Radiologic overview of primary pancreatic neoplasms - beyond adenocarcinoma

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Learning objectives

The purpose of this overview is to:

- Familiarize the Radiologists with the most important radiologic characteristics of pancreatic neoplasms - other than Adenocarcinoma.

- Provide useful key findings (radiologic, epidemiologic and typical clinic manifestation) to orientate the differential diagnosis.
Background

Diagnostic imaging is an important tool to evaluate pancreatic neoplasms. Pancreatic adenocarcinoma account for 90% of all pancreatic malignancies:

We highlight the most important radiologic features of the rest 10% of pancreatic neoplasms, which are far less common and therefore less known.

Primary pancreatic masses will be classified on the basis of its radiologic appearance in solid or cystic lesions:

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| **C) METASTASES TO THE PANCREAS** |

Radiologists and especially radiographers in training should be aware of this minor group of pancreatic neoplasms. The knowledge of some of the most important characteristic key findings may facilitate the right diagnosis.
Findings and procedure details

We made a complete revision of our data base to illustrate the different types of primary pancreatic tumors- other than adenocarcinoma.

We also describe the most important imaging findings:

A) SOLID LESIONS OF THE PANCREAS:

Pancreatic Neuroendocrine Tumor (NETs)

These account for 1%-5% of all pancreatic tumors and typically manifest in patients aged 51-57 years. Most cases are sporadic, but association with syndromes such as multiple endocrine neoplasia type 1, von Hippel-Lindau syndrome, neurofibromatosis type 1 and tuberous sclerosis has been observed. Tumors tend to be multiple when associated with syndromes.

NETs are classified into functioning and nonfunctioning tumors:

- Functioning tumors produce symptoms related to excessive hormone production. In general, functioning tumors manifest early in the course of disease.

- Nonfunctioning tumors manifest when they are large, due to mass effect. Risk of malignancy increases with tumor size (especially in tumors > 5 cm). Because of this fact 90% of nonfunctioning tumors are malignant at presentation.[2]

Small tumors are generally solid and homogeneous, whereas larger tumors are heterogeneous and may show variable amounts of cystic-necrotic degeneration and calcification[1,2,3] (Figure 1).

NETs have a rich vascular supply and therefore enhance avidly during the arterial phase, enhancing more rapidly and intensely than the normal pancreas.

-> That finding helps differentiate NETs from the more common adenocarcinoma which is hypovascular.
Homogeneous enhancement is typical for small tumors (< 2 cm), whereas larger lesions tend to show heterogeneous enhancement.

When NETs have a predominantly cystic component, MDCT and MRI show a hypervascular enhancement in the nonnecrotic or nondegenerated portions of the tumor. Cystic areas are typically hyperintense at MRI on T2-weighted images (Figure 2).

Metastases to lymph nodes and solid organs such as the liver may have an enhancement pattern similar to that of the primary tumor (Figure 3, 4). Cystic metastases to the liver may also be seen\(^1,2\).

**Solid Pseudopapillary Tumor (SPT)**

They account for <2% of all pancreatic tumors. It is most common in young females (mean age 25 years). SPT has a low malignant potential with an excellent prognosis following complete resection.

SPT is typically a large (mean 9 cm), slow-growing, well-encapsulated mass\(^4\). It most commonly occurs in the pancreatic tail. SPT has a tendency to displace rather than invade surrounding structures and rarely causes obstruction of the bile duct or pancreatic duct.

- MDCT usually demonstrates a well-encapsulated lesion with varying solid and cystic components owing to hemorrhagic degeneration. Hemorrhage may progress to cystic changes within the lesions in approximately 20% of cases. SPT shows peripheral heterogeneous enhancement with central cystic spaces.

- MRI typically demonstrates a well-defined lesion with heterogeneous signal intensity on T1- and T2-weighted images. Peripheral calcification is present in 30% of cases\(^4\). The pseudocapsule (composed of compressed pancreatic tissue and reactive fibrosis) has low attenuation at MDCT and low signal intensity at T1- and T2-weighted MRI.

Internal hemorrhagic and cystic degeneration is the hallmark of SPT due to the fragile vascular network of the tumor\(^1\). Although most SPTs exhibit benign behavior, malignant degeneration does occur. Metastases are uncommon, occurring in 7%-9% of cases, mostly to the liver, omentum, and peritoneum.

**Pancreatoblastoma**

Pancreatoblastoma accounts for 0.2% of all pancreatic tumors and is the most common pancreatic tumor in young children (mean 5 years)\(^3\). Pancreatoblastoma rarely occurs
in adults; when it does, however, the tumor is generally more aggressive. The serum alpha-fetoprotein level is elevated in 25%-33% of cases\[^5\].

Pancreatoblastoma is typically slow growing and generally manifests as an asymptomatic large mass (mean 10 cm). Because of the large size of the mass at presentation, in 50% of cases it is not possible to identify the organ of origin at radiology. Therefore, differentiation from other pediatric tumors arising from adjacent organs (e.g., neuroblastoma, Wilms tumor, hepatoblastoma) is challenging, and biopsy is generally required to establish the diagnosis. Metastases occur mostly to the liver.

- At US, the mass is heterogeneous with hypoechoic cystic spaces and hyperechoic internal septa\[^5\].

- At MDCT, pancreatoblastoma generally manifests as a multiloculated inhomogeneous mass with enhancing septa\[^5\].

- On MRI the tumor has low to intermediate signal intensity on T1- and high signal intensity on T2-weighted images, and shows mild contrast enhancement.

**Pancreatic Lymphoma**

Pancreatic lymphoma is most commonly a B-cell subtype of non-Hodgkin lymphoma. Secondary lymphoma is the dominant form and is the result of direct extension from peripancreatic lymphadenopathy. Primary pancreatic lymphoma is rare, representing 0.5% of pancreatic tumors. It is more common in immunocompromised patients\[^4\].

Two morphologic patterns of pancreatic lymphoma are recognized: a focal well-circumscribed form and a diffuse form.

- The focal form occurs in the pancreatic head in 80% of cases and has a mean size of 8 cm. It typically has uniform low attenuation at MDCT. At MRI, it has low signal intensity on T1- and intermediate signal intensity on T2-weighted images and shows faint contrast enhancement.

- The diffuse form is infiltrative leading to glandular enlargement and poor definition, features that can simulate the appearance of acute pancreatitis\[^5,6\].
**B) CYSTIC LESIONS OF THE PANCREAS:**

Cystic lesions account for 10%-15% of all pancreatic neoplasms and represent < 5% of all malignant pancreatic tumors.

**Serous cystadenoma**

It is a benign lesion which typically occurs in older women. The cystic components range from mm- 2cm.

- When the lesion grows a central scar and coarse calcification may be seen (30%). This calcified scar is highly specific and virtually pathognomonic \[^7\] and is best demonstrated at CT.

- MRI shows a cluster of small cyst without visible communication within the cyst or the pancreatic duct. These cysts are hyperintense on T2-weighted images. Central calcified scar is seen as a signal void at MRI (Figure 5, 6). Enhancement of fibrous septa between the cysts are seen on delayed images.

**Mucinous cystic neoplasm (mucinous cystadenoma / cystadenocarcinoma)**

This lesion has a female predominance (80%) in their sixth decade of life.

They preferentially involve the pancreatic body and tail and do not communicate with the pancreatic duct.

- Cross-sectional imaging is ineffective for differentiating between mucinous cystic neoplasms with and without malignant epithelium, except in cases with invasion of adjacent organs, vascular invasion, or metastatic disease. The presence of intracystic enhancing soft tissues are suspicious for malignancy. Peripheral eggshell calcifications are not frequent (16%) but such finding is specific and has a highly predictive value for malignancy.

- On US mucinous cystic neoplasms appear as hypoechogenic multilocular or, less commonly, unilocular masses with posterior acoustic enhancement. Internal septations are usually visualized and better demonstrated at US than at CT[^7].

- CT shows a round to slightly lobulated mass that is well encapsulated with smooth external margins. Because the cyst contents can vary in attenuation according to the
degree of hemorrhage or protein in the mucoid cysts, different levels of attenuation may be seen within the cyst cavities (Figure 7, 8). After intravenous contrast administration septa and peripheral wall enhancement are detected.

- At MR the lesion is hypointense on T1- and hyperintense on T2-weighted images. This lesion may be hyperintense on T1-weighted images due to mucinous content.

**Intraductal Papillary Mucinous Neoplasm of the pancreas (IPMN)**

IPMNs are most frequently identified in elderly men. The most important features are the presence of mucin-producing tumor and cystic dilation of the main pancreatic duct, its branches or both. The dilated ducts often contain profuse mucin. In the past, many IPMNs may have been misdiagnosed as chronic pancreatitis because of their generally benign behavior.

Preoperative determination of the presence or absence of associated invasive carcinoma is crucial; when invasive carcinoma is present, the surgical procedure may be modified to include resection of regional lymph nodes.

Main duct IPMNs are more likely to be malignant. IPMNs are frequently multifocal, and 5%-10% involve the entire pancreas.

- The most important imaging features suggestive of invasive carcinoma in IPMN are the solid structure, the large size of the mass (> 3.5 cm), presence of mural nodules, dilatation of the main pancreatic duct > 15 mm and multifocal involvement.

- MRI is better than CT for evaluating ductal communication. Dilatation of main pancreatic duct or multiple side branches on T2-weighted images is the most common imaging finding. Demonstrating ductal communication can be useful to differentiate between IPMNs and mucinous cystadenoma (the latter has no communication with the pancreatic ductal system) (Figure 9, 10).

- 3D contrast-enhanced US showed similar results as compared with MRI in evaluating "IPMNs" < 1 cm / > 2 cm.

**C) METASTASES TO THE PancreAS:**
Pancreatic metastases account for 2%-5% of all malignant neoplasms. Metastases are most frequently from renal cell carcinoma (RCC) and lung carcinoma. The prognosis is generally more favorable than that for pancreatic adenocarcinoma (Figure 11).

Three morphologic patterns of involvement are recognized:

- solitary (50%-70%),
- multifocal
- diffuse.

At contrast-enhanced CT and MR imaging, the appearances of pancreatic metastases closely resemble that of primary carcinoma but pancreatic adenocarcinoma generally manifests as a hypoenhancing mass, whereas metastases show either peripheral enhancement (in lesions >1.5 cm) or, less commonly, homogeneous enhancement (smaller lesions).

Cystic metastases to the pancreas cannot be differentiated from mucinous cystic neoplasms radiographically. Ovarian carcinoma metastases are the most likely to manifest as a predominantly cystic mass.

A known history of primary malignant disease, combined with the presence of other metastatic foci, are helpful clues in making the diagnosis.
Fig. 1: Pancreatic neuroendocrine tumor. US images (A), axial unenhanced MDCT and coronal MR T2-weighted image show a round, heterogeneous mass, localized in the pancreatic body, with variable amounts of cystic-necrotic degeneration (arrows).

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**Fig. 2:** Same patient shown in figure 1. MR Axial gradient T1 out-of-phase image (A) and T1 fat-suppressed sequence (C) show a hypointense signal in the liquid component of the lesion whereas it reveals a hyperintense signal in the T2-weighted sequence (B) (arrows).

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Fig. 3: Sagittal MDCT image (A) show a heterogeneous pancreatic mass (arrow). Coronal (B) and axial (C) MDCT images show multiple hypervascular metastases in the liver (arrows), showing the same enhancement pattern of the primary mass. Neuroendocrine pancreatic tumor and metastases were histologically proven.

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Fig. 4: Sagittal MDCT image (A) show a heterogeneous pancreatic mass (arrow). Coronal (B) and axial (C) MDCT images show multiple hypervascular metastases in the liver (arrows), showing the same enhancement pattern of the primary mass. Neuroendocrine pancreatic tumor and metastases were histologically proven.

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Fig. 5: Axial nonenhanced MDCT image (A) depicts a polylobulated cystic lesion with a coarse calcification in its center (arrow), which is the pathognomonic central scar for serous cystadenoma.
**Fig. 6:** MRI of the same patient as in Fig. 5, shows a cluster of small cysts (arrows), which are hypointense in T1-weighted images (B) and hyperintense in T2-weighted images (C,D), without visible communication within the cyst or the pancreatic duct. A central signal void is also identifiable.

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Fig. 7: On US (A,B) a hypoechogenic multilocular mass with well-definable internal septations and posterior acoustic enhancement can be seen. Contrast-enhanced MDCT images (C+D) show a big round to slightly lobulated mass with an enhancing capsule and different levels of attenuation within the cyst cavities are seen. Some enhancing components are also detectable.

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Fig. 8: Contrast-enhanced MDCT images (E+F) show a big round to slightly lobulated mass with an enhancing capsule and different levels of attenuation within the cyst cavities are seen. Some enhancing components are also detectable.

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Fig. 9: MDCT image (A) shows cystic dilatation of the main pancreatic duct and some of its branches in the pancreatic tail. Ductal communication with the tumor cannot be clearly identified.

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Fig. 10: In contrast-enhanced axial T1 (B) and T2-weighted (C) MR images and in MRI cholangiography (D) ductal communication can be easily detectable.
Fig. 11: Oblique reformatted enhanced MDCT image reveals a well-defined round mass in the pancreas, slightly hypodense to the pancreatic parenchyma. Pancreatic metastases from melanoma was proven. Note the liver concomitant metastases.

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Conclusion

This synopsis of the minor group of primary pancreatic tumors (different from the most common Adenocarcinoma) describes their most important characteristics and thereby serves as a structured guideline to make a proper diagnosis.

The knowledge of these key findings may facilitate radiologists, and especially radiographers in training, to do an accurate detection and staging of pancreatic neoplasms in order to ensure an appropriate selection of patients who will benefit from surgery and prevent unnecessary surgeries in patients with unresectable disease.
References


