

Groove Pancreatitis: A Malignant Masquerade in the Duodenum

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Abstract

Groove pancreatitis is a rare form of chronic pancreatitis that has been described by several names including para-duodenal wall cyst, pancreatic hamartoma of the duodenum, cystic dystrophy of heterotopic pancreas, and myoadenomatosis. It is characterized microscopically by the presence of a pseudocyst arising from the “groove” between the duodenum, common bile duct, and pancreatic head. It is thought to be caused by functional and/or anatomical minor papilla obstruction from viscous pancreatic secretions. In turn, this leads to impaired pancreatic enzyme outflow, Brunner’s gland proliferation, and resultant pancreatitis. This often appears as a submucosal mass in the region. Thus, the differential diagnosis ranges from benign pathologies such as Brunner’s gland hyperplasia, leiomyoma, and schwannoma to malignant pathologies such as gastrointestinal stromal tumor and ampullary carcinoma. Herein, we present a case of a middle aged male smoker with a remote history of alcohol abuse that presented with one month of epigastric pain associated with bilious emesis and weight loss. Computed tomography (CT) and endoscopic ultrasound (EUS) revealed a submucosal cystic mass that was obstructing the second portion of the duodenum. Cytology demonstrated crowded groups of atypical epithelioid and spindle cells that were suspicious for a neoplasm. The patient subsequently underwent pancreaticoduodenectomy in order to relieve his duodenal obstruction, as well as to establish a definitive tissue diagnosis. Despite the use of multiple imaging modalities including CT and EUS with fine needle aspirates, the final diagnosis of groove pancreatitis was only made following resection. This case now demonstrate that groove pancreatic often mimics submucosal duodenal tumors and malignant periampullary cancers clinically, radiographically, endoscopically, and cytologically.

Keywords: Groove pancreatitis; Cystic dystrophy of heterotopic pancreas; Pancreatic hamartoma of duodenum; Para-duodenal wall cyst; Myoadenomatosis; GIST; gastrointestinal stromal tumor; Brunner’s gland hyperplasia

Introduction

Groove pancreatitis (GP) is a rare form of chronic pancreatitis that has been described by several names including para-duodenal wall cyst, pancreatic hamartoma of the duodenum, cystic dystrophy of heterotopic pancreas, and myoadenomatosis. GP is most common in males aged 40-50-years old with a history of alcoholism and/or smoking that present with abdominal pain, vomiting, and weight loss. Imaging typically reveals duodenal stenosis and cystic lesion(s) near the head of the pancreas [1]. This form of focal, chronic pancreatitis affects the “groove” between the duodenum, common bile duct, and pancreatic head. It is thought to be caused by functional and/or anatomical minor papilla obstruction from viscous pancreatic secretions [2]. Most commonly, this occurs due to alcohol or smoking. In turn, there is impaired pancreatic enzyme outflow, Brunner’s gland proliferation, and resultant pancreatitis [3]. Heterotopic pancreas in the duodenum and peptic ulcer disease are also possible contributing factors. Unlike chronic pancreatitis, there is no known association between groove pancreatitis, autoimmune disease or gallstones [4].

The diagnostic imaging modalities of choice include CT and upper endoscopy with EUS. CT typically reveals a cystic duodenal wall and

local inflammation. Endoscopic imaging may reveal inflamed duodenal mucosa with luminal stenosis. On EUS, cysts are found within the groove and duodenal wall [3,5]. Despite the aforementioned findings, making a tissue diagnosis is often difficult due to the heterogeneity of cells within the duodenal wall and para-duodenal cysts.

Materials and Methods

Report of a case

A 50-year old male smoker with schizoaffective disorder, morbid obesity, Type II diabetes mellitus, and a remote history of alcohol abuse presented with one month of epigastric pain associated with bilious emesis and weight loss. Upon workup he had a total bilirubin of 1.5 mg/dL. Computed tomography (CT) revealed a 7.3×5.9-cm cystic mass that was obstructing the second portion of the duodenum (Figure 1A and B). Upon upper endoscopy he was found to have a near obstructing, submucosal duodenal mass (Figure 1C). Endoscopic ultrasound (EUS) examination demonstrated that this lesion had solid and cystic components. Mucosal biopsies revealed benign duodenal mucosa while cytology from fine needle aspirates displayed foci of spindle cells in the background of intestinal epithelium (Figure 2A). Repeat fine needle aspiration showed foci of somewhat spindle-to-epithelioid cells (Figure 2B). Neuroendocrine-like cells were also present. However, these findings were not diagnostic for malignancy

that may be amenable to preoperative chemotherapy. In order to relieve his biliary and duodenal obstructions, as well as to confirm the diagnosis, he subsequently underwent pancreaticoduodenectomy. Histopathological examination revealed a cystic mass adjacent to fibrotic, atrophic pancreas (Figure 2C) and duodenal mucosa (Figure 2D and E). The pseudocystic changes were seen within the duodenal wall (Figure 2D and E). Even after extensive sampling, no definitive epithelial lining was observed in the large cyst. The cyst was denuded, and cases of paraduodenal wall cysts may indeed be completely denuded. Abundant debris was present within the lumen of the cyst (lower portion of Figure 2D). In Figure 2F, there is myoid spindle cell (e.g., stromal) proliferation at the lower portion of the image, which is most compatible with myofibroblasts and fibroblasts, as well as focal areas containing hemosiderin-laden macrophages noted. Brunner's gland hyperplasia was not identified in this particular case. However, it is notable that Brunner's gland hyperplasia is not one of the features of GP listed in the *Armed Forces Institute of Pathology (AFIP)* criteria. Because the duodenum was quite distorted by the cystic lesion, the minor papilla was not grossly identified, as is classic in GP. However, the lesion does involve, at least partially, the aspect of the duodenum adjacent to the pancreas (Figure 2C), consistent with GP. Immunohistochemical stains for KIT, DOG-1, α -smooth muscle actin, chromogranin A and pan-cytokeratin were negative. Having utilized combined clinical, radiographic, endoscopic, cytological, pathological, and immunohistochemical findings, a final diagnosis of GP was made in our patient.

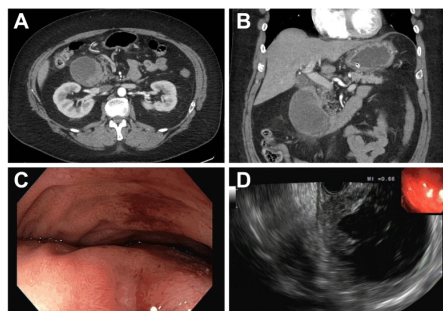


Figure 1: (A and B) CT of the abdomen/pelvis demonstrates a large cystic mass within the duodenal wall. The mass is heterogeneously enhancing with areas suggestive of internal necrosis. A marked mass effect is seen upon the duodenal lumen. (C) Upper endoscopy demonstrates luminal narrowing of the second portion of the duodenum without mucosal involvement. (D) Endoscopic ultrasound demonstrates solid and cystic components in the duodenal wall lesion.

Discussion

The challenge of diagnosing groove pancreatitis preoperatively lies in the ambiguity of imaging findings and cytological variability. Cytology may demonstrate spindle cells, lipid-loaded *macrophages*, and/or debris [6]. In fact, depending upon the area sampled, features may be suggestive of benign pathology such as Brunner's gland hyperplasia (BGH) or submucosal tumors (SMTs), such as gastrointestinal stromal tumor (GIST), leiomyoma, and schwannoma. GIST most often has spindle cell histology, but may also have epithelioid or mixed features. Markers typically include KIT (CD117) and α -smooth muscle actin. A major distinguishing factor between

GIST and other SMTs is that the latter are almost uniformly KIT-negative. However, about one-in-twenty GISTs are also KIT-negative. Thus, the Discovered on GIST-1 (DOG-1) marker can aid in distinguishing these SMTs.

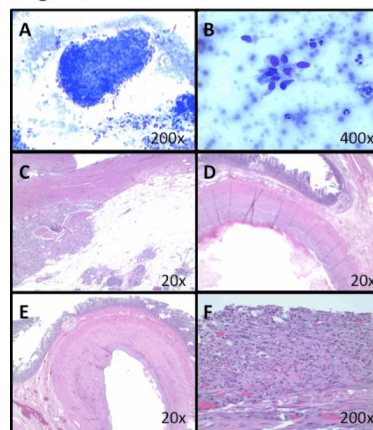


Figure 2: (A) Photomicrograph of cytological specimens demonstrate foci of spindle cells in the background of normal intestinal epithelium (Diff-Quick, 200x). (B) Repeat aspirates demonstrate foci of spindle-to-epithelioid cells (Diff-Quick, 400x). (C) Photomicrograph of the resected specimen reveals cyst wall adjacent to the pancreas, which has evidence of fibrosis and chronic pancreatitis (H&E, 20x). (D and E) Photomicrographs of the resected specimen reveal a cystic mass adjacent to normal duodenal mucosa (H&E, 20x). (F) Photomicrographs of myoid spindle cell (e.g., stromal) proliferation at the lower portion of the image, which is most compatible with myofibroblasts and fibroblasts, as well as focal areas containing hemosiderin-laden macrophages noted (H&E, 200x).

The presence of Brunner's glands on biopsies can lead to misdiagnosing BGH. This can present in a similar fashion to GP because there are excessive Brunner's glands. However, BGH is thought to be caused by hyperactivity of the exocrine modulating factors including the vagus nerve and intestinal mucous membrane factor [7], as well as gastric acid hypersecretion and decreased pancreatic enzyme secretion [2]. Endoscopically, BGH presents with nodular, rather than cystic, nodules in the duodenal submucosa [8]. While Brunner's gland hyperplasia is often asymptomatic, no consensus exists regarding resection (i.e., pancreaticoduodenectomy), except when patients develop severe symptomatology, such as abdominal pain or hemorrhage.

In cases of GP secondary to alcohol, treatment is conservative management; nil per os, analgesia, parenteral nutrition, and alcohol cessation. In patients with GP secondary to anatomic abnormalities or those refractory to conservative management, pancreaticoduodenectomy is recommended with approximately 75% of patients having complete symptom relief postoperatively [3].

Conclusion

In summary, GP is a rare disorder that often mimics submucosal duodenal tumors and malignant periampullary tumors that present with obstructive jaundice. Surgical therapy remains the gold standard

for treatment of this disease when symptoms and obstruction are present. Since resection, our patient is tolerating a diet, and his weight is stable.

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