Cystic Pancreatic Neuroendocrine Tumor

Presented By:

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A 62-year-old female presented to the emergency department after worsening episodes of nausea and sharp abdominal pain with radiation to her back.

Contrast-enhanced CT in the coronal (A) and axial (B) planes in the arterial phase and axial plane in the venous phase (C) shows a 1.1 cm unilocular cystic mass in the body of the pancreas (arrowheads) with a thin rim of peripheral enhancement. There are no solid or nodular enhancing components. The surrounding pancreatic parenchyma appears normal without dilation of the main pancreatic duct, parenchymal atrophy, or inflammation.
Axial MRI shows that the mass (arrowheads) is hypointense on the in-phase T1-weighted image (A), hyperintense on both the T2-weighted fat saturated image (B) and diffusion-weighted image (C), and shows a thin rim of peripheral enhancement on the contrast-enhanced T1-weighted fat saturated image in the arterial phase (D). The mass does not communicate with the normal caliber main pancreatic duct on the 3D MIP MRCP image (E). Subsequent axial $^{68}$Ga-DOTATATE PET (F) shows radiotracer uptake within the mass, confirming expression of type 2 somatostatin receptors. Pathology after resection showed a grade 1 neuroendocrine tumor with $<2$ mitoses per mm$^2$ and a Ki-67 index of $<2\%$. 
Discussion

Cystic pancreatic neuroendocrine tumors (pNETs) account for 6.5-36.1% of all resected pNETs. Once considered rare, the ubiquitous use of cross-sectional imaging has contributed to a rise in their detection over the past few decades.

The etiology, clinicopathologic features, and prognosis of cystic pNETs has been controversial. They were originally thought to be the result of tumoral necrosis, intratumoral hemorrhage, and/or cystic degeneration of a solid tumor and to behave similarly to their solid counterparts.

More recent reports suggest that cystic pNETs are a distinct subtype of pNET with more indolent biologic behavior and a better long-term prognosis than their solid counterparts. Compared to solid pNETs, cystic pNETs are less likely to be functional, are less likely to metastasize, have less microvascular and perineural invasion, have lower Ki-67 indices and mitotic counts, are less likely to be aggressive tumors, and have higher 5- and 10-year disease-free survival rates.
Radiographically, cystic pNETs can present as either partially or entirely cystic masses and are more common in the pancreatic body and tail. Partially cystic pNETs can have internal septations of varying thickness. The solid components will usually show hyperenhancement on arterial and venous phases that is typical of entirely solid pNETs.

Entirely cystic pNETs may be mistaken for other macrocystic pancreatic masses such as mucinous cystic neoplasms (MCNs), intraductal papillary mucinous neoplasms (IPMNs), or oligocystic serous cystadenomas. The key distinguishing feature of an entirely cystic pNET at imaging is a peripheral hypervascular rim, which may necessitate a dedicated pancreatic protocol to detect and is more commonly seen on the arterial phase of enhancement. Cystic pNETs should not communicate with the main pancreatic duct and do not typically cause ductal dilation, which can help differentiate them from IPMNs.

Since radiographic features may not reliably differentiate macrocystic pancreatic masses, endoscopic ultrasound-guided fine needle aspiration biopsy is usually required for a definitive diagnosis. $^{68}$Ga-DOTATATE PET/CT can also be used for confirmation.
Key Points

• Cystic pNETs are not rare and are a distinctive subtype of pNET with unique clinical and pathologic features

• Cystic pNETs have more indolent biologic behavior and a better long-term prognosis than their solid counterparts

• A peripheral hypervascular rim can help differentiate entirely cystic pNETs from other macrocystic pancreatic masses at imaging
References and Suggested Reading


