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CASE REPORT | PANCREAS

Sometimes You Have to Judge a Book by Its Cover: The Case of a Masquerading Pancreatic Mucinous Cyst

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ABSTRACT

Pancreatic cystic lesions are difficult to evaluate amid acute pancreatitis. Without previous pancreatic imaging, it is challenging to discern between pancreatic acute fluid collections and cystic neoplasms. We present a 29-year-old woman with acute pancreatitis and initial cross-sectional imaging suggesting a 2.8-cm cystic lesion in the body/tail of the pancreas. Endoscopic ultrasound completed 5 weeks after index presentation revealed findings worrisome for a cystic neoplasm, but fine-needle aspiration findings suggested lesion to be a pseudocyst (normal carcinoembryonic antigen and cytology, negative mucin stain).

INTRODUCTION

The clinical history and timing of pancreatitis are quintessential pieces of information when assessing pancreatic cystic lesions and deciding further management.^{1–3} We present the case of a young woman with index presentation of acute pancreatitis and a cystic lesion in the tail of pancreas that was found on index presentation. Without previous imaging preceding the pancreatitis episode, the question of acute pancreatic fluid collection vs cystic lesion provoking pancreatitis becomes central. This case demonstrates endosonographic (EUS) findings concerning for cystic neoplasm contrasting with fine-needle aspiration (FNA) findings suggesting a pseudocyst. Surgical resection and pathology ultimately revealed a mucinous cyst with pancreatic duct (PD) communication to be the cause of acute pancreatitis.

CASE REPORT

A 29-year-old woman developed acute symptoms of epigastric abdominal pain with radiation to the back that progressively worsened over a few hours, prompting emergency department presentation. On initial presentation, she was afebrile with normal vital signs. She was found to have normal liver enzymes with a lipase of 2,164 U/L. There were no gallstones on the abdominal ultrasound examination. She reported 1–2 servings of beer the night before presentation. She denied previous abdominal complaints. Abdominal and pelvic computed tomography completed on emergency department presentation revealed a 2.8-cm fluid attenuating structure in the body-tail region of the pancreas without ductal dilatation (Figure 1). A magnetic resonance imaging was completed on hospital day 2 and demonstrated the simple-appearing cystic lesion in the pancreatic body-tail region suggestive of a pseudocyst or cystic neoplasm (Figure 2).

She was managed supportively and discharged home within 3 days of presentation. At the follow-up, EUS was recommended to further characterize cystic lesion. The patient's alcohol use before pancreatitis episode did not seem to be the cause of her pancreatitis. In addition, IgG 4, serum triglycerides, and Ca 19-9 were normal, and no imaging modality had suggested cholelithiasis. The leading differential diagnosis was cystic neoplasm causing pancreatitis vs acute pancreatic fluid collection with progression to pseudocyst.

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Figure 1. Computed tomography on index presentation of acute pancreatitis showing 2.8-cm fluid attenuating lesion in the pancreatic body-tail without dilation of the pancreatic duct (arrow). Differential of this lesion included pancreatic cystic neoplasm such as intraductal papillary mucinous neoplasm and a pseudocyst. Subtle peripancreatic fat stranding also seen.

EUS was completed 5 weeks after the index pancreatitis episode. In the tail of the pancreas, an anechoic pancreatic cyst was confirmed, measuring about 21×17 mm with a large hypoechoic mural nodule (Figure 3). The main PD appeared to be in direct communication with the cyst both downstream and upstream from the cyst. Upstream from the cyst, there was lobularity and stranding to suggest focal inflammatory changes in the tail. However, PD dilation was not seen in the tail of the pancreas. The ampulla was normal; there was no mucin extruding from ampullary orifice. There was no evidence of pancreas divisum, choledocholithiasis, or gall stones. Given cystic findings of large mural nodule and PD communication, the decision was made to proceed with FNA. FNA was performed of the pancreatic tail cyst using a 25-gauge FNA needle

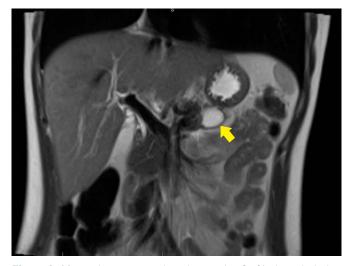


Figure 2. Magnetic resonance imaging on day 2 of index admission for acute pancreatitis showing 2.6×2.8 -cm simple appearing cyst with a thin enhancing wall within the pancreatic body-tail with slight prominence distal to the lesion (arrow). The cyst appeared to communicate directly with the pancreatic duct. Mild edema was seen adjacent to and distal to the cystic lesion in the body of the pancreas.



Figure 3. Linear ultrasound of the pancreatic tail showing 21×17 -mm anechoic cyst with thin wall and a large hypoechoic mural nodule in the tail of the pancreas.

yielded serous, yellow tinged fluid. The mural nodule was mobile and proved challenging to sample directly. Unfortunately, the string test to stratify mucin presence within the aspirate was not performed. The fluid analysis demonstrated amylase of >30,000 U/L and carcinoembryonic antigen (CEA) of 32 U ng/mL. Cytology evaluation showed proteinaceous material and no intracellular or thick extracellular mucin.

Despite relatively reassuring fluid analysis, suggestive of a pseudocyst, the endosonographic findings of large mural nodule raised concern for intraductal papillary mucinous neoplasms or mucinous cyst and surgical evaluation was obtained. Surgical resection was recommended given the collective features of cyst size >2 cm, presentation with pancreatitis, and presence of a large mural nodule. Robotic distal pancreatectomy with possible splenectomy was pursued.

During robotic distal pancreatectomy, patient underwent intraoperative ultrasound. A largely anechoic spherical lesion

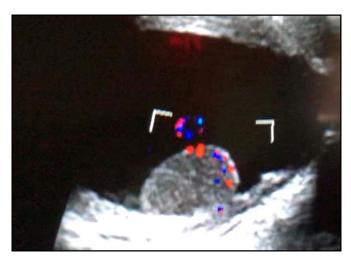


Figure 4. Intraoperative ultrasound imaging showing a spherical lesion was identified with a roughly 1.1-cm mural nodule that did not have the appearance of debris and Doppler ultrasound suggested internal blood flow within nodule.



Figure 5. The distal pancreatectomy specimen demonstrated a 2.7-cm unilocular cyst (black arrow) which appeared to grossly communicate with the main pancreatic duct (yellow arrow).

was seen with a large (greater than 1 cm) mural nodule with internal blood flow seen (Figure 4). A transection margin 2 cm toward the head of pancreas was made, and the entire pancreatic tail was harvested out of splenic hilum with lymph node harvest. The resected specimen and apparent communication of the cyst with PD is seen (Figure 5). Histology demonstrated mucinous cystic neoplasm with ovarian stroma, low-grade dysplasia noted with margins negative for mucinous cystic neoplasm (Figure 6). Features of chronic pancreatitis were seen in the tail of the pancreas. The patient tolerated the procedure well and was discharged postoperative day 3. She has not had any further episodes of recurrent acute pancreatitis in the 5 months since her surgery and is doing well clinically.

DISCUSSION

The central question, in this case, was the temporal/causal relationship between the cyst and the occurrence of acute pancreatitis. Therefore, the importance of the index cross-sectional imaging study cannot be overstated. The presence of a cystic lesion on imaging during the index admission should and did raise concern for neoplastic origin over a pancreatic fluid collection in the differential diagnosis. In addition, the collective high-risk features on imaging with suspected pancreatitis related to the cyst warranted surgical resection in this young patient; explicitly, the presence of a mural nodule greater than or equal to 5 mm. Furthermore, the American Gastroenterological Association and the American College of Gastroenterology guidelines are in agreement with international consensus Fukuoka guidelines, recommending surgical management in cases where the pancreatic cyst has a solid component and association with a dilated PD. Section 1.5.

EUS and cross-sectional studies suggested PD communication with the cyst. Traditionally, mucinous cystic neoplasms (MCNs) do not feature communication with the PD. In a large multicenter study from Japan in 2011 of resected MCNs (n=156), preoperative pancreatogram demonstrated communication of PD

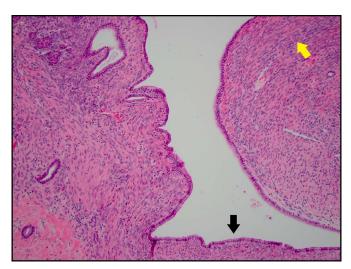


Figure 6. Mucinous cystic neoplasm with low-grade dysplasia with margins negative for mucinous cystic neoplasm. Black arrow: columnar mucinous epithelium lining. Progesterone and estrogen receptor immunostain positivity was seen in underlying spindled ovarian-type stroma surrounding the cyst (yellow arrow) (hematoxylin and eosin stain).

and cyst in 18.1% of cases. It is unclear whether these findings were related to erosion of the cyst wall into the duct rather than true origin from the PD.6 FNA fluid analysis in this case was not suggestive of MCN and demonstrates limitations in FNA in the preoperative evaluation of pancreatic cysts. In a case series of resected pancreatic cysts (n = 32), the sensitivity of FNA (positive cytology and/or CEA >192 ng/mL) was 61%.7 Hence, negative cytology and low CEA are insufficient in and of themselves to remove MCN from the differential diagnosis of a pancreatic cystic lesion. In this case, a high index of suspicion with careful attention to clinical timeline and imaging led to the optimal management and outcome for this young patient. This case also highlights the need for follow-up imaging when no trigger is identified for an acute pancreatitis presentation, such as biliary stones, alcohol, autoimmune, medications, hypertriglyceridemia, or trauma. This case exemplifies some of the complexities in assessing cystic lesions of the pancreas that are best addressed by a multidisciplinary model of assessment within high-volume centers.

DISCLOSURES

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