Gastric duplication cyst (GDC) associated with ectopic pancreas: Case report and review of the literature

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ABSTRACT

INTRODUCTION: Duplication of the alimentary tract is a relatively rare congenital anomaly. It can affect any part of the gastrointestinal tract, with ileum being the most common site. These malformations are believed to be congenital, formed before the differentiation of epithelial lining, and therefore named for the organ with which they are associated. Duplication cysts of the stomach represent four percent of all alimentary tract duplications.

CASE REPORT: Here, we report a rare case of symptomatic duplication cyst of stomach associated with ectopic pancreas presenting in adult.

DISCUSSION: Gastrointestinal duplication is a relatively rare anomaly that may occur at any level from oral cavity to rectum with ileum being the most common site. Duplication cysts of the stomach are quite rare, and most of them have been reported in children. Duplication cysts of ileum are usually located on the mesenteric border, whereas the usual location for gastric duplication cysts is along the greater curvature. The duplication cyst is entirely separated from the adjacent bowel but shares a common wall. Complete removal is the treatment choice to avoid the risk of possible complications such as obstruction, torsion, perforation, hemorrhage, and malignancy. A non-communicating GDC is classically treated by complete excision of the cyst and resection of the shared wall between stomach and the duplication cyst.

CONCLUSION: This unusual developmental anomaly should be included in the differential diagnosis of cystic masses of the gastrointestinal tract, and the possibility of malignancy should also be considered, so as be treated surgically by complete resection.

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1. Introduction

Duplication of the alimentary tract is a relatively rare congenital anomaly. It can affect any part of the gastrointestinal tract, with ileum being the most common site. These malformations are believed to be congenital, formed before the differentiation of epithelial lining, and therefore named for the organ with which they are associated. Duplication cysts of the stomach represent four percent of all alimentary tract duplications [1]. Approximately 67% of gastric duplication cysts (GDCs) are identified within the first year of life. Duplication cysts in adults are generally asymptomatic and encountered as incidental findings at endoscopy or laparotomy. Here, we report a rare case of symptomatic duplication cyst of stomach associated with ectopic pancreas presenting in adult.

2. Case report

A 38-year-old man was referred to the surgical department of our hospital for evaluation of recurrent, non-radiating epigastric pain associated with nausea, mild fever and occasional vomiting for two months. He denied any change in bowel habits and weight loss. His medical history was not significant for any medical or surgical condition. Physical examination was unremarkable except for mild epigastric tenderness.

Diagnostic evaluation included abdominal CT scan with contrast medium, which demonstrated the cyst as a lesion with thick-walled cystic origin and with enhanced outline of the inner layer. The calcification of the cyst is occasionally a characteristic CT finding (Fig. 1).

Endoscopic ultrasound showed a cystic formation in the stomach with an echogenic mucous layer and an intermediate muscular layer, characteristics of GDC (Fig. 2). It is very likely that the wall of the cyst cannot be always distinct in endoscopic ultrasound and the diagnosis may be difficult.

An x-ray examination with barium meal was also conducted, also revealing the gastric duplication cyst (Fig. 3).

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Fig. 1. CT scan demonstrated a lesion with a thick-walled cystic origin and with an enhanced outline of the inner layer, suggestive of duplication cyst.

Fig. 2. Endoscopic ultrasound showed a cystic formation in the stomach with an echogenic mucous layer and an intermediate muscular layer, suggestive of GDC.

Fig. 3. X-ray examination with barium meal.

Diagnostic workup in gastric duplication cyst also includes a full blood count and patient’s biochemical profile, upper GI endoscopy and MRI of the abdomen.

An exploratory laparotomy was conducted, revealing the cystic lesion along the greater curvature of the stomach and in contact with ectopic pancreatic tissue. Surgical removal of the cystic lesion was the procedure of choice (Figs. 4–6).

Patient’s postoperative course was uneventful. He was discharged on postoperative day 7 and has been asymptomatic since then.

Cut surface of the specimen revealed one cyst filled with clear mucinous fluid. On microscopy, cyst wall was composed of mucosa, submucosa, and muscularis propria. Mucosa was predominantly of pyloric type (Figs. 7–8).

3. Discussion

Gastrointestinal duplication is a relatively rare anomaly that may occur at any level from oral cavity to rectum with ileum being the most common site. Duplication cysts of the stomach are quite rare, and most of them have been reported in children. Duplication cysts of ileum are usually located on the mesenteric border, whereas the usual location for gastric duplication cysts is along the greater curvature. The duplication cyst is entirely separated from the adjacent bowel but shares a common wall.

The essential criteria for diagnosis of a gastric duplication cyst are (a) the wall of the cyst is contiguous with the stomach wall; (b) the cyst is surrounded by smooth muscle, which is continuous with the muscle of the stomach; and (c) the cyst wall is lined by epithelium of gastric or any other type of gut mucosa.

Our case fulfilled these criteria excluding other diagnoses.
Gastric duplication cysts comprise 4% of all gastrointestinal duplications. Various other congenital anomalies such as alimentary tract duplications, esophageal diverticulum, or spinal cord abnormalities are encountered in up to 50% patients. These malformations are believed to be congenital, formed before the differentiation of epithelial lining, and therefore named for the organs with which they are associated. Duplications result from the disturbances in embryonic development, and various theories have been proposed for the actual mechanism. Bremer proposed the theory of errors of recanalization and fusion of longitudinal folds. He suggested 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 month of intrauterine life. McLetchie suggested that adhesion of notochord and embryonic endoderm might not elongate as quickly as its surrounding structures, causing traction diverticulum leading to duplication cyst formation. Other theories of enteric duplication include abortive twinning, persistent embryological diverticula, and hypoxic or traumatic events. There is no single theory that is satisfactory for all types of duplications.

Greater than 80% of gastric duplications are cystic and do not communicate with the lumen of the stomach. The remainders are tubular with some communication. The structure is defined as tubular when the lumen is contiguous and cystic when the lumen is not contiguous with stomach lumen. The mucosal lining of duplication may be histologically similar to the segment of gut to which it is topographically related. However, some duplications may include lining from other segment of the alimentary or respiratory tract [2]. The presence of respiratory epithelium in the cysts of thorax, tongue, liver, and stomach suggests that the undifferentiated epithelium of foregut might undergo transition to differentiated specialized epithelium during embryonic period.

Gastric duplications typically become symptomatic during childhood. 67% are diagnosed within the first year of life, and less than 25% are discovered after age 12. The duplication cysts of the stomach are usually diagnosed intra-operatively in adults. In our first patient, the preoperative CT and MRI findings were interpreted as being most consistent with a pancreatic neoplasm, and diagnosis of GDC was suspected only during surgery.

The clinical presentation of gastric duplication cysts can be highly variable and nonspecific ranging from vague abdominal pain to nausea, vomiting, epigastric fullness, weight loss, anemia, dysphagia, dyspepsia with abdominal tenderness and epigastric mass on physical examination. Because most cases occur along the greater curvature of the stomach, the cysts can potentially compress the adjacent organs such as pancreas, kidney, spleen, and adrenal gland. Accordingly, the differential diagnosis would include lesions arising from these organs. The cysts may also be manifested by complications such as infection, gastrointestinal bleeding, perforation, ulceration, fistula formation, obstruction, compression, or carcinoma arising in the cysts [3]. Up to 10% of gastric duplications may contain ectopic pancreatic tissue which may lead to pancreatitis and mimic a pancreatic pseudocyst.

Duplication cysts have the potential for neoplastic transformation [4]. The production of oncofetal antigens raises the problem of a precancerous condition in long standing intestinal duplications. Another issue of investigation and debate is the fact of coincidence of epithelial and non-epithelial components of GDC which can give rise respectively to epithelial and non-epithelial gastric neoplasms [5]. Out of 11 reported cases of malignancy arising within the duplication cysts, 8 were adenocarcinomas (epithelial tumors). Five of the carcinomas originated from gastric duplications. Adenomyoma (non-epithelial tumor) arising from a gastric duplication has also been reported. Malignancies arising from duplication cysts are likely to be present at advanced stages because of their unusual symptoms and difficulty of diagnosis.

Additionally, GDC can mimic other malignancies as neuroendocrine and gastrointestinal stroma (GIST) tumors [6].

Although it is difficult to diagnose GDC preoperatively, recent imaging modalities have provided some informative findings. CT scan and endoscopic ultrasound (EUS) are the best ways to identify GDC. Classically, radiographic studies show an intramural filling defect indenting the gastric contour. Contrast-enhanced CT scan typically demonstrates GDC as a thick-walled cystic lesion with enhancement of the inner lining. Calcification is occasionally observed on CT. These findings are of diagnostic significance for...
GDCs. However, since mucinous cystic tumors of the pancreas also show similar radiological features, GDCs adjoining the pancreas are indistinguishable from pancreatic mucinous cystic tumors based on these CT findings. Moreover, because the wall is sometimes thin, enhancement of the inner cyst wall is not always demonstrated. Generally, MRI can provide additional information about the cyst content compared to CT scan. However, the nature of the fluid in the GDC was reported to differ in each case according to bleeding, chronic inflammation, or infection. Therefore, MRI seems to be of less significance than expected in diagnosing GDCs. EUS is useful in distinguishing between the intramural and extramural lesions of the stomach. When EUS demonstrates a cyst with an echogenic internal mucosal layer and a hypoechoic intermediate muscular layer, the diagnosis of GDC is highly likely.

Complete removal is the treatment choice to avoid the risk of possible complications such as obstruction, torsion, perforation, hemorrhage, and malignancy [4]. A non-communicating GDC is classically treated by complete excision of the cyst and resection of the shared wall between stomach and the duplication cyst. Communicating GDC usually requires no intervention when both gastric lumens are patent. Drainage and marsupialization of the cyst have been suggested. However, marsupialization into the stomach exposes the unprotected mucosa of the cyst to gastric contents with the risk of ulceration. Drainage procedures such as cystojejunosotomy may be complicated by stenosis of the Anastomosis or blind loop syndrome and therefore discouraged. Furthermore, leaving the cyst in place is ill-advised given the potential for malignant transformation.

4. Conclusion

In summary, this extremely rare condition, which has a severe diagnostic difficulty, should be considered in the differential diagnosis of cystic masses, taking into consideration its imaging ability to mimic other malignancies or even more its own malignant potential. Regarding the latter, researchers must take into consideration that this unique entity can produce either epithelial or nonepithelial cancers as well. Finally, due to the risk of malignant transformation and other complications, complete surgical resection can offer definitive diagnosis and treatment as in our case.

Conflict of interest

The authors declare that they have no conflicts of interest.

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Ethical approval

The ethical committee of our military hospital has approved the case report to be sent for publication.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request [7].

Author contribution

G. Chatzoulis: study concept and design, main operator, General Surgeon.
ID Passos: data collection and writing the paper, General Surgery Resident.
K. Milias: data collection, General Surgeon.
C. Christoforakis: data collection.
P. Spyridopoulos: study concept and design, second operator, General Surgeon.
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