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CASE REPORT | STOMACH

Gastric Duplication Cyst With Occult GIST Component

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ABSTRACT

A 37-year-old man underwent screening examinations, and a pale-colored submucosal tumor was detected on gastric cardia. Endosonography showed a 15-mm cyst with hypoechoic nodules in the muscularis propria, and endoscopic ultrasound-guided fine-needle aspiration obtained mucinous fluid with atypical spindle cells positive for *c-kit*, indicating a gastrointestinal stromal tumor. Surgical resection was recommended, but he initially agreed to surveillance. After becoming larger for 8 years, partial gastrectomy was performed for the 22 × 22 × 15-mm capsulized lesion. Surprisingly, its histology was gastric duplication cyst without gastrointestinal stromal tumor. Gastric duplication cyst is a rare entity with the possibility of malignant complications, but careful assessment of endoscopic ultrasound-guided fine-needle aspiration might also be required.

INTRODUCTION

Enteric duplication cyst is a rare congenital anomaly, mainly diagnosed during the first year of life. Gastric duplication cyst (GDC) accounts for 2%–9% of all enteric duplication cysts.[1] Its criteria were proposed as follows: (i) cystic structures located in or adjacent to the gastric wall; (ii) those surrounded by smooth muscle fibers, fusing with a layer of the muscularis propria of the original wall; and (iii) those lined with epithelium consisting of gut mucosa.[2] Its endoscopic findings are mainly a submucosal tumor (SMT) covered with smooth epithelium that does not communicate with the gastric lumen. The accuracy of endoscopic biopsies was reported to be very low. Most adult cases are diagnosed incidentally after surgery, whereas the risk for complications, including perforation, hemorrhage, and malignancy, might increase during an asymptomatic course. The prognosis of cases with malignant complications is poor according to previous reports of 10 GDC cases with concomitant adenocarcinoma, 1 with a neuroendocrine tumor, 1 with squamous cell carcinoma, and 2 with gastrointestinal stromal tumor (GIST).[1,3–5] Recently, endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) has been recommended for making a preoperative diagnosis of SMTs, but its yield for gastric cystic lesions remains uncertain.

CASE REPORT

A 37-year-old asymptomatic male without medical history visited a general hospital for screening examinations. A physical examination and laboratory data revealed no abnormalities. Esophagogastroduodenoscopy (H260, Olympus, Tokyo, Japan) depicted a pale-colored SMT covered with normal-appearing mucosa on the lesser curvature of the gastric cardia. EUS (UM-2000, Olympus) demonstrated a 15-mm anechoic cyst with small hypoechoic nodules in the muscularis propria (Figure 1). Computed tomography (CT) revealed a well-circumscribed cystic mass with irregularly thickened walls without enhancement of iodine contrast agents and fluorodeoxyglucose positron emission tomography (SUVmax 1.2). EUS-FNA was performed in 3 sessions using a 19-gauge EchoTip Ultra Endoscopic Ultrasound Needle (Cook Med., Bloomington, IN), which obtained mucinous fluid with whitish fragments composed of proliferative and atypical spindle cells, whose immunostaining was strongly and diffusely positive for *c-kit* and negative for α-smooth muscle actin (Figure 2).

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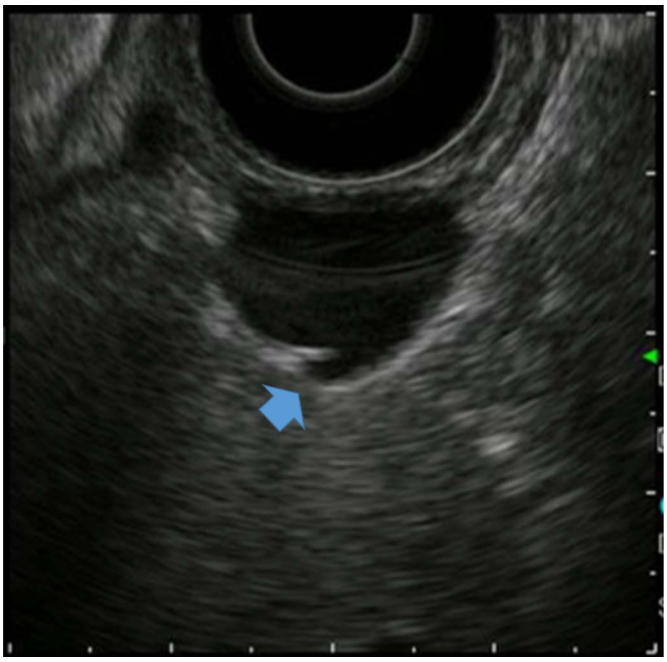

Figure 1. Gastric duplication cyst which is 15 × 9 mm in size with small hypoechoic nodules (arrow) was in the layer of the muscularis propria of the original gastric wall in the 12-MHz endoscopic ultrasound findings.

With a diagnosis of GIST with cystic degeneration, surgical resection was recommended, but the patient chose surveillance. After 8 years, the endoscopy showed that it had become larger (Figure 3). The patient had a partial gastrectomy by laparoscopy endoscopy cooperative surgery (Figure 4). Macroscopically, it was a 22 × 22 × 15-mm capsulized lesion containing a clear mucoid substance in the muscularis propria. Its histology was GDC with a single layer of columnar epithelium, staining positive for cytokeratin 7 and thyroid transcription factor-1 and negative for cytokeratin20, MUC5AC, and MUC6, whereas the inner side of the wall lacked GIST components, and the whole lesion was surrounded by smooth muscle fibers shared with the muscularis propria (Figure 5). Postoperative workup showed no recurrence.

DISCUSSION

GDC is a rare anomaly with clinically miscellaneous features. The present case was unusual in terms of its location, size, and the co-existence of an occult GIST component.[1] During gut development, GDCs are frequently located on the greater curvature of the proximal stomach, and only 5.5% of reported cases were on the lesser curvature. The size generally ranges between 1.6 and 40 cm, and the symptoms are multiplex. CT and EUS are thought to be useful diagnostic modalities. CT reveals a fluid-attenuation cystic mass adjacent to the gastric wall, but the finding of GDC with a high protein content can be confused with a solid mass. EUS may help to identify the origin, border, and echo level of tumors. The finding of GDC is typically that of an anechoic SMT, lined with a hypoechoic epithelial layer and a hyperechoic subepithelial layer, and surrounded by a hypoechoic layer, which is fused with the muscularis propria of the original wall.[5] However, because of the limited resolution of EUS, the echo levels and numbers of cystic components were reported to be too variable to distinguish GDC from other malignancies.

Recently, histological assessment by EUS-FNA for solid SMTs has been proposed. The sensitivity, specificity, and accuracy of EUS-FNA in diagnosing neoplasms of the alimentary tract were reported to be 61%–96%, 79%–88%, and 67%–92%, respectively.[6] Moreover, a previous report demonstrated the usefulness of EUS-FNA for diagnosing GDCs, whose characteristics were (i) mucous membrane with a well-organized layer of smooth muscle, (ii) granulomatous inflammation with

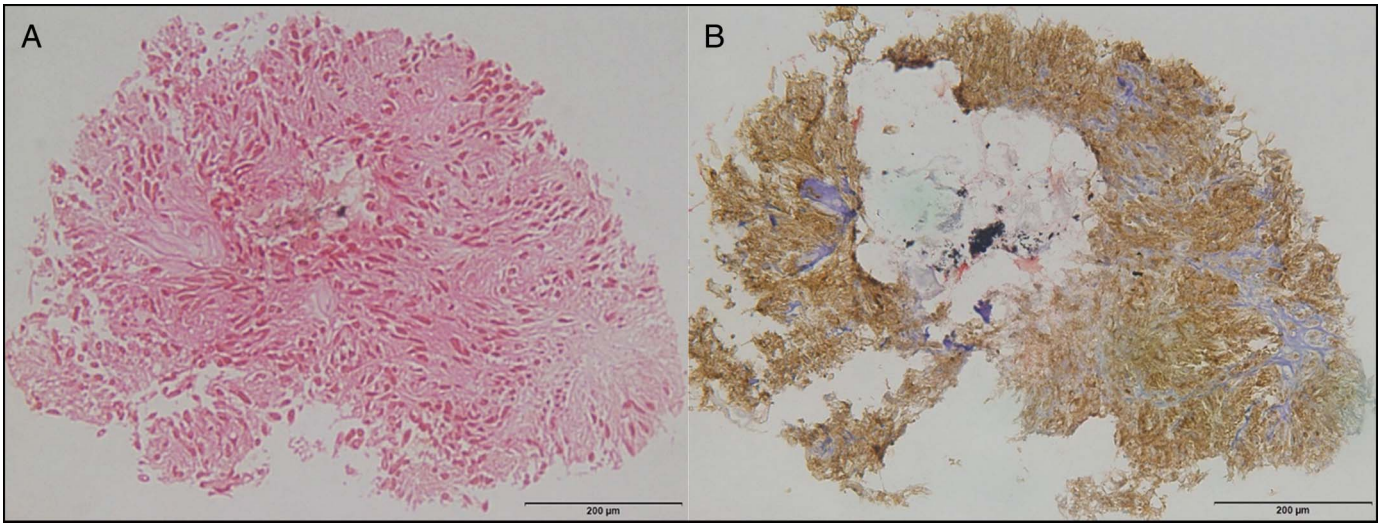

Figure 2. Histology of the sample obtained through endoscopic ultrasound fine-needle aspiration showing that (A) proliferative and atypical spindle cells (haemotoxylin and eosin staining) and (B) some of the proliferative and atypical spindle cells were strongly and diffusely positive for *c-kit*.

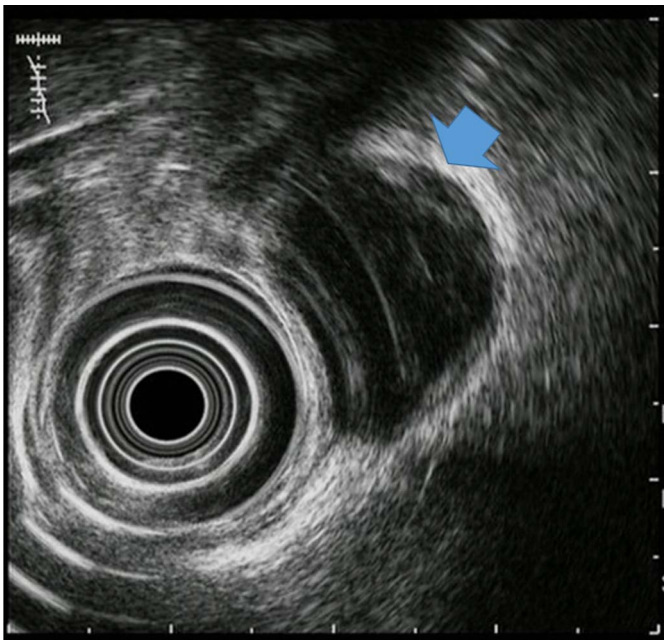

Figure 3. Endoscopic image of the tumor at the 8-year follow-up showing that the tumor had grown to 18.1 × 13 mm.

denuded mucosa, and (iii) foreign body giant cells, but none of these were observed in our case. Considering that EUS-FNA might not be required for asymptomatic congenital or benign tumors, its yield for asymptomatic GDCs remains controversial.[7,8]

Certainly, malignant complications might occur in adult GDC cases, and 2 cases of GDC concomitant with GISTs have been reported.[3,4] In those cases, small nodules were macroscopically detected inside the wall of 20–95 mm-sized cystic masses in the surgical specimens, but none of them were proven by EUS-FNA. In our case, the EUS-FNA was targeted for a tiny nodule in the mass using a 19-gauge needle, resulting in histological diagnosis of a *c-kit*-positive GIST. The follow-up EUS demonstrated that the echo level of the nodular component had changed in the larger mass, but there was no GIST component in the surgical specimen. Therefore, we could not exclude the possibility of GDC concomitant with the smallest GIST.

Of course, there remains the other possibility of a false-positive in the EUS-FNA assessment. Two previous reports described that samples from a 60 mm-sized GDC were cytologically interpreted as Class IIIb and that the carcinoembryonic antigen and CA19-9 levels and the existence of mucin in the intracystic fluid might be unreliable markers of malignancy.[9,10] In addition, several studies pointed out that the aggregates of spindle-shaped neoplastic cells of GIST might be misinterpreted as smooth muscle hyperplasia of the alimentary wall when their cellularity is low.[7] Although it is recommended to apply on-site cytological interpretation and an immunocytochemical study of EUS-FNA samples, the staining/sample condition might cause misdiagnosis.

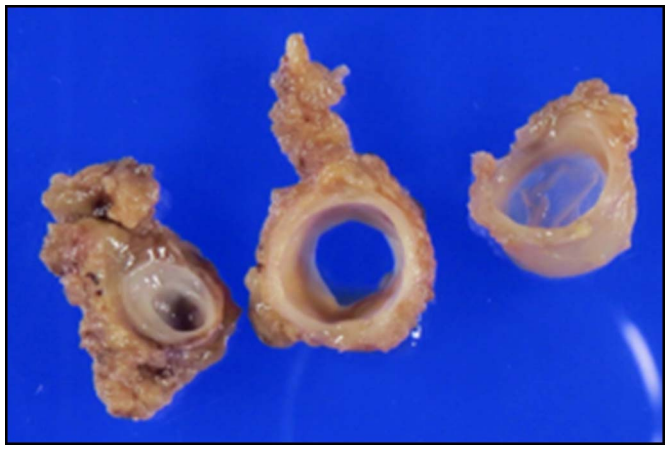

Figure 4. Specimen of partial gastrectomy by laparoscopy endoscopy cooperative surgery revealed a 22 × 22 × 15 mm-capsulized lesion containing clear mucoid substance in the muscularis propria of the gastric wall.

We demonstrated a case of GDC with an occult GIST component for the first time. Nowadays, the demand for EUS-FNA for the diagnosis of SMTs has been increasing, but the diagnosis of GDC is difficult because of its rarity with miscellaneous features, together with the uncertainty in obtaining appropriate samples. Therefore, we should pay careful attention when evaluating the cytological results of EUS-FNA samples from gastric cystic tumors.

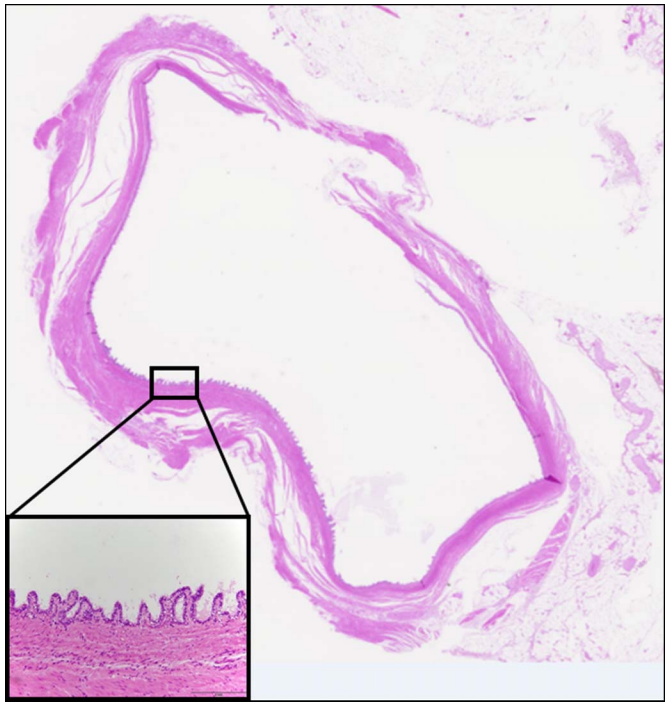

Figure 5. Histological examination demonstrated a single layer of columnar epithelium in the inner side of the wall without the nodules of gastrointestinal stromal tumor, and the whole lesion was surrounded by smooth muscle fibers shared with the muscularis propria (haemotoxylin and eosin staining 20×, column 200×).

DISCLOSURES

Author contributions: S. Lee, K. Uno, F. Fujishima, W. Hatta, T. Koike, T. Aoki, N. Tanaka, H. Musha, and R. Kikuchi acquired the data. S. Lee and K. Uno wrote the manuscript. S. Lee, K. Uno, T. Koike, and A. Masamune revised the manuscript and approved the final version. K. Uno is the article guarantor.

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Informed consent was obtained for this case report.

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