

CASE REPORT

Extra-Gastrointestinal Stromal Tumor Presenting as Hemorrhagic Pancreatic Cyst Diagnosed by EUS-FNA

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ABSTRACT

Context Pancreatic gastrointestinal stromal tumors (GIST) are rare mesenchymal tumor with only 6 cases reported to date. We report a case of pancreatic GIST presenting as hemorrhagic cyst. **Case report** A 63-year-old female with past medical history of hypertension and pancreatic mass presented with fatigue. She was found to have anemia requiring blood transfusion. An abdominal CT scan revealed an 11x16 cm cystic mass at the pancreatic body that had increased in size compared with previous CT scan. Endoscopic ultrasound confirmed a large complex pancreatic mass and fine needle aspiration demonstrated gross bloody fluid. Cytology revealed a spindle cell lesion. Immunohistochemistry from cyst wall biopsy was strongly positive for CD34 and CD117 confirming the diagnosis of pancreatic GIST. **Conclusion** We report a case of pancreatic GIST which presented as hemorrhagic cyst. Endoscopic ultrasound guided fine needle aspiration plays an important role in the diagnosis. Although it is an uncommon tumor, pancreatic GIST should be in differential diagnosis of hemorrhagic pancreatic cystic lesions as well as a rare cause of solid pancreatic lesions.

INTRODUCTION

Gastrointestinal stromal tumors (GIST) are the most common mesenchymal tumors mainly occurring in the stomach and small intestine. They are occasionally found in the colon, rectum, esophagus and appendix. Extra-gastrointestinal stromal tumors (EGIST) are rare and are found in the omentum, mesentery, retroperitoneum or other intra-abdominal sites [1]. Pancreatic GISTs are extremely rare with a few case reports in the literature. We report the case of an EGIST arising from the pancreas and radiologically mimicking a pancreatic cystic mass that prompts the practitioner to be aware.

CASE REPORT

A 63-year-old woman presented with increasing fatigue and generalized weakness without significant gastrointestinal symptoms. Her past medical history included hypertension and a pancreatic mass. The

pancreatic mass was found by CT scan when she presented to the hospital with flank pain 4 years prior. However, the patient refused surgical treatment at that time and was lost to follow-up. The patient was non-compliant with her recommendations to undergo surgery and have follow up imaging. Surgery was recommended for the resection of a symptomatic presumably solid mass of the pancreas. Physical examination revealed a healthy-appearing woman with tachycardia and soft systolic murmur but otherwise unremarkable. Laboratory data revealed a low hemoglobin of 4.9 g/dL (reference range: 12.0-15.5 g/dL) and a mean corpuscular volume of 86.1 fL (reference range: 78-100 fL). She received 4 units of packed red blood cells. An abdominal CT scan (Figure 1) demonstrated an 11x16 cm cystic mass at the pancreatic body that had increased in size compared with the 6 cm cystic mass from the previous CT scan of 4 years earlier. The cystic mass did not demonstrate a central scar, solid component or calcifications. No clear communication with the pancreatic duct was identified and no evidence of pancreatic calcifications or acute pancreatitis was demonstrated. The cystic mass did not appear to encase any vascular structures on CT. Upper endoscopy was normal. Colonoscopy revealed diverticulosis and a large sigmoid polyp. Endoscopic ultrasound (EUS) demonstrated a large complex cystic structure arising at the pancreatic body characterized by hyperechoic debris within the cyst consistent with a

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Abbreviations EGIST: extra-gastrointestinal stromal tumor

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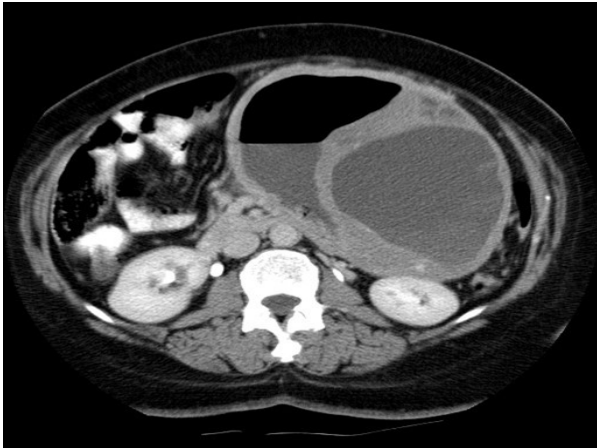


Figure 1. Computed tomography of the abdomen demonstrated an 11x16 cm cystic mass at the pancreatic body.

hemorrhagic cyst (Figure 2). Fine needle aspiration (FNA) was performed and thick bloody fluid was obtained which was found to have an amylase of 20,891 U/L and a cyst fluid CEA of 0.5 ng/mL consistent with a cyst. Fluid cytology revealed scattered spindle cells within the cyst aspirate. Immunohistochemistry studies were positive for muscle specific actin, smooth muscle actin and CD34 but negative for CD117. The patient underwent exploratory laparotomy due to concern for continued hemorrhage. The exploration revealed a cystic mass arising from the pancreatic body. Drainage of the cyst with cystojejunostomy and biopsy of cyst wall was performed. Histologically, the tumor was composed of spindle cells with a mitotic rate less than 5 per 50 high power fields. Immunophenotyping stain was strongly positive for CD34 and CD117 (Figure 3). All of these findings support the diagnosis of low grade pancreatic GIST (based on the mitotic rate but this lesion would be considered high risk due to the size of the lesion) presenting as a hemorrhagic pancreatic cyst. The patient recovered without complication postoperatively and was subsequently referred to oncology for adjuvant therapy with imatinib and for definitive resection of the

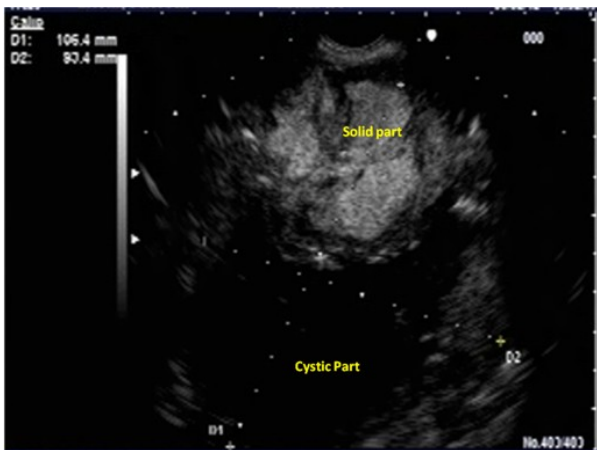


Figure 2. Curved linear array endosonographic image of a complex heterogeneous mass of the retroperitoneum with hyperechoic and cystic component (9x10 cm).

GIST but the patient has again refused definitive resection of the mass.

DISCUSSION

Pancreatic gastrointestinal stromal tumor is an uncommon solid tumor of the pancreas. The primary localization in the pancreas has been rarely reported in only 6 cases [2, 3, 4, 5]. They are usually asymptomatic depending on the location, size of the tumor and lack of mucosal involvement [3]. The clinical symptoms include abdominal pain, early satiety, flatulence, ileus, bleeding, anemia, weight loss. It can be diagnosed incidentally from radiologic imaging [1]. Our patient presented with severe anemia, which could be explained from bleeding inside the pancreatic cyst.

The origin of GIST is from the interstitial cell of Cajal (ICC), which is the pacemaker cell of the GI tract. These cells express the c-kit protein (CD117), CD34 and vimentin [2]. The definite diagnosis is based on the immunohistochemical examination. CD117 expression is the most sensitive marker because it can be found in 95% of GIST. CD34 presents in 75% of GIST and it is mostly expressed in the esophagus and large intestine. There is limited data with regards to predicting the malignant potential of EGIST. Reith *et al.* [6] analyzed

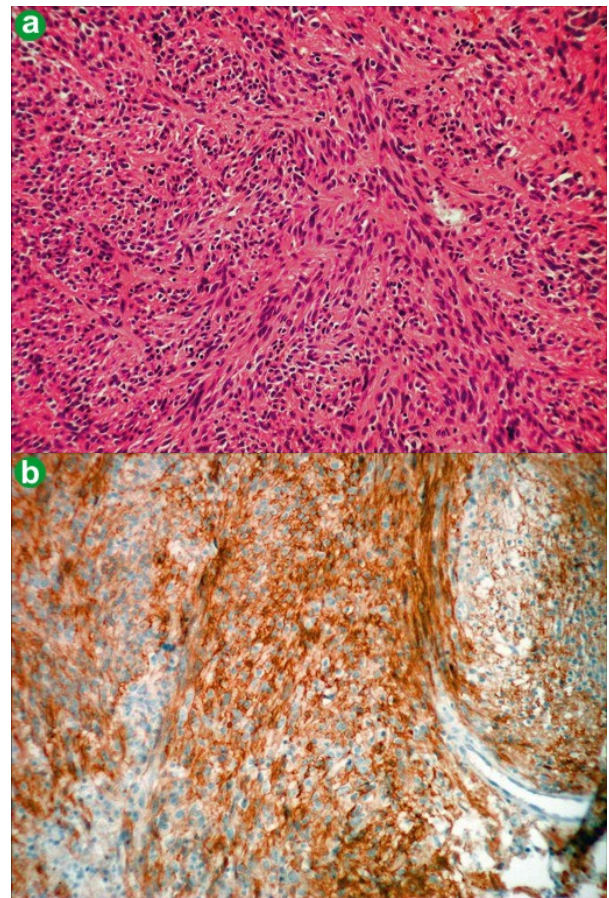


Figure 3. a. H&E stain demonstrated spindle cells with mitotic rate less than 5 per 50 high power fields. **b.** Immunohistochemical analysis results demonstrated the tumor cells are strongly positive for CD117 (c-kit).

48 cases and reported that the clinical malignancy of the tumors increases in the more distal part of GI tract. Other prognostic factors are the tumor size and mitotic rate. Showalter *et al.* [3] reviewed 5 reported cases of pancreatic GIST and found that most of the tumors were in the body and tail of the pancreas. The mitotic rate in most of these was low.

The accuracy of CT imaging to predict a malignant vs. benign cystic lesion of the pancreas ranges between 76% and 82% [7]. The accuracy of CT determination of the histopathological diagnosis of a pancreatic cystic lesion is less than 50% [7]. Endoscopic ultrasound guided FNA plays an important role in the diagnosis of pancreatic cystic lesions. Successful diagnosis of GIST via FNA depends on the adequacy of the specimen. FNA of a solid mass has higher accuracy rate compared to a cystic lesion [5]. In our case, FNA cytology demonstrated a spindle cell lesion but perhaps did not provide sufficient material for immunohistochemistry analysis which may explain the negative CD117 in the FNA but positive CD117 in the surgical biopsy. Multiple passes are often required for adequate fluid analysis. However, EUS with FNA was still helpful in establishing the diagnosis of a hemorrhagic pancreatic cystic lesion and identified the spindle cells within the specimen.

This case demonstrates a rare pancreatic GIST which presented with severe anemia due to cyst formation and hemorrhage within the GIST. A recent report of pancreatic GIST presenting as a solid tumor of the pancreas diagnosed by EUS-FNA was described by Yan *et al.* [4]. To our knowledge, this is the first case report of a pancreatic GIST presenting as a hemorrhagic cyst diagnosed by EUS-FNA. Although these are uncommon, pancreatic GIST should be included in the differential diagnosis of both cystic and

solid masses of the pancreas. Endoscopic ultrasound with fine needle aspiration is a key in the diagnosis of these lesions and can be accomplished with the combination of cytopathology, immunohistochemistry and cyst fluid analysis.

Conflict of interest The authors have no potential conflicts of interest

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