ACELLULAR IS NOT ALWAYS BENIGN: A CASE OF PRIMARY PANCREATIC SARCOMA

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1. Introduction

Pancreatic Cystic Neoplasms (PCN) are encountered more frequently given the more widespread usage of cross sectional imaging. Primary pancreatic sarcomas represent 0.1% of all pancreatic malignancies and tend to be aggressive and have a poor prognosis.

2. Case Description

A 74 yo F w/ PMHx significant for HLD presented to ER with persistent nausea and vomiting of two days duration. She became concerned when she was unable to tolerate oral diet and decided to seek medical attention. There was no abdominal pain. Her only medication was atorvastatin. No family history of malignancy. Social history yielded occasional alcohol use (1-2/year) and no smoking. She was independent and ROS was unremarkable. Her PE was unremarkable with no abdominal tenderness. CXR and CBC were unremarkable. CMP was significant for a mildly elevated bilirubin 1.3. Given the fact that she was unable to tolerate PO she was admitted for further evaluation and supportive management. Due to unresolving symptoms she underwent a CT scan with IV contrast. It was significant for a 10.4 x 9.1 x 7.2cm cystic lesion centered in the root of the jejunal mesentery. The pancreatic body and tail were displaced and the main pancreatic duct appeared normal. The origin was unclear. MRI with and without contrast could not delineate the origin of the cyst either. AFP and CEA were unremarkable. EUS was undertaken and was significant for a large 10cmx8cm complex cyst in the pancreatic body. The cyst was heterogenous and had a solid component (90%). The bile ducts and pancreatic ducts were normal in caliber. There was no lymphadenopathy. A 22g needle was used to aspirate 1.5mL of fluid. Grossly the fluid was thin and serous appearing. Intraoperative microscopic review showed an acellular fluid with blood only. The patient improved with supportive management and was discharged home. Pathology subsequently showed high grade spindle cells positive for MDM2 with patchy SMA staining. They were negative for S100, SOX10, and CD117. This is consistent with a high-grade sarcoma however the specimen has been sent to a tertiary care center for a second opinion.

The patient is currently awaiting oncology input but is systemically well

References

1. **Ambe *et al* Primary Sarcoma of the Pancreas, A Rare Histopathological Entity: A Case Report and review of literature. World J of Surg Oncology 2011.**
2. **Khalid *et al* Classification of Pancreatic Cysts. UptoDate.com**

4. Endoscopic Ultrasound Findings Findings and Pathology

Figure 1: Endoscopic Finding

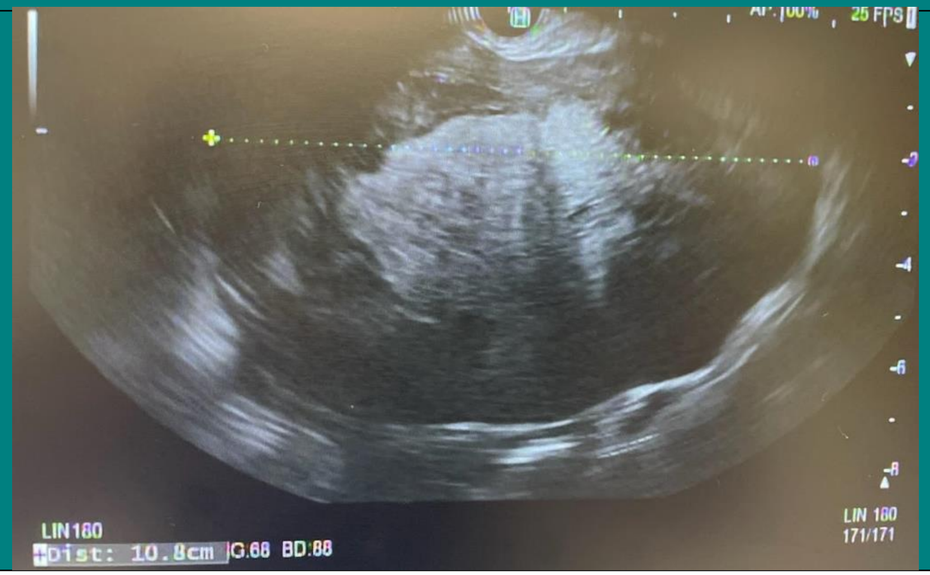

Figure 2: Primary Sarcoma of the Pancreas[1]

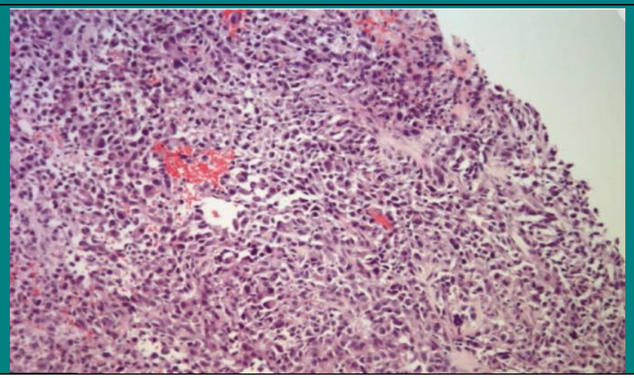

Figure 3: Key Features of Typical Pancreatic Cystic Neoplasms[2]

Key demographic and clinical features of patients with pancreatic cystic neoplasms[1-4]

	Serous cystic tumor	Mucinous neoplasm	Main-duct intraductal papillary mucinous neoplasm	Branch-duct intraductal papillary mucinous neoplasm	Solid pseudopapillary neoplasm
Age of presentation	Variable, usually 5th to 7th decade	Variable, usually 5th to 7th decade	Variable, usually 5th to 7th decade	Variable, usually 5th to 7th decade	Usually 2nd to 3rd decade
Gender distribution	Females >males	Almost exclusively females	Females = males	Females = males	Females >males
Typical clinical presentation	Incidental or abdominal pain or mass effect	Incidental or abdominal pain or malignancy related	Incidental or pancreatitis or pancreatic insufficiency or malignancy related	Incidental or pancreatitis or malignancy related	Incidental or abdominal pain or mass effect
Typical imaging characteristics	Microcystic/honeycomb appearance Oligocystic appearance less common	Unilocular or septated cyst ± wall calcifications Solid component, if present, may suggest malignancy	Dilated main pancreatic duct ± parenchymal atrophy Solid component, if present, may suggest malignancy	Dilated pancreatic duct branch or branches Solid component, if present, may suggest malignancy	Solid and cystic mass ± calcifications
Typical aspirate characteristic	Thin, often bloody	Viscous	Viscous	Viscous or thin	Bloody
Typical cytology findings	Cuboidal cells that stain positive for glycogen; yield <50%	Columnar cells with variable atypia Stains positive for mucin; yield <50% High yield from solid component for malignancy	Columnar cells with variable atypia Stains positive for mucin; yield <50% High yield from solid component for malignancy	Columnar cells with variable atypia Stains positive for mucin; yield <50% High yield from solid component for malignancy	Characteristic branching papillae with myxoid stroma High yield from solid component
Typical carcinoembryonic antigen (CEA) level	<5 to 20 ng/mL in majority of lesions	>200 ng/mL in approximately 75% of lesions	>200 ng/mL in approximately 75% of lesions	>200 ng/mL in approximately 75% of lesions	Insufficient data
Typical glucose level	>50 mg/dL in majority	<50 mg/dL in majority	<50 mg/dL (limited data)	<50 mg/dL in majority	Insufficient data
Typical DNA analysis	Allelic loss affecting chromosome 3p and VHL mutation specific	K-ras mutation specific (>90%), not sensitive (<50%) TP53, PTEN, PIK3CA, high DNA amount or high-amplitude allelic loss seen in malignancy	K-ras and GNAS mutation specific (>90%), not sensitive (<50%) TP53, PTEN, PIK3CA, high DNA amount or high-amplitude allelic loss seen in malignancy	K-ras and GNAS mutation specific (>90%), not sensitive (<50%) TP53, PTEN, PIK3CA, high DNA amount or high-amplitude allelic loss seen in malignancy	CTNNB1 mutation specific
Relative malignant potential	Negligible	Moderate	High	Low to moderate	Moderate to high
Treatment	Resect if symptomatic	Resection	Resection and post-resection surveillance	Closely monitor or resect Post-resection surveillance required	Resection

References:
1. Khalid A, Brugge WR. ACG practice guidelines for the diagnosis and management of neoplastic pancreatic cysts. Am J Gastroenterol 2007; 102:2339.
2. Wu J, Jiao Y, Dal Molin M, et al. Whole-exome sequencing of neoplastic cysts of the pancreas reveals recurrent mutations in components of ubiquitin-dependent pathways. Proc Natl Acad Sci U S A 2011; 108:21188.
3. Singhi AD, McGrath K, Brand RE, et al. Preoperative next-generation sequencing of pancreatic cyst fluid is highly accurate in cyst classification and detection of advanced neoplasia. Gut 2018; 67:2131.
4. McCarty TR, Garg R, Rustagi T. Pancreatic cyst fluid glucose in differentiating mucinous from nonmucinous pancreatic cysts: a systematic review and meta-analysis. Gastrointest Endosc 2021; 94:698.

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5. CONCLUSIONS

It is of vital importance that large cystic lesions in close proximity to the pancreas be evaluated urgently with EUS. While intraoperative FNA results are helpful, what can appear benign can in fact be a highly aggressive tumor. Primary pancreatic sarcomas are a rare but important consideration of PCN.