CASE REPORT

Gastrointestinal Stromal Tumor Mimicking a Pancreatic Cystic Lesion: A Case Report

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ABSTRACT

Gastrointestinal stromal tumors are a small portion of gastrointestinal malignancies that usually present in the stomach. This is a case of a sixty-four-year-old male who presented with large gastrointestinal stromal tumors that had undergone cystic degeneration, and was initially thought to be a pancreatic pseudocyst due to radiographic and biochemical findings. The diagnosis was established after an endoscopic ultrasound was performed with fine needle aspiration showing histology and endoscopic findings consistent with a neoplasm. This is an interesting case due to the atypical presentation of the gastrointestinal stromal tumors.

INTRODUCTION

Gastrointestinal Stromal Tumors (GISTs) are the most common mesenchymal neoplasms of the gastrointestinal tract, although they only account for 0.1%-3% of all gastrointestinal malignancies. The majority arise from the stomach and present as solid masses. In rare cases, they can undergo cystic degeneration and be misdiagnosed as cystic lesions of the pancreas due to similarities in clinical presentation and appearance on imaging studies [1, 2, 3].

CASE REPORT

A sixty-four-year-old male presented with acute right upper quadrant and epigastric abdominal pain that had been intermittent over the past 2 months. He had associated abdominal distension, nausea, vomiting, early satiety and melena.

Laboratory workup revealed a serum amylase of 210 U/L (range 40-140 U/L) and lipase of 336 U/L (range 0-160 U/L). Liver function tests, CEA and CA19-9 levels were within normal limits. His hemoglobin was 11.2 g/dL (MCV of 86 fl), serum iron 13 mcg/dL, TIBC 276 mcg/dL, and Ferritin 55 ng/mL. CT scan of the abdomen demonstrated a large, predominantly necrotic cystic mass measuring 15.8×14.7×13.4 cm, in the left upper abdomen. Based on his clinical presentation and imaging findings, the patient was diagnosed with possible acute pancreatitis with pseudocyst.

Pt underwent an upper endoscopy which showed an area of extrinsic compression on the gastric fundus and proximal gastric body along the greater curvature **(Figure 1a)**. Endoscopic Ultrasound (EUS) revealed a large, unilocular cystic lesion measuring 15×14 cm that was contiguous with the anterior aspect of body and tail of the pancreas and abutted the muscularis propria of the gastric body **(Figure 1b)**. Furthermore, EUS demonstrated areas of intramural calcification suggestive of a malignant cystic lesion **(Figure 1c)**.

Fine needle aspiration (FNA) obtained 200 mL of viscous, brown fluid with debris. Cystic fluid analysis revealed an amylase level of 57 u/L and CEA of 0.8 ng/mL, suggesting this was neither a pseudocyst nor a mucinous cystic neoplasm of the pancreas.

The patient presented again three weeks later with melena, repeat upper endoscopy an ulcer at the site of gastric compression. As well as a fistula at the center of the ulcer draining a copious amount of brown fluid **(Figure 1d)**. During endoscopic visualization there was no active bleeding or high-risk stigmata of bleeding warranting endoscopic therapy. We obtained biopsy of the thick wall of the cystic lesion in the region where EUS demonstrated its origin from the gastric muscularis propria with EUS-19G Fine Needle Biopsy (FNB) needle that provided a sample adequate for histology and forceps biopsy of the aspect of the lesion with mucosal ulceration.

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Abbreviations EUS Endoscopic ultrasound; GIST Gastrointestinal stromal tumors
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Histology showed spindle cells occurring singly and in groups **(Figure 2a)**. Immunohistochemistry revealed tumor cells positive for c-Kit and CD 34 **(Figure 2b)**, and negative for desmin, pankeratin, and S-100. Ki-67 was also performed revealing a proliferation index of about 25%-30%. The findings are diagnostic of a GIST. The patient also underwent PET scan, which revealed a hypermetabolic mass in the left upper quadrant with central necrosis and metastases to the liver along with a mesenteric nodule adjacent to the mass. The patient's case was discussed at our interdisciplinary tumor board and based on the PET scan revealing metastatic disease, neoadjuvant Imatinib therapy was recommended. This decision was also made based on the surgical oncologist reviewing the imaging and determining that the large cystic lesion was not resectable at this time.

The patient was initiated on Gleevec (Imatinib), with an excellent response with resolution of metastatic disease and significant reduction in size of the gastric lesion. The tumor was subsequently resected with a subtotal gastrectomy with negative margins (R0 resection) and showed over 90% necrosis **(Figure 2c)**.

DISCUSSION

This is an unusual presentation of a patient with an exophytic Gastric GIST that had undergone cystic degeneration, presenting as a pancreatic pseudocyst. The differential diagnosis of pancreatic cystic lesions generally includes pseudocyst, serous cystadenomas, mucinous cystadenoma, mucinous cystadenocarcinoma, cystic neuroendocrine tumors, lymphangioma and other rare cystic neoplasia that are lumped into the "miscellaneous" category [4, 5, 6]. As illustrated by this case, an extra pancreatic cyst or neoplasm with cystic degeneration may appear to arise from the pancreas on imaging studies and confound the diagnosis, so they must also be considered in the differential diagnosis.

A GIST arising from the stomach are often asymptomatic, the most common clinical manifestation of a GIST is acute or chronic upper gastrointestinal bleeding (61%) ranging from asymptomatic occult bleeding to massive bleeding or a palpable abdominal mass. Large GISTs are usually associated with central ulcerations which penetrate deeply into the tumor mass resulting in intraluminal bleeding [7]. GISTs can also rarely present with bowel or biliary

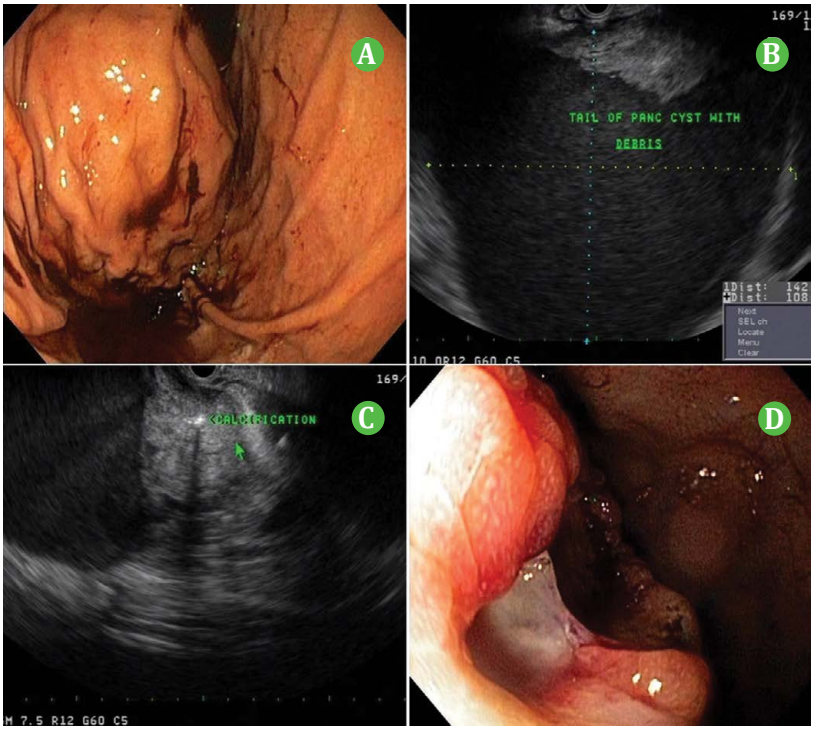

Figure 1. (A). Endoscopic image displaying extrinsic compression on gastric fundus; **(B).** Endoscopic Ultrasound (EUS) image showing 15.0×14.0 cm cystic lesion in tail of pancreas; **(C).** EUS demonstrating eccentric cyst wall thickening with areas of intramural calcification; **(D).** Endoscopic image showing mucosal ulceration at the site of gastric compression seen on previous imaging.

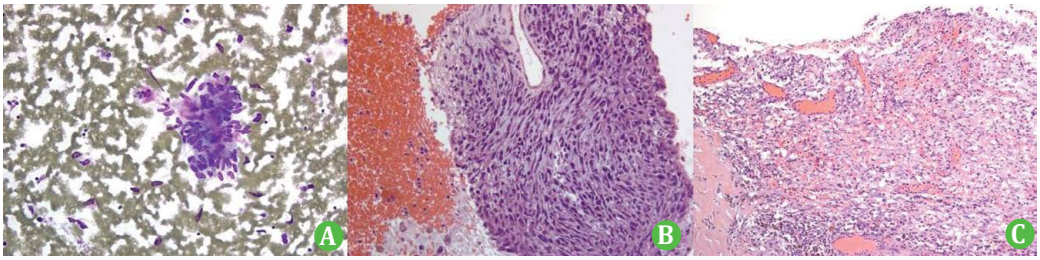

Figure 2. (A). Fine needle aspiration showing small fragment with spindle cells with wavy nuclei, and several stripped, single nuclei; **(B).** Immunohistochemistry was strongly and diffusely positive for CD 117; **(C).** Resection specimen displaying wall of the cystic cavity lined by histiocytes, and reactive fibrobalsts, surrounded by hyalinized fibrosis.

obstruction, dysphagia, intussusception, hypoglycemia, or even with fistulous tract formation between cystic cavity and gastric lumen.

There have been other reports of GISTs presenting as a pancreatic cyst, although it is quite rare [3, 8, 9, 10, 11]. EUS-FNA enables high-resolution imaging of the pancreatic and peripancreatic cystic lesions and enables image guided tissue sampling facilitating a specific diagnosis [12]. EUS is considered superior in the evaluation of intramural lesions of the upper gastrointestinal tract as it provides high-resolution imaging of submucosal lesions and loco-regional staging [13]. EUS, when combined with FNA (Fine Needle Aspiration) contributes to an improved diagnosis with an accuracy of 80%-85% [4]. In our case, EUS-guided FNA played a key role in establishing the diagnosis of a GIST and had a significant impact on specific management and the patient's outcome. Currently we have Fine Needle Biopsy (FNB) needles available that provide core of tissue for histology, but we used a 19 G FNA needle as the patient had recent bleeding and our goal was to both aspirate the cyst fluid and cyst wall with the same needle. As is often the case we were able to obtain an adequate sample for histology with the 19G FNA needle.

The standard of care for non-metastatic GIST that is assessed on cross-sectional imaging to be resectable is surgical resection. For metastatic GIST, large tumors and small GISTs in locations that are considered marginally resectable on technical grounds, neoadjuvant imatinib is recommended [14, 15, 16]. An interdisciplinary tumor board discussion is recommended prior to management decisions on all GISTS as is the clinical pathway at our institution.

CONCLUSION

As shown by this case, clinical symptoms, cross-sectional imaging, and even the gross appearance of the fluid, are not reliable in differentiating among the various possible entities considered in the differential diagnosis of cystic abdominal lesions. Although rare, GIST should be included in the differential diagnosis of pancreatic and peri-pancreatic cystic lesions and its' presentation may mimic a pancreatic pseudocyst or present with gastric fistula. A careful EUS examination and EUS guided fine needle aspiration or biopsy of both the cyst fluid is crucial in making a specific diagnosis and particularly cyst wall sampling may offer specific histological diagnosis as in our patient.

Conflict of Interest

The authors declare that they have no conflict of interest.

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