Rare Tumors and Lesions of the Pancreas

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KEYWORDS

• Pancreatectomy • Pancreatic neoplasm • Anaplastic carcinoma
• Adenosquamous carcinoma • Solid pseudopapillary tumor • Acinar cell carcinoma
• Primary pancreatic lymphoma • Unusual pancreas tumors

KEY POINTS

- Rare pancreatic tumors of the pancreas include adenocarcinoma variants, such as anaplastic carcinoma, adenosquamous carcinoma, colloid, hepatoid, and medullary carcinoma.
- Other neoplasms include acinar cell carcinoma, solid pseudopapillary tumor, sarcomas, or lymphomas.
- Benign solid or cystic masses, such as hamartoma, hemangioma, lymphangioma, or others also may mimic neoplastic disease.
- The pancreas may be the site of isolated metastatic disease, such as renal cell cancer, colorectal cancer, melanoma, and other carcinomas.
- Pancreatic inflammatory diseases may mimic solid neoplasms of the pancreas.

Primary pancreatic ductal adenocarcinoma (PDAC) is the most common neoplasm of the pancreas. Pancreatic neuroendocrine tumors (PNETs) are much less common but their incidence has increased over the past decade due to the increased use of cross-sectional imaging.[1] Cystic lesions, such as intraductal papillary mucinous neoplasm (IPMN), mucinous cystic neoplasms (MCN), and serous cystic neoplasms (SCN) are also relatively common. The pancreas is a complex organ that harbors a wide array of diseases. There are a variety of non-neoplastic conditions that mimic PDAC, such as groove pancreatitis (GP) and autoimmune pancreatitis (AIP).[2,3] Additionally, there are a handful of other rare neoplastic lesions infrequently found in patients with pancreatic masses that range from well known (eg, solid pseudopapillary neoplasm and acinar cell carcinoma) to less well known (eg, leiomyosarcoma and hepatoid carcinoma). Rare cystic lesions can be misdiagnosed for the more common

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mucinous, serous, or inflammatory pancreatic lesions. Peripancreatic solid lesions or duodenal pathology occasionally can be mistaken for pancreatic pathology as well. Finally, the pancreas is a potential site for metastatic disease, such as renal cell carcinoma (RCC), or can be involved with other diseases, such as primary pancreatic lymphoma. Any of these lesions can be mistaken for PDAC.

Contrast-enhanced computerized tomography (CT) is the most common modality to detect and diagnose pancreatic pathology. Endoscopic ultrasound (EUS) and MRI with cholangiopancreatography (MRCP) have distinct advantages in hepatopancreatobiliary imaging that may clarify the diagnosis of an unknown pancreatic mass. EUS allows gastroduodenal mucosal evaluation, and has perhaps the highest sensitivity for small lesions of the pancreas (eg, subcentimeter PNET and mural nodules within IPMN), as well as enabling direct tissue sampling with fine-needle aspiration. High-quality MRI with MRCP provides detailed anatomic information of the pancreas and ductal structures that cannot be obtained with any other modality. Additionally, with an appropriate protocol, the MRI can provide conspicuity of any liver lesions associated with pancreatic disease surpass the images of the highest-quality triple-phase CT.[4,5] Complete evaluation by an experienced team is warranted for all with pancreatic neoplasms. A multidisciplinary approach with knowledgeable surgeons, gastroenterologists, radiologists, oncologists, pathologists, and others limit misdiagnosis and/or mismanagement of the following rare pancreatic findings.

ADENOCARCINOMA VARIANTS

Ductal adenocarcinoma of the pancreas with tubular morphology accounts for more than 90% of pancreatic carcinoma. There are variants of adenocarcinoma with a different prognosis that should be distinguished from PDAC.

Anaplastic (Undifferentiated) Adenocarcinoma (Also with Osteoclastlike Giant Cells)

Anaplastic pancreatic carcinomas (APCs) are rare neoplasms that represent 2% to 7% of all exocrine pancreatic tumors. First described by Sommer and Meissner in 1954[6] they are referred to as undifferentiated carcinoma with or without osteoclastlike giant cells, carcinosarcoma, sarcomatoid carcinoma, pleomorphic carcinoma, pleomorphic giant-cell carcinoma, and pleomorphic large-cell carcinoma of the pancreas. This undifferentiated carcinoma is an aggressive epithelial neoplasm that does not display significant components of differentiated lesions. Anaplastic foci can be seen within PDAC but as a minor component. The male-to-female ratio is 3 to 1 and generally affects older men (**Fig. 1**). The lesions are distributed throughout the pancreas and are often quite large when diagnosed (average of up to 9–10 cm).[7,8] The literature focuses on histology, immunohistochemistry, electron microscopy, and gene expression of APCs. Many studies note a poor outcome after resection due to its systemic nature, but other larger studies have shown benefit to resection.[7,9] Hoshimoto and colleagues[10] reported 60 cases of APC resected in Japan with a mean age at diagnosis of 61.5 years, 63% male, and a median size of 6 cm. Nearly one-fourth required resection of adjacent organs and vascular involvement was present in (12%). Although half died within 1 year of surgery, the 5-year survival rate was 12%.[10] Strobel and colleagues[11] reported a single institutional experience of 18 patients with APC who underwent attempted resection and compared them to a similar group with PDAC. They noted a median survival of only 5.7 months, but a margin-negative resection extended survival and 17% were long-term survivors.[11] Paniccia and colleagues[12] matched 192 patients with APC from the National Cancer Data Base with 960 PDAC patients. They too noted a 1-year survival that was lower than PDAC with a similar overall long-term survival.