An Adult Gastric Duplication Cyst Mimicking a Gastrointestinal Stromal Tumor

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Abstract

We herein describe a rare case of a 24-year-old man who presented with severe epigastralgia after consuming a considerable amount of broiled meat. Computed tomography revealed a cystic lesion adjacent to the distal stomach, with high intensity on T2-weighted magnetic resonance imaging. Upper endoscopy showed a cystic mass measuring 6 cm in diameter, mimicking a submucosal tumor adjacent to the pyloric valve, with duodenum invagination, characteristic of ball valve syndrome. Endoscopic ultrasonography showed that the lesion was contiguous through the first to the third layer of the stomach. Therefore, we performed distal gastrectomy. Pathology showed that the lesion was a gastric duplication cyst without malignancy.

Key words: gastric duplication cyst, upper endoscopy, ball valve syndrome

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Introduction

A gastric duplication cyst (GDC) is a congenital anomaly that is rarely seen in adults (1). As it is typically diagnosed during childhood with various symptoms such as vomiting, abdominal pain, or weight loss, >70% of reported cases have been found in patients <12 years of age (2). As GDCs are usually asymptomatic in adult patients, they are incidentally discovered through ultrasonography, computed tomography (CT), or upper endoscopy (3); however, GDCs are not widely recognized by endoscopists due to their extreme rarity. Although it is difficult to diagnose GDCs preoperatively, recent imaging modalities have provided some informative findings. CT and endoscopic ultrasonography (EUS) are of diagnostic significance for GDCs. When EUS demonstrates a cyst with an echogenic internal mucosal layer and a hypoechoic muscular layer, the diagnosis of a GDC is more likely. However, in some instances, mucinous cystic tumors of the pancreas also show similar radiological features, and GDCs adjoining the pancreas are indistinguishable. Furthermore, the reported radiographic and endosonographic findings for a definitive preoperative diagnosis leave some ambiguity. Comprehensively, the definitive diagnosis of GDC is difficult, despite the use of diagnostic modalities including endoscopy. Complete resection is the only method to definitively diagnose GDCs. We herein report a rare case of adult symptomatic GDC mimicking a submucosal tumor (SMT), such as a gastrointestinal stromal tumor (GIST), which presented with outlet obstruction due to ball valve syndrome (BVS).

Case Report

A 24-year-old man with no history of disease, who presented to our emergency department with acute severe epigastralgia a few hours after consuming a considerable amount of broiled meat, was admitted to the gastroenterology department of our institution. A physical examination revealed severe epigastric tenderness without any peritoneal signs. His facial appearance expressed agony and he presented with excessive sweating. His blood pressure was 96/40 mmHg, and his pulse rate was elevated at 95 beats/min. All hematological parameters were within normal ranges, including the serum levels of carcinoembryonic antigen and CA19-9. He received conservative therapy with fasting and nasogastric intubation. CT revealed a cystic lesion around the distal stomach (Fig. 1A). The lesion showed high intensity on the coronal view on T2-weighted magnetic resonance imaging (MRI) (Fig. 1B), and upper endoscopy displayed a
Figure 1. Computed tomography showing the cystic lesion around the distal stomach (A). The lesion shows high intensity on a T2-weighted coronal slice magnetic resonance image (B).

Figure 2. Upper endoscopy shows a giant cystic mass, approximately 6 cm in diameter, mimicking a submucosal tumor (such as a gastrointestinal stromal tumor) adjacent to the pyloric valve.

cystic mass mimicking a SMT of the stomach (Fig. 2). Moreover, outlet obstruction of the stomach due to duodenal invagination of the cystic lesion was noted, referred to as BVS. The vomiting reflex released the lesion from the duodenum, and obstructive symptoms quickly disappeared the day after endoscopy. EUS, performed for further evaluation, demonstrated that the cystic lesion was three-layered, hypoechoic, and contiguous with the stomach through the first to the third layer of the proper stomach (Fig. 3A). A cytological examination using EUS-guided fine-needle aspiration (FNA), performed for the definitive diagnosis, indicated class IIIb cytology, which could be suggestive of malignancy. Differential diagnoses included GIST, carcinoma of the stomach, heterotopic gastric gland, or an extragastric lesion such as pancreatic cystic lesion or lymphatic cyst of the greater omentum; however, we could not establish a definitive preoperative diagnosis. As the patient desired symptom relief, we decided to perform distal gastrectomy for the therapeutic diagnosis. The resected specimen is shown in Fig. 3B. A pathological examination demonstrated that the inner side of the cystic lesion comprised a single layer of columnar epithelium with glandular formation. In addition, the lesion shared the layer of the mucosa, muscularis mucosa, and submucosa of the proper stomach (Fig. 4). The patient has been well since hospital discharge, and the postoperative radiological work-up and upper endoscopy showed no evidence of recurrence during the 3-year follow-up period.

Discussion

Rowling et al. described the following essential features of GDC: 1) the cyst wall is contiguous with the stomach; 2) it is surrounded by a smooth muscle coat, which fuses with the muscle layer of the stomach; and 3) it is lined with alimentary epithelium (4). To date, the etiopathogenetic origin of gastric duplication remains unknown. However, the lesions are typically located dorsal to the primitive gut during development, so that most gastric duplication cysts appear along the greater curvature of the stomach, adjacent to the gastric wall (1). In the present case, the duplication cyst was located at the antrum on the side of the greater curvature of the stomach. The differential diagnoses were an SMT such as GIST, neuroendocrine tumor, unusual gastric carcinoma, heterotopic gastric gland, pancreatic cystic lesion, or pancreatic heterotopia. According to the CT findings, the lesion appeared to be separate from the pancreas. Thus, we could have ruled out pancreatic cystic lesions in the differential diagnosis; however, there was a possibility that the tumor originated from the pancreas due to the distribution frequency of cystic lesions around the stomach. Moreover, the operative procedures and complications for the stomach and the pancreas differ completely. This point was very important for obtaining informed consent from the present patient before surgery. Although CT and MRI showed the cystic le-
Figure 3. Endoscopic ultrasonography shows that the cystic lesion is three-layered, hypoechoic, and contiguous with the stomach through the first to the third layer of the proper stomach (A). The resected specimen of the distal gastrectomy is shown. The cyst is smaller after aspiration (B).

Figure 4. The mass was longitudinally-incised. A pathological examination shows the inner side of the cystic lesion composed of a single layer of columnar epithelium with glandular formation (B). In addition, the lesion shares the layer of the mucosa and submucosa of the proper stomach, indicating that it is a gastric duplication cyst (A, C).

...sion around the distal stomach, these diagnostic modalities were not beneficial for the qualitative diagnosis. We could not preoperatively diagnose GDC, despite using diagnostic modalities. Subsequent upper endoscopy only suggested that the lesion had originated in the stomach. Eventually, we came close to a preoperative diagnosis by EUS, which clearly showed that the cystic lesion was surrounded by the first to the third layer of the stomach (Fig. 3A). At the time, we speculated that the cystic lesion was probably a GIST with cystic change. Surgery was required for the therapeutic diagnosis, and the definitive diagnosis of GDC could not be made until removal of the lesion. Fig. 4 illustrates the histo-
pathological findings of the present case. The lesion shared both the mucosa and submucosa of the proper stomach, with a common blood supply. The smooth muscle coat was fused with the muscularis propria of the proper stomach. The inside of the cyst was lined by gastric epithelium. These findings fit the definition of GDC proposed by Rowling. (4).

In general, the preoperative diagnosis of GDC is difficult due to its extreme rarity. One of the reasons for the difficulty in diagnosing GDC may be that most endoscopists are unfamiliar with GDCs. Namely, few endoscopists list GDC in a differential diagnosis when they encounter SMT-like lesions. Furthermore, in most cases, GDCs are recognized during childhood. As a result, adult GDCs are often difficult to diagnose by endoscopists because they are asymptomatic and tend to be small in size.

Regarding treatment, there is no therapeutic algorithm for GDCs. On one hand, surgical treatment is recommended for symptomatic cases; on the other hand, some investigators also recommend surgical removal for asymptomatic cases to prevent complications in the medium- and long-term. Malignancy is a complication of GDCs. Liu et al. described the clinicopathological diagnosis for cases of malignancy arising from GDCs. Adenocarcinoma is likely the most common histologic type of gastric duplication cyst reported (5). In the present case, we selected surgical treatment for the following reasons: 1) the patient presented with outlet obstruction due to BVS by the GDC and 2) because the endoscope-guided FNA cytology was class III, malignancy could not be excluded. Thus, surgical treatment was the only option. We speculate that the primary reason for the absence of symptoms in the patient until adulthood is that BVS did not occur because of the GDC; the patient’s excessive eating incidentally induced BVS.

In conclusion, when endoscopists encounter a cystic lesion mimicking an SMT, GDC should be considered in the differential diagnosis.

The authors state that they have no Conflict of Interest (COI).

References

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