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Imaging features of rare pancreatic tumors

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KEYWORDS
Pancreas; Imaging; Lesion characterization; Pancreatic tumor

Abstract The increasing use of abdominal imaging has led to a growing incidence of traditionally uncommon pancreatic tumors. These rare tumors have specific imaging features whose knowledge may heighten confidence in characterization and may avoid unnecessary surgical procedures when imaging findings suggest a benign condition. Computed tomography (CT) is the modality with which rare pancreatic tumors are incidentally detected in the majority of cases. Magnetic resonance imaging (MRI) is often performed as a second line examination for further characterization. This review provides an update on CT and MRI findings of rare tumors of the pancreas.

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Abbreviations

<table>
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<tr>
<td>AFP</td>
<td>alphafoetoprotein</td>
</tr>
<tr>
<td>CA</td>
<td>carbohydrate antigen 19-9</td>
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<tr>
<td>CEA</td>
<td>carcinoembryonic antigen</td>
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<tr>
<td>EUS-FNA</td>
<td>endoscopic ultrasound fine needle aspiration</td>
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<tr>
<td>(^{18}\text{FDG-PET}/\text{CT})</td>
<td>(^{18}\text{Fluoro-deoxy-glucose positron emission tomography coupled with computed tomography})</td>
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<tr>
<td>CT</td>
<td>Computed tomography</td>
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<tr>
<td>MRI</td>
<td>magnetic resonance imaging</td>
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<td>MRCP</td>
<td>magnetic resonance cholangiopancreatography</td>
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Introduction

With an incidence between 7 and 9 per 100,000 in men and 4.5 and 6 per 100,000 in women, the overall incidence of pancreatic cancers remains stable [1]. However, the incidence of unusual pancreatic tumors has increased during recent years, assumably because of an expanded use of cross-sectional imaging [2]. Pancreatic ductal adenocarcinoma accounts for approximately 90% of all pancreatic tumors [2]. Among the rare pancreatic tumors, primary pancreatic tumors are divided into epithelial and non-epithelial tumors. Primary epithelial tumors include exocrine and endocrine tumors according to the World Health Organization classification. Primary non-epithelial tumors include tumors from mesenchymal origin (including vessel, stroma, fat and neural cell-derived tumors) and lymphomas. Pancreatic tumors may also be secondary to the dissemination of primary tumors. Computed tomography (CT) is the modality with which pancreatic tumors are incidentally detected in the majority of cases. Magnetic resonance imaging (MRI) is often performed as a second line examination for further characterization. This article provides an update on CT and MRI features of rare pancreatic tumors (Table 1).

Clinical considerations

Most of rare pancreatic tumors are clinically asymptomatic and often diagnosed at an advanced-stage because of mass effect related symptoms. In this regard, the majority of rare pancreatic tumors have a mean diameter larger than 5 cm at the time of diagnosis. The median age of diagnosis is the fifth decade. Patients may present with abdominal pain, nausea, emesis, anorexia or obstructive jaundice when the tumor is located in the pancreatic head. Rarely, patients may present with a palpable mass. Serum tumor marker levels are usually within the normal range. Fine needle aspiration obtained through endoscopic ultrasound (EUS-FNA) may be a helpful tool in case of suspicion of a benign condition when surgery cannot be considered [3]. Because these rare tumors may mimic actual malignant tumors, per procedure histological frozen section examination may be recommended to avoid aggressive surgical procedure and favor tumor enucleation [4].

Primary epithelial tumors: exocrine tumors

Solid pseudopapillary tumors (SPT)

SPT, or Frantz tumor, accounts for 3% of all pancreatic tumors and 6% of all exocrine pancreatic tumors [5,6]. SPT is mostly found in women in the 2nd and 3rd decade because of its progesterone dependency [6]. The mean size of SPT is 50 mm at the time of diagnosis and the pancreatic head is the most common location [7,8]. SPT is considered as a benign condition even if distant metastases or recurrence after resection have been reported [8,9]. Histopathologically SPT consists of an encapsulated mixed tumor with cystic and pseudopapillary component.

On ultrasound, SPT presents as a solid, well-circumscribed, heterogeneous tumor with internal cystic changes [10,11]. CT and MRI show a well-circumscribed tumor with necrosis, solid component and hemorrhagic areas [12]. Of note, hemorrhagic areas are found in 50% of SPTs [12]. Unenhanced CT shows calcifications in one third of the tumors [13]. After intravenous administration of iodinated contrast material, SPT shows vivid enhancement during the arterial phase that persists during the portal and late phases. On MRI, SPT has regular margins. On T1-weighted images, SPT can be homogeneous and hypointense (47%) or heterogeneous and hypointense (41%), and more rarely heterogeneous and hyperintense (12%) [8]. On T2-weighted MR images, SPT is heterogeneous and hyperintense (94%) or homogeneous and hyperintense (6%) [8]. The solid portion is hypointense on T1-weighted images and hyperintense on T2- and diffusion-weighted images with a low apparent diffusion coefficient (ADC) value [13]. The hemorrhagic areas are hyperintense on T1-weighted images and may display fluid-fluid level (Fig. 1). Because of mass effect, dilatation of the pancreatic duct may be present and best depicted with MR cholangiopancreatography (MRCP) but secondary pancreatic duct dilatation is absent [8,13]. After intravenous administration of a gadolinium-chelate, SPT shows marked enhancement on T1-weighted images obtained during the arterial phase.

Acinar cell carcinoma

Pancreatic acinar cell carcinoma exhibits exocrine pancreatic enzyme secretion (trypsin, lipase, chymotrypsin, and amylase) [14–16]. Patients may present with extra-intestinal clinical symptoms, such as subcutaneous nodules, ectopic fat necrosis, and polyarthritis [16]. It carries a poor prognosis, between ductal adenocarcinoma and endocrine tumors, with a median survival of 19 months. Metastases at the time of diagnosis are often present [15–17]. On cross-sectional imaging, it appears as a well-defined, predominantly oval or round exophytic mass [14]. It usually presents as a dense and predominantly solid tumor without notable cystic changes, although rare cystic variants have been described [14,16,17]. Calcifications may be seen in one third of patients and most of the tumor enhances continuously, but less than the surrounding pancreatic parenchyma [14,18]. After intravenous administration of contrast material, the tumor
<table>
<thead>
<tr>
<th>Tumor type</th>
<th>Predominantly solid</th>
<th>Predominantly cystic</th>
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<tr>
<td>SPT</td>
<td>+</td>
<td>−</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>+</td>
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<td>+</td>
<td>−</td>
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<td>−</td>
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<td>−</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>Centripetal filling-in fluid-fluid levels</td>
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<tr>
<td>Hemangioma</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>+</td>
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<td>−</td>
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<td>Diffuse enlargement</td>
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<td>−</td>
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<td>No Wirsung duct dilatation</td>
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<td>+</td>
<td>−</td>
<td>+</td>
<td>−</td>
<td>+</td>
<td>+</td>
<td>Enlarged lymph nodes</td>
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<td>Leiomyosarcoma</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>±</td>
<td>−</td>
<td>±</td>
<td>Fat, calcification and solid portion</td>
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<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>±</td>
<td>±</td>
<td>Metastases</td>
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<td>+</td>
<td>−</td>
<td>−</td>
<td>−</td>
<td>+</td>
<td>−</td>
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<tr>
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<td>−</td>
<td>+</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>+</td>
<td>Intrapancreatic fat</td>
</tr>
<tr>
<td>SFT</td>
<td>+</td>
<td>−</td>
<td>−</td>
<td>+</td>
<td>−</td>
<td>+</td>
<td>Small and solid hypervascular</td>
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<tr>
<td>Intrapancreatic accessory spleen</td>
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<td>±</td>
<td>−</td>
<td>+ serpiginous</td>
<td>−</td>
<td>−</td>
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<td>−</td>
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<td>18FDG uptake when malignant</td>
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<td>Parallels spleen enhancement</td>
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SFT: solitary fibrous tumor; 18FDG: 18Fluoro-deoxy-glucose; SPT: solid and papillary tumor.
shows heterogeneous enhancement [16]. Compared to ductal adenocarcinoma, acinar cell carcinoma shows greater degrees of enhancement, with well-circumscribed margins [18].

**Pancreatoblastoma**

Pancreatoblastoma is the most common pancreatic tumor in children within the first decade [19], whereas it is exceptional in adults. Its etiology is unknown, however, it has been reported in association with genetic syndromes such as familial adenomatous polyposis syndrome [20] and Beckwith-Weidemann syndrome [21]. In contrary to the children population, there is no male predominance in adults. Pancreatoblastoma involves the pancreatic head in almost half of the cases [19,22]. Non-specific elevation of carcinoembryonic antigen (CEA), alphafetoprotein (AFP), and carbohydrate antigen (CA) 19-9 serum level has been reported. In most of the cases, pancreatoblastoma is discovered at a late stage, with metastases, predominantly hepatic and a poor prognosis [22]. Imaging features are similar in adults and children [23,24].

On imaging, pancreatoblastoma appears as a large, heterogeneous, exophytic multiloculated and well-circumscribed necrotic lesion with enhancing septa [25]. Of interest, higher-grade tumors may have progressively less-defined margins [24]. Rim-shaped or clustered calcifications, biliary and pancreatic ductal dilatation, ascites, hepatic and pelvic metastases, adenopathy and vascular invasion may be present [23]. On MRI, tissular component is hyperintense on T2-weighted images, with low to intermediate signal intensity on T1-weighted images. However, heterogeneous areas within the tumor may be observed, due to calcifications, hemorrhage and necrosis [24]. After intravenous administration of contrast material, pancreatoblastoma shows mild enhancement. However, rapid, arterial enhancement and late washout have been described and could be a very suggestive imaging feature (Fig. 2) [23,24,26].

**Pancreatic hamartoma**

Pancreatic hamartoma is considered as a strictly benign malformation rather than a tumor [27]. Pancreatic hamartoma has no gender predilection and may present as a solid, or solid and cystic lesion [27,28]. Most of pancreatic hamartomas are located in the pancreatic head [27]. Pathologically, pancreatic hamartomas is a well-demarcated mass consisting of mature acini and ducts with distorted architecture embedded in a fibrous stroma [28]. The solid component of the tumor consists of fibrous and adipose tissue, whereas the cystic component consists of dilated ducts [28]. Islets cells of Langerhans may be present [27–29]. Tumor serum markers are usually normal [27].
On imaging, pancreatic hamartoma presents as a well-demarcated, cystic or solid tumor. Ultrasound shows hyperechoic solid tumor with or without cystic changes [30]. The tumor is well-circumscribed, solid and/or cystic, iso- or hypoattenuating on CT. Fatty component within the solid portion may be present as a hypoattenuating area (−30 and −130 HU) on CT [27]. On MRI, the tumor is hyperintense on T1- and T2-weighted images due to fat content and becomes hypointense on fat-attenuated sequences [27]. The solid component is isointense on T1-weighted images, and with a discrete hyperintensity on T2-weighted images (Fig. 3). MRC is useful to rule out connection with the main pancreatic duct. Mural or septal thickening shows progressive and delayed enhancement [27–30]. Morphological changes with time may be pathognomonic, with a shrinking of the cystic part, an increase in the number of cysts and a honeycomb pattern arrangement [27]. In the absence of fatty component, because of the extreme rarity of the lesion, surgical resection with peroperative histological examination is recommended to avoid carcinologic resection and restrict the procedure to a less aggressive surgical enucleation [4].

**Primary non-epithelial tumors**

**Hemangioma and lymphangioma**

Vascular tumors include hemangioma, cystic lymphangioma, hemolymphangioma, hemangioendothelioma, hemangiolastoma, hemangiopericytoma, and angiosarcoma. They represent only 0.1% of all pancreatic tumors [31].

Pancreatic hemangiomas are more frequent in women [32]. An association with Van Hippel-Lindau disease has been reported [33]. Typical imaging findings parallel those of cavernous hemangioma in the liver. However, according to Mulliken et al., the pathogenesis of hemangiomas display three different phases: proliferation, involution and involuted that would result in various macroscopic and various imaging features [34]. On ultrasound, pancreatic hemangioma appears as a well-circumscribed, complex cyst with thick septations [35]. Isoechoic and hyperechogenic components with slow-flow on Doppler imaging and without any ductal or vascular changes are observed. On CT, peripheral or speckle calcifications with heterogeneous solid and cystic component and fluid-fluid levels are observed.

Figure 2. A 21-year-old man with pancreatoblastoma presenting with weight loss. Ultrasound reveals hepatic tumors. a: multidetector row computed tomography in the transverse plane obtained before intravenous administration of iodinated contrast material shows a well-delineated tumor of the pancreatic head with calcifications (arrow); b: multidetector row computed tomography in the transverse plane obtained during the arterial phase following intravenous administration of iodinated contrast material shows early enhancement of a solid mural thickening (arrow); c: diffusion-weighted MR imaging using high b-value shows heterogeneous hyperintense tumor of the pancreatic head (arrow); d: gadolinium-chelate enhanced fat-suppressed T1-weighted MR image in the transverse during the venous phase shows a heterogeneous tumor of the pancreatic head, well-delineated, that moderately enhances, with enhancing septas, and areas of arterial enhancement that decreases (arrow).
may be seen [36,37]. On MRI, pancreatic hemangioma appears as a heterogeneous, multiseptated cystic mass, hypointense on T1-weighted images and hyperintense on T2-weighted images. Some areas of hemorrhage may display hypersignal on T1-weighted images [32]. After intravenous administration of contrast material, pancreatic hemangioma typically appears as a hypervascular lesion with peripheral and septal enhancement and a progressive, peripheral, centripetal filling-in, which is a pathognomonic feature [32]. However, a poorly-enhancing cystic lesion does not exclude the diagnosis [32,37]. No ductal dilatation is seen on MRCP [32].

Pancreatic cystic lymphangioma is predominantly solitary but may be diffuse in case of lymphangiomatosis. It occurs more frequently in women and is often located in the distal pancreas [38–40]. Pancreatic cystic lymphangioma is an endothelium-lined tumor that is the result of congenital malformation of the lymphatic vessels, leading to blockage of the lymphatic flow and thus, to lymphangiectasia [41]. Cystic lymphangiomas are considered from a pancreatic origin when they develop from pancreatic parenchyma, adjacent to the organ, or connected by a pedicle [41]. On imaging, pancreatic cystic lymphangioma presents as a well-circumscribed, encapsulated, polycystic tumor with thin septa, and an attenuation on CT approaching that of water [41–43]. In appearance, cystic lymphangioma is similar to cystadenoma and classically contains no calcification [41–43]. On MRI, pancreatic cystic lymphangioma appears as a markedly hyperintense, homogeneous, multiloculated cystic tumor on T2-weighted images [44]. The septa and walls contain various amounts of smooth muscle fascicles and collagenous connective tissue that may enhance after intravenous administration of a gadolinium-chelate [44]. Serological screening for tumor markers usually returns to normal value and EUS-FNA or CT-guided biopsy may be a valuable diagnostic solving tool [44]. Pancreatic cystic lymphangioma is a benign tumor but can be locally invasive [45]. Surgical enucleation is a diagnostic and curative procedure but partial pancreatectomy may be necessary in some cases [40,45]. Incomplete excision is likely to lead to local recurrence, so that imaging follow-up during the first years following surgery is mandatory [40,45].

Lipoma

The pancreas is a rare location of lipoma [46,47]. Head and tail of the pancreas are the most frequent locations [46]. Embryological theory suggests a trapping of the retroperitoneal or mesenteric fat during the fusion of the ventral and dorsal bud of the pancreas. It is mostly composed of lobules of mature adipose cells with a thick connective tissue capsule [46]. It has characteristic imaging features so that histological biopsy sample is not needed for a definite diagnosis.

Pancreatic lipoma appears as a well-defined, homogeneous, encapsulated tumor with an extracellular fatty component [46–48]. Typically, it is a hypointenuating tumor with fat-like HU values on unenhanced CT (Fig. 4). Of interest, opposed-phase gradient-echo recalled T1-weighted

Figure 3. A 41-year-old woman with pancreatic hamartoma that was discovered incidentally during MDCT. MR imaging was performed for further characterization. a: fat-suppressed T2-weighted MR image in the transverse plane reveals a moderately hyperintense, predominantly solid and well-delineated tumor of the pancreatic head (arrow); b: fat-suppressed T1-weighted MR image in the transverse plane during the arterial phase shows no tumor enhancement (arrow); c: fat-suppressed T1-weighted MR image in the transverse plane during the late phase (3 min) shows progressive, centripetal delayed enhancement.
images show a thin hypointense demarcation between the lipoma and the adjacent pancreatic parenchyma [48]. No enhancement is observed after intravenous administration of contrast material [46–48].

Mainly asymptomatic, pancreatic lipoma has a benign course [47]. However, short-term interval observation is recommended to ascertain stability and help differentiate between pancreatic lipoma and early well-differentiated liposarcoma [47]. Extensive follow-up is not necessary [47]. Surgical removal of the lesion is not recommended. However, in case of atypical findings (male gender, thick septa, calcification, and large tumor) [49] that may suggest well-differentiated liposarcoma, EUS-FNA is recommended to exclude malignancy [3].

Schwannoma and neurofibroma

Schwannoma is the most common peripheral nerve tumor and is only composed of Schwann cells [50]. It has no gender predilection and is considered as a benign condition. However, 5 cases of malignant degeneration have been reported, but only one has been immunohistologically confirmed [4,51,52]. The pancreatic head is the most common location (40%) [53]. The association with type 1 Recklinghausen neurofibromatosis has only been reported in two patients [54]. Serum tumor marker levels are usually normal [50]. Macroscopically, schwannomas are characterized by two histologic components, namely Antoni A and Antoni B areas [55]. Antoni A area is hypercellular and characterized by closely packed spindle cells with nuclear palisading and Verocay bodies, whereas Antoni B area is hypocellular with degenerative changes and contains loosely arranged tumor cells with abundant myxoid stroma [55]. Degenerative changes like hyalinisation, cystic changes, xanthomatous calcification or hemorrhage are often recognized in the Antoni B areas [56]. Imaging findings reflects histological components, thus, Antoni A areas usually correspond to solid and hypervascular enhancing areas, while Antoni B to cystic and hypovascular non-enhancing areas. Polymorphism of imaging features of pancreatic schwannomas results of the relative proportions of Antoni A and Antoni B areas contained within the tumor [57]. Of note, small pancreatic schwannomas predominantly contain Antoni A areas whereas large pancreatic schwannomas predominantly contain Antoni B areas [4].

On ultrasound, pancreatic schwannoma presents as a well-circumscribed, hypoechoic tumor [58]. Unenhanced CT usually demonstrates a well-defined, encapsulated, hypodense or hyperdense mass with central necrosis or fat [57]. Tumors with a predominant Antoni A component are isoattenuating on CT and hypointense on T1-weighted images and moderately hyperintense on T2-weighted images on MRI with central necrosis or fat [58]. The tumor displays early and intense enhancement with a delayed and persistent filling-in [59]. Selective duodeno-pancreatic arteriography coupled with CT shows intense arterial vascularization [50]. Conversely, Antoni B predominant pancreatic schwannoma (representing two third of the lesions) usually appears as a pseudocystic tumor, with minimally or not enhancing liquid portions following intravenous administration of iodinated contrast material [58,59]. Calcifications and hemorrhage have also been reported in Antoni B areas [56].

Because of a potential malignant transformation, surgical enucleation should be systematically considered with peroperative frozen section analysis to avoid unnecessary radical resection of a benign schwannoma [4].

Leiomyosarcoma

Pancreatic leiomyosarcoma develops from the stromal component of the pancreas and is supposed to derive either from smooth muscle cells of the pancreatic ducts or from the wall of small intrapancreatic vessels [60]. Pancreatic leiomyosarcoma has no pancreatic location or gender predilection [60,61]. Serum tumor markers levels are within the normal range [62]. On imaging and histologic examination, pancreatic leiomyosarcoma has the same morphologic features as its counterpart in other organs [62]. Indeed, it has no specific imaging features [62]. It appears as a heterogeneous solid, cystic or mixed egg-shaped, well-circumscribed hypervascular tumor [62] (Fig. 5). As the tumor volume increases, hemorrhagic, necrotic, and cystic changes can be observed [61]. The presence of liver metastases is common while there is usually no lymph node involvement [60,61]. This latter feature could be a valuable clue for radiologists to guide pathologist for specific stromal immunohistochemistry examinations [62]. Local surgical excision is the treatment of choice, but pancreatic leiomyosarcoma carries a poor prognosis, with a median survival time of 48 months [61].

Solitary fibrous tumor

In general, solitary fibrous tumor is a mesenchymal tumor, consisting of spindle cells derivateing from fibroblastic tissue. Solitary fibrous tumors may be found in various sites but the most common location is the pleura. Solitary fibrous
tumors have been reported more frequently in women. Mostly benign, 12% have demonstrated malignant features with metastasis and local recurrence [63]. Serum tumor markers levels are normal [63].

Based on the few reported cases, solitary fibrous tumor of the pancreas is a well-circumscribed, heterogeneous, solid tumor with a central necrotic or hemorrhagic component. On ultrasound, solitary fibrous tumor is a heterogeneous hypoechogenic mass [63]. On CT, the lesion appears isoattenuating to the adjacent parenchyma. On MRI, the lesion shows iso- or hyposignal on T1-weighted images and iso- or hypersignal on T2-weighted images. After intravenous administration of contrast material, CT or MRI shows a hypervascular, early enhancing tumor. Of note, 18-Fluorodeoxy-glucose positron emission tomography coupled with CT may be useful to distinguish between benign and malignant solitary fibrous tumor, and detect local recurrence during the follow-up [63].

**Lymphoma**

Primary pancreatic lymphoma is usually a non-Hodgkin lymphoma. Primary pancreatic lymphoma is extremely rare whereas secondary involvement of the pancreas by an abdominal lymphoma is more common [64]. Mostly located in the head, primary pancreatic lymphoma may mimic pancreatic carcinoma [65]. Behrens et al. have reported suggestive criteria for the diagnostic of primary pancreatic lymphoma [66]. They include a tumor predominantly located within the pancreas, with lymph node involvement confined to the peripancreatic region, no superficial or mediastinal lymphadenopathy, hepatic or splenic involvement, and normal white blood cell count [66].

On imaging, two patterns have been described. One is a localized, well-circumscribed lesion and the other is a diffuse enlargement with an infiltration of the whole pancreas [67,68]. On CT and MRI, primary pancreatic lymphoma appears as a homogeneous tumor, hypoattenuating on CT and hypointense on both T1- and T2-weighted images, with degrees of enhancement similar or slightly lower than that of the adjacent pancreatic parenchyma [69] (Fig. 6). A bulky, localized pancreatic head tumor without Wirsung duct dilatation, enlarged lymph nodes below the level of the renal veins, and invasive and infiltrating growth toward the retroperitoneal or upper abdominal organs and the gastrointestinal tract are clues for the diagnosis of primary pancreatic lymphoma [69]. Usually no calcification or necrosis are seen. Compared to pancreatic ductal adenocarcinoma, the presence of a vascular invasion and metastasis is uncommon. EUS-FNA helps make a definite diagnosis [64]. Chemoradiotherapy is

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**Figure 5.** A 16-year-old girl presenting with acute abdominal pain. a: multidetector row computed tomography in the transverse plane obtained before intravenous administration of iodinated contrast material shows a large well-defined, heterogeneous, solid and cystic tumor of the pancreatic head well-delineated tumor with calcifications (arrow); b: multidetector row computed tomography in the transverse plane obtained during the arterial phase following intravenous administration of iodinated contrast material shows early enhancement of a solid mural thickening (arrowheads); c: multidetector row computed tomography in the transverse plane obtained during the venous phase following intravenous administration of iodinated contrast material shows thick enhancing septa (arrowheads). Percutaneous biopsy showed primary sarcoma.
Imaging shows cysting calcifications. Reportedly, unfortunately, intensity changes in the cystic body may vary according to the examination conditions. Imaging of the cystic body in the transverse plane obtained before intravenous administration of iohexol contrast material reveals a homogenous, well-delineated hypoattenuating mass of the pancreatic body (arrows); b: computed tomography in the transverse plane obtained during the arterial phase shows early peripheral tumor enhancement (arrows); c: computed tomography in the transverse plane obtained during the venous phase shows no enhancement lower than that of the adjacent pancreatic parenchyma (arrow) and no main pancreatic duct dilatation. Histopathological analysis of tissue sample obtained from percutaneous biopsy showed large B cell lymphoma.

Currently, the treatment of choice before considering surgery [65].

Mature teratoma (dermoid cyst)

Mature teratoma or dermoid cysts are frequent germ cell-derived tumors, but the pancreas is the rarest location [65]. Most of the dermoid cysts are located in the pancreatic head of the with no gender predilection [65,70]. Pathologically, mature dermoid teratomas contain skin-like and skin adnexal structures. Imaging findings depend on the various ectodermal components found in the dermoid cyst that include skin, hair, fatty sebaceous debris and calcifications. The classical hair/fluid or cyst/fluid level is pathognomonic of pancreatic dermoid cysts but is unfortunately rarely present [72]. Thus, there is no typical appearance.

On ultrasound, dermoid cyst is a well-circumscribed heterogeneous tumor [73]. On CT, it appears a lobulated and hypoattenuating oligocystic tumor with a thin capsule [73]. Internal or peripheral calcification may be present [73]. MRI shows an oligocystic tumor with a thin capsule [73]. Fatty changes are hyperintense on T1- and T2-weighted imaging and vanish after fat-suppression [70]. MRCP has been reported as a helpful imaging sequence because the hyperintensity of the tumor disappears [70]. Serum tumor marker levels are usually normal. Increased CEA levels in the cyst fluid and whitish necrotic material with no malignant cells may be found on cyst fluid analysis after EUS-FNA [74,75].

Mature teratoma still remains a preoperative challenge and final diagnosis is often made histopathologically after surgical removal of the tumor [70].

Intrapancreatic accessory spleen

Intrapancreatic accessory spleen is the second location of accessory spleens accounting for 17% of all locations [76]. It is usually located 3–4 cm from the splenic hilum, adjacent to the pancreatic tail [77]. Embryologically, accessory spleen is a result of failure of fusion of splenic tissue buds of the dorsal mesogastrium [78]. Pathologically, pancreatic accessory spleen has the same histological features as the main spleen, surrounded by a thin capsule [78].

On imaging, intrapancreatic accessory spleen appears as a less than 4 cm solid enhancing mass with a smooth, round, ovoid or minimally lobulated shape within the tail of the pancreas [76,78]. The patterns of enhancement parallel those of the main spleen with an arterial serpiginous pattern followed by a homogenous enhancement during the portal and late phases (Fig. 7). Rarely, epidermoid cyst may arise from intrapancreatic accessory spleen. It usually appears as a uniloculated or multiloculated cystic tumor with normal splenic tissue surrounding the cyst, either with a mass-like or a rim-like appearance [79]. Identification of intrapancreatic accessory spleen is important to avoid unnecessary surgical procedure. In this regard, heterogeneous arterial enhancement is a main feature to distinguish intrapancreatic
spleen from neuroendocrine tumor [78]. Diffusion-weighted MRI is also helpful to improve lesion characterization [80,81].

Secondary tumors
The pancreas is an unusual site of secondary tumors. Melanoma, lung cancer and breast carcinoma are the most common origins of multiple pancreatic metastases, whereas renal cell carcinoma usually leads to a single pancreatic metastasis [82]. The interval between the diagnosis of the primary tumor and the development of pancreatic metastases varies between 1 and 3 years, except for renal cell carcinoma for which pancreatic metastases may appear after more than 20 years [83]. The presence of other extra-pancreatic metastases is common and facilitates the diagnosis [84]. Pancreatic metastases may present as solitary or multiple tumors, with sometimes a diffuse parenchymal involvement [84]. Pancreatic metastases usually display the same imaging features than their primary counterpart [85]. Hypovascular tumors are well delimitated, homogeneous with a lack of distinct intrapancreatic capsule and show a necrotic center when larger than 1.5 cm [84,86]. Of note, metastases from non-small cell lung cancer may demonstrate an enhancing peripheral rim [86]. Special attention must be given to renal cell carcinoma pancreatic metastases, which demonstrate rapid arterial enhancement and then a washout, isodense or isoointense to the normal pancreatic parenchyma during the portal phase [86–88] (Fig. 8). Their identification is of importance because patients with this condition may benefit from surgical procedures in case of solitary metastasis from renal cell carcinoma [89].

Figure 7. A 62-year-old woman with intrapancreatic accessory spleen discovered incidentally. a: T2-weighted MR image in the transverse plane reveals a moderately hyperintense, solid and well-delineated tumor of the pancreatic tail (arrow); b: diffusion-weighted MR imaging using high b-value shows homogeneous hyperintense tumor (arrow) similar to spleen hyperintensity (arrowhead); c: apparent diffusion coefficient map shows a hypointense tumor with a value of 940 mm²/s, similar to that of the spleen (arrowhead); d: fat-suppressed T1-weighted MR image in the transverse plane during the arterial phase shows serpiginous enhancement of the tumor (arrow) that parallels spleen enhancement (arrowhead); e: fat-suppressed T1-weighted MR image in the transverse plane during the venous phase shows delayed enhancement of the tumor (arrow) that parallels spleen enhancement (arrowhead).
Imaging features of rare pancreatic tumors

Figure 8. A 56-year-old woman with pancreatic metastases from renal cell carcinoma 7 years after left total nephrectomy. a: fat-suppressed T2-weighted MR image in the transverse plane reveals two homogeneous, hyperintense, well-defined tumors (arrowheads) of the pancreatic isthmus and body; b: diffusion-weighted MR imaging using high b-value shows two homogeneous hyperintense tumors (arrowheads); c: fat-suppressed T1-weighted MR image in the transverse plain obtained during the arterial phase following intravenous administration of a gadolinium-chelate shows early arterial enhancement of the tumors (arrowheads); d: fat-suppressed T1-weighted MR image in the transverse plain obtained during the late phase following intravenous administration of a gadolinium-chelate shows persisting enhancement (arrowheads).

Conclusion

Rare pancreatic tumors include a wide range of tumors that represent less than 5% of all pancreatic tumors [2]. However, knowledge of specific imaging features is important for radiologists in order to warn the gastroenterologist or surgeon of the possibility of an alternate diagnosis. Thus, in case of fatty or hemorrhagic tumor content, atypical enhancement or chronological changes and specific clinical context, radiologists must keep consider rare pancreatic tumors whose treatment may substantially differs from those of the more frequent ductal adenocarcinoma or cystic lesions. Histological samples obtained from EUS-FNA can be considered in order to reach a definite diagnosis and avoid unnecessary aggressive surgical procedure.

- An enhancing pancreatic tumor without upstream dilatation of the main pancreatic duct should suggest primary pancreatic lymphoma.
- The presence of a fatty component within a pancreatic tumor is highly suggestive of a benign lesion.
- Secondary pancreatic tumors have the same imaging features as their primary counterpart.
- In case of unusual imaging features, endoscopic ultrasonography with fine needle aspiration should be considered to reach a definite diagnosis and avoid an aggressive surgical procedure.

Clinical case

A 32-year-old woman had a pancreatic mass discovered incidentally on multidetector row computed tomography (MDCT) of the abdomen (Fig. 9).

Questions

1. Among the following item, which one is true? MDCT before intravenous administration of iodinated contrast material (Fig. 9a) shows:

Take-home messages

- The presence of hemorrhage within a mixed solid and cystic tumor suggests solid and pseudopapillary tumor.
- Imaging features of a small tumor of the pancreatic tail should be compared to that of the spleen to rule out the possibility of an intrapancreatic accessory spleen.
Figure 9. A 32-year-old woman with a pancreatic mass discovered incidentally on multidetector row computed tomography (MDCT) of the abdomen. a: MDCT in the transverse section before intravenous administration of iodinated contrast material shows an isoattenuating tumor of the pancreatic body (arrowheads); b: MDCT in the transverse section obtained after intravenous administration of iodinated contrast material during the portal phase shows a homogeneous hypoattenuating well-delineated tumor of the pancreatic body (arrowheads) without dilatation of the pancreatic main duct; c: T2-weighted MRI in the transverse section shows a well-defined heterogeneous high signal intensity tumor of the pancreatic body (arrowheads); d: T1-weighted MRI with fat-suppression in the transverse section shows a low intensity well-delineated and homogeneous tumor of the pancreatic body (arrowheads); e: gadolinium enhanced T1-weighted MRI with fat-suppression in the transverse section obtained during the arterial phase shows a heterogeneous well-circumscribed encapsulated pancreatic mass with early peripheral enhancement (arrowheads).

A, Dilatation of the intra-hepatic biliary tract.
B, Focal liver lesion.
C, Pancreatic tumor.
D, Duodenal tumor.

2. Among the following items, which one is present on MDCT obtained after intravenous administration of iodinated contrast material during the portal phase (Fig. 9b)?
A, A homogeneous and well-defined solid tumor of the pancreatic body suggesting malignancy.
B, No upstream dilatation of the main pancreatic duct suggesting a pancreatic benign lesion.
C, A single focal liver lesion suggesting malignancy.
D, A global bile duct dilatation.

3. Among the following items, which one is present on MR imaging (Fig. 9c–e)?
A, A hyperintense, heterogeneous solid and cystic, predominantly cystic and well-delineated tumor of the pancreatic head on T2-weighted MR images.
B, A spontaneously hyperintense, well-circumscribed and heterogeneous mass of the pancreatic tail on T1-weighted images.
C, A homogeneous, early enhancing pancreatic mass with a washout on dynamic T1-weighted MR images.
D, A heterogeneous encapsulated pancreatic mass with early, peripheral enhancement with a progressive fill-in on dynamic T1-weighted MR images.

4. Among the following items, which one corresponds to the most plausible diagnosis?
A, Pancreatic ductal adenocarcinoma.
B, Pancreatic endocrine tumor.
C, Solid and pseudopapillary tumor.
D, Pancreatoblastoma.

Answers
1. Answer C.
2. Answer B. The patient underwent magnetic resonance imaging (MRI) of the abdomen for further characterization.
3. Answer D.
4. Answer C.

Disclosure of interest
The authors declare that they have no competing interest.

References


