction, bleeding, fistulization, pancreatitis, acute abdomen, and malignant transformation have been described.[7,9]

CT is useful as an initial diagnostic test for detection of GDC, as it is readily available and non-invasive.[13] CT typically reveals fluid-attenuation cystic mass in close contact with the stomach. However, these lesions may be confused with solid masses due to the high-protein content of the cyst. Ultrasound has not been evaluated as a diagnostic tool for GDC. Upper endoscopy may reveal a gastric mass with intact, erythematous or ulcerated overlying mucosa.[8] Endoscopic ultrasound (EUS) may help to differentiate between the solid and cystic component and the relation between the cyst wall and adjacent gastrointestinal structures.[14,15] EUS-guided fine-needle aspiration has been proposed as a valuable tool for the diagnosis and characterization of the cyst, and can be therapeutic.[16]

GDC may mimic pancreatic pseudocyst in imaging studies. However, pseudocysts are usually related to recent episodes of pancreatitis, which may be associated with elevation of lipase or CA 19-9, and walls are microscopically different.[13,14,17] Other differential diagnoses include GIST, omental cysts, mesenteric cysts, choledochal cysts, and ovarian cysts.[15] About 70% of GIST occur in the stomach and may contain areas of cyst formation. Histopathology obtained by cyst biopsy or surgical resection is of paramount importance to differentiate between GDC and cystic GIST.

Malignant transformation of GDC is uncommon.[7,9,18] Only 9 cases of GDC with malignant transformation were found in

the literature. The histopathological findings were adenocarcinoma (6 patients), followed by neuroendocrine carcinoma, epithelial malignancy, and mixed adenocarcinoma with squamous cell carcinoma. Most patients had no recurrence of disease in at least 12 months of follow-up, except for 3 patients who developed progression of the disease and metastasis after surgery. GIST has been previously described arising from ileal and colonic duplication cysts, but, to our knowledge, this is the first case reporting a GIST arising from a GDC.[2,4]

The recommended treatment for symptomatic patients with GDC is surgical resection.[1,11,19,20] Complete cystectomy is the best surgical option and alternatively segmental gastrectomy can be performed. The management of asymptomatic patients with GDC is controversial.[21] As malignant transformation has been reported in patients with GDC, different experts recommend prophylactic surgical resection.[1] As the association with malignancies has not been completely elucidated, some authors advocate for a more conservative approach with close clinical and imaging follow-up.[22] Further studies of the role of GDC to predispose for malignancies are needed to clarify what is the best therapeutic approach in asymptomatic patients. As the diagnosis of GDC is barely made solely on imaging findings, surgery plays a major role to clarify the diagnosis in most patients. Neodajuvant or adjuvant therapy has not been evaluated.[23] It is not clear how long patients with GDC with concomitant malignancy should be followed to reassure the absence of recurrence.

Disclosures

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