Rare presentations of common pancreatic neoplasms

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Learning objectives

Common pancreatic neoplasms may present with atypical imaging appearance. The radiologist should be aware of the possible more common variants and the pathological correlation to better report and manage the each single case.

To describe rare presentations of common pancreatic neoplasms, both solid and cystic, providing examples and radiologic-pathological correlations.
Background

Rare solid and cystic pancreatic tumors are heterogeneous neoplasms, infrequently seen in any given individual clinical practice. Distinct pathological entities can be found, according with the World Health Organization (WHO) nomenclature [1]. Atypical presentations of common pancreatic tumors are more frequent and clinically relevant. The typical aspect of these tumors at different imaging modalities are well known and used in the everyday clinical practice; the atypical, uncommon aspects of these tumors must be known in order to avoid relevant misdiagnosis.
Findings and procedure details

SOLID NEOPLASMS

DUCTAL ADENOCARCINOMA

Ductal adenocarcinoma is the most common primary malignancy of the pancreas [2]. Macroscopically, it presents as a white-yellow firm mass, owing to the presence of marked desmoplasia and fibrosis. Mean vascular density (MVD) of pancreatic adenocarcinoma is low. The combination of marked desmoplasia and low MVD, together with the presence of necrosis or mucin, justify its typical hypovascularity at imaging (Fig. 1). At conventional US, pancreatic adenocarcinoma typically presents as a solid hypoechoic lesion [3], showing poor enhancement at contrast-enhanced ultrasound (CEUS) [4, 5]. At CT, the tumor is isodense in pre-contrast phase; the tumor becomes better visible during arterial phase, even if the tumor-to-pancreas contrast difference is still poor, making sometimes suboptimal the detection. Pancreatic phase allows better tumor detection, since during this phase there is the maximum difference of density between the hyperdense pancreatic parenchyma and the markedly hypodense tumor [6]. At MRI, ductal adenocarcinoma usually shows inhomogeneously hypointensity on T1-weighted images and a variable signal intensity on T2-weighted images, especially if necrotic degeneration is present. After contrast medium administration, ductal adenocarcinoma shows poor enhancement; at CT and MRI, late phase acquisitions may show a slightly intralesional pooling of contrast medium, reflected by a slight hypervascularization during this phase, related to the high amount of fibrotic tissue within the tumor [6].

Rare presentations (Fig. 2-9): necrotic, cystic, microcystic, pancreatitis-like shape, pancreatitis epiphenomena, cysts epiphenomena.

NEUROENDOCRINE TUMORS

Pancreatic endocrine tumors arise from neuroendocrine cells. These tumors can be divided into functioning or non (hyper)functioning, based on the presence or absence of symptoms related to hormone production [7]. Nonfunctioning tumors are frequently larger at presentation, with inhomogeneous aspect mainly due to necrotic areas and calcifications. Typically, small endocrine tumors are well-defined, rounded lesions, homogeneously hypoechoic at US, hypodense at CT; at MRI, endocrine tumors are better visualized with fat-suppression sequences, and they present as well-defined rounded lesions, homogeneously hypointense on T1-weighted images and slightly hyperintense on T2-weighted images. Endocrine pancreatic neoplasms are typically hypervascular (Fig. 10) at CEUS and contrast-enhanced CT and MRI [8, 9].
Rare presentations (Fig. 11-15): hypovascular, intravessels-growing, cystic, serotoninoma, calcified.

CYSTIC NEOPLASMS

SEROUS CYSTADENOMA

Serous cystadenoma (SCA) is a cystic tumor, generally detected in 50-70 year-old females (sex-ratio: 2:1), usually located in the pancreatic head as a solitary lesion. SCA has a typical multilocular "honeycomb" architecture due to the presence of multiple microcysts (<20 mm), thin wall and multiple septa oriented toward a central scar (Fig. 16, 17). The typical radiologic lobulated "cloud-like" morphology is clearly demonstrable at ultrasound. The cystic content appears homogeneously anechoic at US, hypodense at MDCT and hypointense on T1-weighted images at MRI. T2-weighted images clearly demonstrates the microcystic pattern; after intravenous administration of contrast material, the hypervascularization of the central scar and of internal thin septa may be seen. [4, 10, 11]. When a microcystic lesion whit US findings comparable with SCA is found, the final report must address the need of an MRI with MRCP. SCA does not communicate with the pancreatic ductal system and this can be well demonstrated on MRCP: this finding remains crucial for the differential diagnosis in respect to branch duct IPMNs [12-14].

Rare presentations (Fig. 18-25): unilocular, oligocystic, pseudosolid, huge dimensions.

INTRADUCTAL PAPILLARY MUCINOUS NEOPLASMS

Intraductal papillary mucinous neoplasms (IPMNs) are exocrine mucin-producing neoplasms. Three types of IPMNs have been described: the branch duct type, the main duct type and the mixed type (Fig. 26). The segmental or diffuse involvement of the main pancreatic duct identifies lesions with higher risk of malignancy. The demonstration of the communication with the ductal system is needed for an appropriate diagnosis [14, 16, 18]. MRI is the imaging modality of choice for the noninvasive diagnosis of IPMNs. At imaging, features of malignancy consist of the presence of solid enhancing mural nodules or septa [19, 20].

Rare presentations (Fig. 27, 28): pan-ductal-ectasic (wirsungocele/santorinicele), colonizing growth.

MUCINOUS CYSTIC NEOPLASMS

Mucinous cystadenoma (MCA) is a pancreatic cystic tumor with female sex predilection. MCA is a benign mucin-producing lesion with a proved malignant potential. Thus, in respect to serous cystadenoma, MCA requires surgical resection. MCA appears as a
single lesion, usually located in the body-tail of the pancreas, without communication with the pancreatic ductal system. MCA usually presents as a macrocystic lesion, with rounded "ball-like" morphology, irregular septa, thick wall and complex content that can be particle, viscous and dense owing mainly to mucinous content (Fig. 29). This content makes very often the lesion heterogeneously hypoechoic at US, hypodense at CT and slightly hyperintense on T2-weighted images at MRI. On T1-weighted images, the signal intensity can vary from hypointensity to hyperintensity, depending on mucin concentration. CPMR clearly demonstrates the lack of communication with the pancreatic ductal system. Differing from SCA, on post-contrast enhanced phase, the intralesional septa are disorganized and peripherally located, describing a "bridge" along the cystic wall with a "pseudonodular" appearance. Peripheral calcifications along the thick wall can be detected [14-17].

Rare presentation (Fig. 30-32): site (head), disepitelized, gender (male).

SOLID-PSEUDOPAPILLARY NEOPLASMS

Solid-pseudopapillary tumors (SPTs) are epithelial neoplasms with low malignant potential, occurring predominantly in young women. They usually develops as solid tumors and then undergo massive hemorrhagic degeneration, giving rise to a cystic appearance at imaging (Fig. 33). The cystic areas often presents hematic content, usually well depicted at CT and even better at MRI. At dynamic imaging, homogeneous enhancement of the solid components and the peripheral thick wall is well demonstrated [21, 22].

Rare presentations (Fig. 34): small and solid.
Images for this section:

**Fig. 1:** DUCTAL ADENOCARCINOMA Typical fibrotic mass with low mean vascular density resulting hypoechoic at US, hypovascular at CEUS, MDCT and MRI and hypointense on T1 MRI

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Fig. 2: NECROTIC PANCREATIC ADENOCARCINOMA

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Fig. 3: NECROTIC PANCREATIC ADENOCARCINOMA Necrotic pancreatic body mass at MDCT

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Fig. 4: NECROTIC PANCREATIC ADENOCARCINOMA Necrotic pancreatic tail mass at MRI

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Fig. 5: CYSTIC ADENOCARCINOMA Pancreatic body-tail huge cystic/solid mass with hypovascular component

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Fig. 6: MICROCYSTIC ADENOCARCINOMA Microcystic pancreatic body adenocarcinoma with microcystic appearance at MRI mimic serous cystadenoma

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Fig. 7: PANCREATITIS-LIKE SHAPE Pancreatic body-tail adenocarcinoma hypoechoic at US and hypovascular at MDCT involving all the gland
**Fig. 8:** PANCREATITIS EPIPHENOMENA Pancreatic body adenocarcinoma hypoechoic at US with dilation of the main pancreatic duct and hypovascularization of the body-tail at MDCT.

**Fig. 9:** CYSTIC EPIPHENOMENA Very small adenocarcinoma hypovascular at MDCT with secondary irregular dilation of the adjacent collateral branches and the main pancreatic duct mimic IPMN at MRI.

**Fig. 10:** ENDOCRINE TUMORS Typical mass with well defined margins and high mean vascular density resulting hypoechoic at US and hypervascular at CEUS and MDCT.
Fig. 11: HYPOVASCULAR ENDOCRINE TUMOR Double duct sign with small ill defined hypointense on T1 and T2 MRI and hypovascular pancreatic head mass

INTRAVESSEL GROWING ENDOCRINE TUMOR
Pancreatic body endocrine tumor growing into the splenic vein
**Fig. 12:** INTRAVESSEL GROWING ENDOCRINE TUMOR Pancreatic body endocrine tumor growing into the splenic vein

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**Fig. 13:** CYSTIC ENDOCRINE TUMOR Pancreatic head cystic mass with thick wall hypervascular at MDCT

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SEROTONINOMA
Small fibrotic nodule isointense on T2 MRI and slightly hyperintense on late phase dynamic MRI obstructing the main pancreatic duct uniformly upstream dilated

Fig. 14: SEROTONINOMA Small fibrotic nodule isointense on T2 MRI and slightly hyperintense on late phase dynamic MRI obstructing the main pancreatic duct uniformly upstream dilated

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Fig. 15: CALCIFIED ENDOCRINE NEOPLASM Pancreatic body calcified mass and small hypervascular round nodule at the pancreatic isthmus

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**Fig. 16:** SEROUS CYSTADENOMA Typical microcystic appearance at specimen, MRI and ultrasound

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**Fig. 17:** SEROUS CYSTADENOMA Typical microcystic appearance at specimen and MRI

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**Fig. 18:** UNILOCULAR SEROUS CYSTADENOMA Macrocyst arising from the tail of the pancreas with tick enhancing wall at MRI and thin enhancing septa at CEUS
**Fig. 19:** UNILOCULAR SEROUS CYSTADENOMA Marcocyst of the tail of the pancreas. Macroscopic appearance.

**Fig. 20:** PSEUDOSOLID SEROUS CYSTADENOMA Hypoechoic small pancreatic body lesion resulting typically microcystic and not communicating with the main pancreatic duct at MRI
**Fig. 21:** OLIGOCYSTIC SEROUS CYSTADENOMA Oligocystic mass of the body-tail of the pancreas with thick enhancing septa at CT Macroscopic appearance

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**Fig. 22:** PSEUDOSOLID SEROUS CYSTADENOMA Inhomogeneous hypoechoic lesion typically microcystic and not communicating with the main pancreatic duct at MRI

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Fig. 23: PSEUDOSOLID SEROUS CYSTADENOMA Pancreatic lesion inhomogeneously hypervascular and hyperdense at CT resulting typically microcystic at MRI

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**Fig. 24:** PSEUDOSOLID SEROUS CYSTADENOMA Pancreatic lesion solid and homogenously hyperechoic at ultrasound resulting typically microcystic at MRI

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**Fig. 25:** HUGE SEROUS CYSTADENOMA Pancreatic body-tail lesion inhomogeneously hyperechoic with microcysts and big central calcification at ultrasound resulting typically microcystic at MRI

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**Fig. 26:** INTRADUCTAL PAPILLARY MUCINOUS NEOPLASM Typical mass with papillary proliferation and dilatation of the main pancreatic duct (w) visible at US with enhancing vegetation at CEUS and communication of the cystic mass with the main pancreatic duct at MRI.

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**Fig. 27:** PAN-DUCTAL-ECTASIC IPMN Santorinicele.
**Fig. 28:** DUODENAL COLONIZATING IPMN Huge papillary vascularized pancreatic mass colonizing the lumen of the duodenum from the major papilla.

**Fig. 29:** MUCINOUS CYSTADENOMA Typical round macrocystic mass of the pancreatic body with thick wall and enhancing septa at CEUS and at CT
Fig. 30: PANCREATIC HEAD MUCINOUS CYSTADENOMA Unilocular cystic mass in the head of the pancreas

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Fig. 31: DISEPITELIZED MUCINOUS CYSTADENOMA Huge unilocular cystic mass of the pancreatic body

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**Fig. 32:** MUCINOUS CYSTADENOMA IN MALE PATIENT Cystic mass with enhancing septa in the tail of the pancreas

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**Fig. 33:** SOLID-PSEUDOPAPILLARY NEOPLASM Typical hemorrhagic cystic round pancreatic body mass with blood content at MRI and enhancing solid component at CEUS

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Fig. 34: SMALL SOLID SOLID-PSEUDOPAPILLARY NEOPLASM Small solid hypoechoic at US, hypodense at CT and hypointense on MRI lesion in the pancreatic body

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Conclusion

Common pancreatic neoplasms, both solid and cystic may rarely present with atypical appearance; their histopathological bases and radiologic features must be known in order to avoid misdiagnosis.
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