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Case Report

Gastric gastrointestinal stromal tumor (GIST) with co-occurrence of pancreatic neuroendocrine tumor ☆

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ABSTRACT

Mesenchymal tumors make up only about 1% of primary GI tumors, with Gastrointestinal Stromal Tumors (GIST) being the most common nonepithelial GI neoplasms. They are derived from the Interstitial cells of Cajal (ICC), and occur predominantly in older individuals, with a mean age of diagnosis of 64 years. Here we discuss the case of a 39-year-old female with atypical thoracic back pain wrapping around to the front and migrating diffuse abdomen pain that sometimes radiates into the chest. Upon imaging, a gastric GIST of the greater curve of the stomach was found incidentally on investigation of a pancreatic mass that was revealed to be a co-occurring pancreatic neuroendocrine tumor. For management of the gastric GIST and pancreatic neuroendocrine tumor, this patient underwent partial gastrectomy with gastrojejunostomy, partial pancreatectomy, splenectomy, and cholecystectomy with no complications.

Introduction

Gastrointestinal stromal tumors (GISTs) are rare tumors originating from the Interstitial cells of Cajal (ICC). Approximately 80% of GIST tumors are primarily located in the GI tract. 60% of these GI GIST tumors are found in the fundus of the stomach, 30% are in the jejunum and ileum, 5% in the duodenum, 4% in the colorectal, and few have been reported to be in the esophagus and appendix [14]. Symptoms of GISTs vary depending on the location of the tumor. It may present with symptoms of abdominal pain, bloody stools, fatigue, dyspepsia, nausea or vomiting, constipation, or diarrhea, etc. but are often asymptomatic. This may be due to the generally exophytic growth pattern of stromal tumors involving the outer muscular layer [13]. The clinical silence of early GIST tumors often means discovery of the tumor only after remarkable size is achieved, which also happens to correlate with the tumor's malignant potential and worsened prognosis [2,5]. In fact, up to 25% of GIST tumors are discovered incidentally, and diagnosis is based on histologic assessment of biopsied tissue and identification of the **KIT** proto-oncogene receptor tyrosine kinase, that is selectively expressed by the ICC in the gastrointestinal tract [8].

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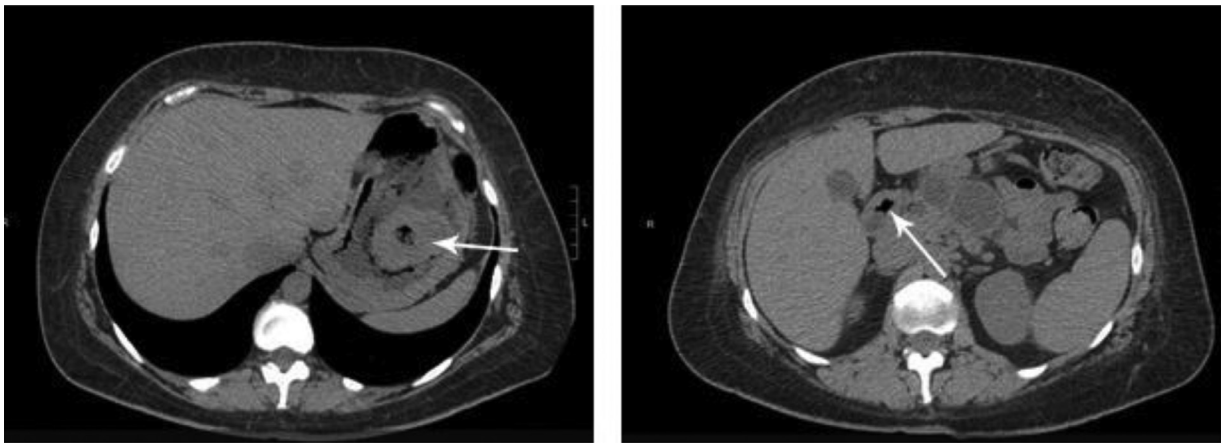

Fig. 1 – CT Abdomen w/o contrast showing Gastric body mass with central ulceration (left image, white arrow) and cystic dilation in pancreas (right image, white arrow)

Generally, in imaging of a GIST tumor, Computed tomography (CT) is the preferred choice for initial imaging studies to investigate a possible GIST. Magnetic resonance imaging (MRI) may provide more information on internal features of the tumor such as hypervascularity and is also better at detecting liver metastasis [9]. CT of a GIST will vary in appearance depending on the size, aggressiveness, and progression of the tumor. Small GIST tumors are usually smoothly contoured, homogeneous masses that enhance brightly with contrast, but large GIST tumors and metastatic tumors are hypervascular, enhance brightly with contrast and appear heterogeneous due to necrosis, hemorrhage, or cystic degeneration [7]. In addition, ulceration, cavitation, and fistula formation are also associated with larger lesions. Positron emission tomography (PET) imaging is often used with CT to stage GISTs and are also useful in equivocal CT/MRI results, as GISTs typically show flourodeoxyglucose (FDG) uptake [1,6].

Surgical resection with clear margins is the most definitive form of treatment for nonmetastatic primary GISTs. According to the National Comprehensive Cancer Network, all GIST tumors greater than 2 cm in size should be resected. However, surgical intervention is not the preferred treatment for tumors that are poorly positioned for resection and tumors smaller than 2 cm. Imatinib mesylate has been shown to be effective in the treatment of smaller and unresectable tumors as well as for decreasing the tumor size prior to surgery and reducing risk of recurrence through neoadjuvant and adjuvant therapy, respectively [3].

Case summary

A 39-year-old female with a history of hypertension and tobacco use initially presented to the ED with atypical thoracic back pain wrapping around to the front and migrating diffuse abdominal pain that radiated to the chest. Lab work and chest X-ray were both unrevealing.

GI ultrasound showed gallstones, as well as an abnormality in the pancreas, warranting further investigation. Abdominal CT with and without contrast was performed, revealing a cystic lesion and dilation of the pancreatic duct, atrophy of the pancreatic tail, and a gastric body mass (Fig. 1). The gastric mass was a noncalcified and lobulated, arising from the cephalad and left lateral wall of the gastric fundus to body, measuring 6.2 × 5.8cm, and had diffuse heterogeneous internal enhancement. In addition, central ulceration of the surface of the mass was evident with some gas or debris within the defect. Abdominal MRI was consistent with CT results, and showed the gastric mass extending both intra and extraluminally, with high suspicion for transdiaphragmatic extension as well, although there were no additional features indicating a specific histology of the mass (Fig. 2).

Esophagogastroduodenoscopy demonstrated a 6 cm lobulated mass on greater curvature of the body of the stomach, which was biopsied and found to be GIST. She was also referred for EUS of her pancreatic mass, which revealed 9mm dilation of the pancreatic duct and a 4.4 × 3.7cm pancreatic lesion with internal debris and fluid. EUS also showed a large, ulcerated mass in the stomach consistent with GIST. Biopsy of the pancreatic mass showed atypical cells consistent with a neuroendocrine tumor. Amylase was 5250 u/L and CEA was low at 2.3.

Gallium Dotatate PET scan demonstrated PET avidness of the pancreatic lesion, and she was found to have elevated pancreatic polypeptide. The gastric mass was found to have no associated radiotracer uptake, further increasing suspicions that it was a separate lesion from the pancreatic tumor (Fig. 3).

The patient was referred for surgery, where she underwent exploratory laparotomy, cholecystectomy, extended distal pancreatectomy, use this one OK splenectomy, wedge resection of liver section 7 and 3, abdominal lymphadenectomy, and omentectomy, as well as intraoperative US of liver and pancreas.

Pathology report of the specimens showed Neuroendocrine carcinoma of the Pancreas and malignant GIST. The GIST was determined to be low grade spindle type GIST positive for CD34 and c-KIT.

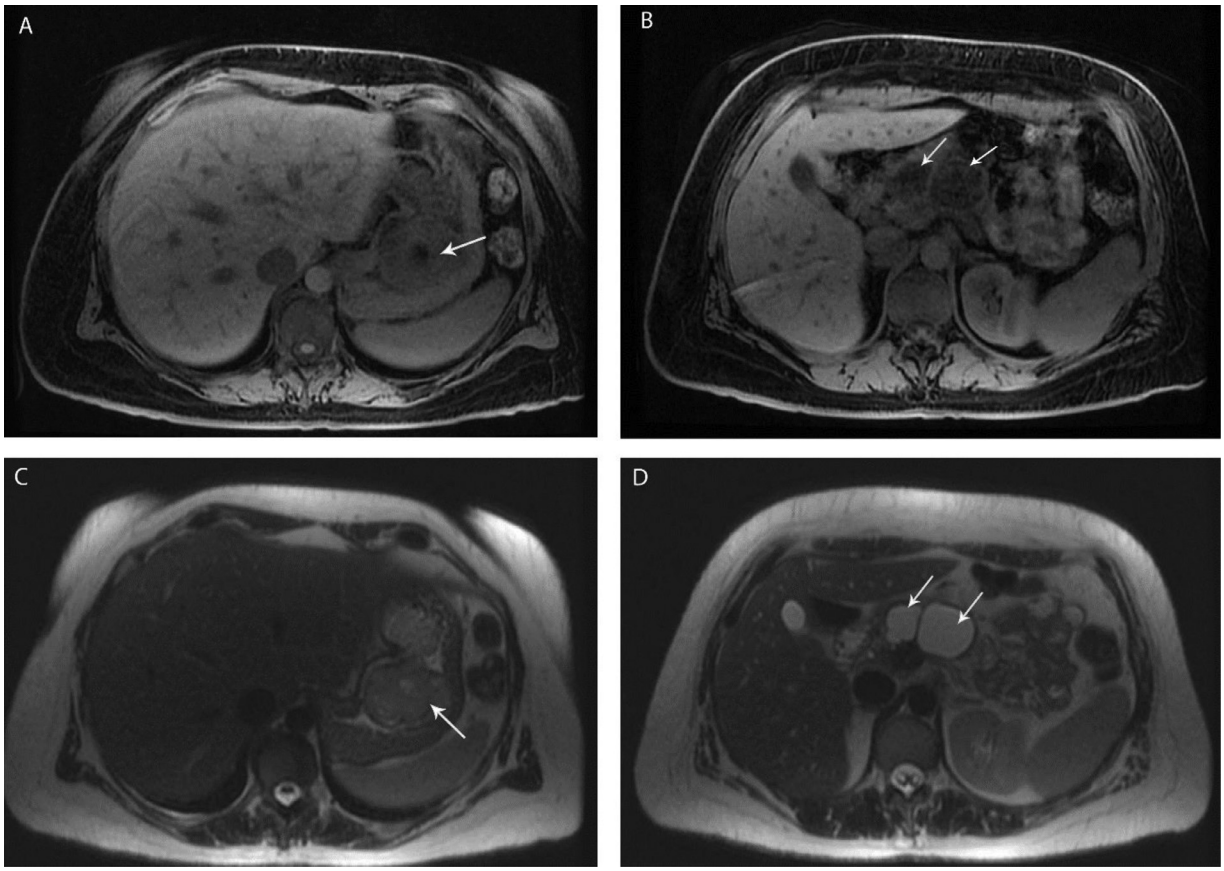

Fig. 2 – Axial Water LAVA-Flex MRI Abdomen w/o contrast showing ulcerated gastric body mass (A, white arrow) and cystic dilations in pancreas (B, white arrows). Axial SSFSE MRI Abdomen w/o contrast showing gastric body mass (C, white arrow) and cystic dilations in pancreas (D, white arrows)

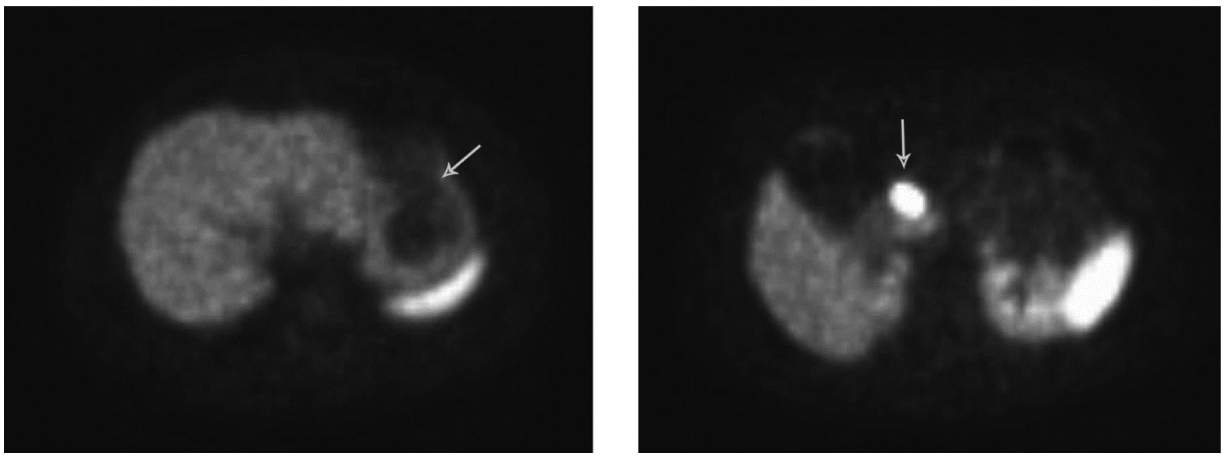

Fig. 3 – Gallium Dotatate PET Abdomen showing lack of radiotracer uptake in gastric tissue(left image, white arrow), compared to PET avid pancreatic lesion (right image, white arrow)

Postoperatively she was started on insulin POD2 for hyperglycemia. UGI Xray was obtained and Regulin was started for delayed gastric emptying. Otherwise, the patient tolerated the procedure well with no complications.

Discussion

GISTs are usually sporadic, with only 5% of occurrences being as part of a familial syndrome [4], including Carney Stratakis Syndrome (CSS), Carney triad, Neurofibromatosis type 1 and

primary familial GIST syndrome. Here, the gastric GIST was found along with pancreatic NET, and co-occurrence of GIST with another primary tumor is usually seen in familial syndromes. In addition, the median age of discovery for GISTs is 63 years. Only 9.3% of tumors are discovered before the age of 40 [10], making our 39-year-old patient an unusual case and raises the question of whether her GIST and concurrent pancreatic NET are coincidental or not. In the known hereditary syndromes, GISTs are associated with co-occurring paragangliomas or pulmonary chondromas as with Carney's triad and Carney-Stratakis syndrome. GISTs also have an incidence of 7% in Neurofibromatosis type 1 patients, where they often occur with nerve plexus hyperplasia or endocrine tumors of the periampullary regions, which is most similar to the case we present here [11,12]. However, our patient has no documented genetic syndromes at the time of writing this case report.

Although extremely rare, GISTs have been described as co-occurring with other neoplasms in literature. However, a large majority of the co-occuring neoplasms consist of adenocarcinomas of the GI tract, lymphomas, or adenomas. The co-occurrence of a GIST along with a pancreatic neuro-endocrine tumor is extremely rare and normally presents in patients with familial syndromes or other genetic backgrounds such as Neurofibromatosis Type 1 patients. In this case, we presented a symptomatic 39-year-old patient without genetic conditions, presenting with pancreatic NET and incidental GIST, who was managed surgically with no post-operative complications. We conclude that although certain genetic syndromes have a predisposition for co-occurring neoplasms and some evidence does point to mutations within the NF1 gene causing GIST and NET occurrence, more research needs to be done to identify a similar mechanism in patients without NF1.

Patient consent

Patient consent for this case report was waived in accordance with our institutional IRB's policies. Letter of determination from the IRB is attached to the submission. All identifiable information has been removed in the writing of this case report

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