Rare Pancreatic Tumors

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KEYWORDS

• Pancreatic cancer • Rare pancreatic tumors • Rare pancreatic lesions • MR imaging
• Computed tomography

KEY POINTS

- Pancreatic ductal adenocarcinoma is the most common pancreatic malignancy and represents greater than 90% of all pancreatic neoplasms.
- Magnetic resonance (MR) and computed tomography (CT) imaging provide complementary high contrast, spatial, and temporal resolution for imaging and characterization of pancreatic tumors.
- The absence of classic imaging features of pancreatic ductal adenocarcinoma, such as hypoenhancement, ductal dilation, and distal pancreatic atrophy, should raise consideration for a nonductal tumor.
- Tissue sampling is often required to make a definitive diagnosis, but MR and CT imaging can potentially narrow the differential diagnosis and guide management.

INTRODUCTION

Pancreatic ductal adenocarcinoma (PDAC) (90%) and neuroendocrine tumors (5%) comprise most malignant pancreatic neoplasms.[1] A variety of cystic neoplasms of the pancreas, ranging from intraductal papillary mucinous neoplasms to serous and mucinous cystadenomas to solid pseudopapillary neoplasms, are also well known to most radiologists. However, a variety of rare benign and malignant pancreatic tumors may be encountered that radiologists are unfamiliar with. An important consideration in evaluating solid pancreatic lesions is an appearance atypical of PDAC, such as the lack of pancreatic ductal dilation or distal atrophy, that might lead the radiologist to consider a rare tumor. Pancreatic neoplasms may be of exocrine, endocrine, mesenchymal, or extrapancreatic origin; imaging features of the rare pancreatic tumors are often nonspecific. Knowledge of the patients' history for clues to guide the radiologist is key, such as a history of neurofibromatosis type I or a primary renal cell carcinoma (RCC) or melanoma; but in many cases tissue sampling will be required to confirm the diagnosis. An MR imaging protocol for comprehensive evaluation of the pancreas is described in **Table 1**. This article reviews a variety of rare pancreatic tumors (**Table 2**) with magnetic resonance (MR) and computed tomography (CT) case examples.

ACINAR CELL CARCINOMA

The exocrine pancreas is composed primarily of ductal cells, the precursors for PDAC, and acinar cells, which secrete enzymes such as amylase and lipase.[2] Carcinomas arising from acinar cells, also known as acinic cell carcinomas (ACCs), represent less than 1% of adult pancreatic neoplasms but 15% of pediatric pancreatic exocrine neoplasms.[3] These tumors have a male predominance in the sixth and seventh decades of life.[4]

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Table 1
Magnetic resonance protocol

Sequence	TR (ms)	Base TE (ms)	FOV (mm)	Matrix	Slice Thickness (mm)
T2W coronal SSTSE	1300	90	400 × 400	640 × 640	6 mm
T2W coronal SSTSE FS (thin slice)	1000	84	320 × 350	320 × 300	4 mm
T2W axial SSTSE (thin slice)	1000	84	320 × 260	260 × 320	4 mm
T2W sagittal SSTSE (thin slice)	1000	84	284 × 350	320 × 260	4 mm
T2W coronal/oblique thick slab MRCP × 3	4500	750	300 × 300	384 × 348	70 mm
T1W axial GRE in and out of phase	112	2.38	320 × 260	520 × 640	7 mm
EPI axial DWI (b = 50, 400, 800)	5800	65	380 × 285	144 × 192	6 mm
PD axial GRE multi-echo Dixon	122	2.38	320 × 320	192 × 192	10 mm
T1W 3D FS spoiled GRE axial	4.62	2.3	340 × 255	480 × 640	3.2 mm
Administer 0.1 mmol/kg intravenous gadolinium contrast agent at 2 mL/s, with bolus tracking on the abdominal aorta					
T1W 3D FS spoiled GRE axial × 3	4.62	2.3	340 × 255	480 × 640	3.2 mm
T1W 3D FS spoiled GRE coronal	4.62	2.3	330 × 380	320 × 280	3 mm
T1W 3D FS spoiled GRE axial 5 min	4.62	2.3	340 × 255	480 × 640	3.2 mm
T2W 3D TSE free breathing MRCP	4084	703	380 × 380	384 × 384	1 mm

Abbreviations: 3D, 3 dimensional; DWI, diffusion-weighted imaging; EPI, echo planar imaging; FOV, field of view; FS, fat suppressed; GRE, gradient echo; MRCP, magnetic resonance cholangiopancreatography; PD, proton density; SSTSE, single-shot turbo spin echo; TE, echo time; T1W, T1 weighted; TR, repetition time; T2W, T2 weighted.

Table 2
Rare pancreatic tumors

Pancreatic Tumor	Key Features
Acinar cell carcinoma	Exocrine malignancy arising from acinar cells; usually well-circumscribed, large, hypoenhancing on early postcontrast phases, less likely to see ductal dilation than with ductal adenocarcinoma
Hepatoid carcinoma	Extrahepatic malignancy resembling hepatocellular carcinoma with arterial phase hyperenhancement and delayed washout
Nerve sheath tumors	Schwannomas may be cellular and solidly enhancing or hyaline/myxoid with a cystic appearance; neurofibromas have characteristic T2 target appearance and if plexiform may resemble a bag of worms
Plasma cell tumors	Multiple myeloma or solitary plasmacytoma may be infiltrative or masslike and may be T1 hyperintense from proteinaceous content
Leiomyoma	Benign smooth muscle tumor, well circumscribed with intermediate T2 hypointensity and delayed enhancement
Lipoma	Benign fatty tumor following fat intensity on all sequences
Metastatic disease	Renal cell and melanoma notoriously metastasize to the pancreas but also from common cancers (lung, breast, colon)
Lymphangioma	Benign lymphatic tumor, circumscribed cystic mass with thin septations; may not be distinguishable from more common cystic lesions by imaging
Dermoid	Mature cystic teratoma; imaging appearance varies depending on composition between fat, calcification, and soft tissue
Lymphoma	Usually secondary, may present as diffuse or segmental masslike enlargement, frequently without pancreatic ductal dilation or atrophy
Sarcoma	Variable imaging appearance depending on histology, frequently locally aggressive and prone to metastasize to liver and lymph nodes
Pancreatoblastoma	Most common malignant pancreatic tumor in young children; circumscribed, lobulated heterogeneous mass with peripheral calcification, central hemorrhage/necrosis, and mild enhancement