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

John Miller Jr, MD1, Diana Agostini-Vulaj, DO2, Victoria Howard, PA1, Luke Schoeniger, MD, PhD, FACS3, and Truptesh Kothari, MD, MS, FACP, FASGE1

1Division of Gastroenterology and Hepatology, Department of Medicine, Strong Memorial Hospital, University of Rochester Medical Center, Rochester, NY
2Department of Pathology and Laboratory Medicine, Strong Memorial Hospital, University of Rochester Medical Center, Rochester, NY
3Division of Surgical Oncology, Department of Surgery, Strong Memorial Hospital, University of Rochester Medical Center, Rochester, NY

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.
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 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 neoplasm 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
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.4 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.3,5

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 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

Author contributions: All authors contributed equally to this manuscript. J. Miller is the article guarantor.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received April 23, 2020; Accepted October 4, 2020

REFERENCES

1. Siddiqui UD. Pancreatic cysts—Making sense of differing guidelines. 2016 ACG Postgraduate Course, Las Vegas; 2016.
2. Moran R, Lennon AM. Chapter 15: Endoscopic ultrasonography in the evaluation of pancreatic cysts. In: Hawes R, Fockens P, Varadarajulu S (eds). *Endosonography*. 4th edn. Elsevier: Philadelphia, PA, 2019, pp 185–96.

3. Elta GH, Enestvedt BK, Sauer BG, Lennon AM. ACG clinical guideline: Diagnosis and management of pancreatic cysts. *Am J Gastroenterol* 2018;113(4):464–79.

4. Tanaka M, Fernandez-Del Castillo C, Kamisawa T, et al. Revisions of international consensus fukuoka guidelines for the management of IPMN of the pancreas. *Panreatology* 2017;17(5):738–53.

5. Vege SS, Ziring B, Jain R, Moayyedi P; Clinical Guidelines Committee; American Gastroenterology Association. American gastroenterological association institute guideline on the diagnosis and management of asymptomatic neoplastic pancreatic cysts. *Gastroenterology* 2015;148(4):819–22.

6. Yamao K, Yanagisawa A, Takahashi K, et al. Clinicopathological features and prognosis of mucinous cystic neoplasm with ovarian-type stroma: A multi-institutional study of the Japan pancreas society. *Pancreas* 2011;40(1):67–71.

7. de Jong K, van Hoof JF, Nio CY, et al. Accuracy of preoperative workup in a prospective series of surgically resected cystic pancreatic lesions. *Scand J Gastroenterol* 2012;47(8–9):1056–63.

Copyright: © 2021 The Author(s). Published by Wolters Kluwer Health, Inc. on behalf of The American College of Gastroenterology. This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.