

Rare pancreatic neoplasm: MDCT and MRI features of a typical Solid Pseudopapillary Tumor

Stefano Palmucci^{1*}, Alessandra Uccello¹, Giorgia Leone², Giovanni Failla¹, Giovanni Carlo Ettore¹

1. Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele", Catania, Italy

2. Section of Anatomic Pathology, G.F. Ingrassia, University of Catania, Catania, Italy

* **Correspondence:** Stefano Palmucci, MD, Radiodiagnostic and Oncological Radiotherapy Unit, University Hospital "Policlinico-Vittorio Emanuele", Via Santa Sofia 78, 95123 Catania, Italy

(✉ spalmucci@sirm.org)

Radiology Case. 2012 Jan; 6(1):17-24 :: DOI: 10.3941/jrcr.v6i1.823

ABSTRACT

Solid pseudopapillary tumor of the pancreas is a rare neoplasm, predominantly observed in young women and with greatest incidence in the second and third decade. It has clinically good behavior, although large at the time of diagnosis. We report the case of a thirty-year-old woman with a giant mass in the pancreas, incidentally discovered during an abdominal ultrasonography. The mass was later investigated by multidetector computed tomography and magnetic resonance imaging. The cystic-solid appearance of the encapsulated lesion suggested to radiologists the possibility of a solid pseudopapillary tumor. Imaging features of this pancreatic neoplasm and differential diagnosis from other cystic pancreatic tumors are discussed in our report, in order to help radiologists and clinicians achieve correct diagnosis and management.

CASE REPORT

CASE REPORT

A thirty-year-old woman was admitted to our hospital due to the presence of an abdominal mass incidentally revealed in an ultrasonography (US) exam that had been performed in a private radiological center. The patient had been referred for US exam due to hypercholesterolemia and clinical suspicion of gallstone disease. The mass was described as a giant round-shaped solid lesion, containing multiple internal anechoic areas, located in the epigastric region; no intralesional vascular sign was revealed by US color-doppler examination (Figure 1); the axial diameters of the lesion were approximately 85 x 65 mm.

Abdominal physical examination of the patient was normal; laboratory tests - except for the above-mentioned hypercholesterolemia - were insignificant. The patient was subsequently referred for a multidetector computed

tomography (MDCT) scan and for a magnetic resonance imaging (MRI) exam, in order to better characterize the pancreatic mass and define its relationships with other organs. The MDCT (Figure 2), performed before and after contrast administration (Iomeprolo), showed the large mass centered on the body of the pancreas, with round-shaped cystic appearance; no fluid level was observed. On unenhanced images the mass was homogeneously hypodense, but post-contrast acquisition revealed a weak intralesional enhancement. This progressive fill-in enhancement gave rise to a suspicion of the presence of solid intralesional tissues. For this reason the patient underwent an MRI exam (Figure 3), using a 1.5 Tesla scanner (GE Signa HDxt, Milwaukee, USA). T2-weighted images showed multiple internal slightly hypointense papillary areas; based on the presence of cystic and solid components, radiologists diagnosed a solid pseudopapillary tumor (SPT). The diagnosis was confirmed by open-surgery biopsy and histological findings from the

specimen (Figure 4). Immunohistochemical findings were: CD10: + (15%); Vimentin: + (100%); Enolase: + (95%); Progesterone receptor: + (90%); Cyclin D1: focal positivity (<15%).

DISCUSSION

SPT accounts for 0.13-2.7% of all exocrine pancreatic neoplasms [1]. Its morphological features were described for the first time by Dr. Frantz in 1959 [2-4]. Macroscopic and histological features of this pancreatic tumor have been variously described in literature: solid cystic tumor, papillary cystic neoplasm, papillary cystic tumor, papillary epithelial neoplasia, solid and papillary epithelial neoplasia, papillary epithelial tumor, Frantz's tumor, solid and papillary tumor, solid-cystic-papillary epithelial neoplasm, benign or malignant papillary tumor of the pancreas and adenocarcinoma of the pancreas in childhood [2]. In 1996 the World Health Organization (WHO) finally defined this neoplasm as "SPT of the pancreas" [2].

Unlike pancreatic cancer, SPT occurs most frequently in young women, "with greatest incidence in the second and third decade", and a female to male ratio of 10:1 [4]. In many published works, a low level of aggression and a good prognosis are reported for this tumor [3-5].

In its typical appearance, SPT is a well-defined encapsulated mass composed of a mixture of cystic and solid components [6-8]. In our report we describe the morphological appearance of this tumor, considering the most important elements to make a differential diagnosis from common pancreatic cystic neoplasms.

SPT of the pancreas is often asymptomatic and discovered incidentally during diagnostic exams; the first symptom frequently reported is a vague abdominal pain; "there are no abnormalities in laboratory tests; in addition, serum markers of pancreatic neoplasms are normal" [9].

The tumor has a low grade of malignancy and a good prognosis is reported for patients; however, atypical manifestations - metastases (involvement of liver and peritoneum), main pancreatic duct obstruction, capsular invasion or parenchymal infiltration - are also described in literature [6]. Even when aggressive, the tumor maintains a good prognosis for the patient and a long life-expectancy has been reported in cases of diffuse metastases [10].

Imaging shows several macroscopic and microscopic features that are important in easily differentiating the pseudopapillary tumor [7] from other cystic lesions: as mentioned above, it is often discovered incidentally and, as reported in our clinical history, "at the time of diagnosis appears as a large pancreatic mass", limited by a capsule and with variable intralesional solid, cystic and hemorrhagic components [11]. MDCT reveals a large inhomogeneous mass limited by a capsule; after contrast administration there is a progressive enhancement, with gradual intralesional "fill-in

enhancement" in the portal and venous diagnostic phase. Thanks to the high contrast resolution, MRI identifies the presence of the capsule and the intratumoral hemorrhage better than MDCT [12]; magnetic resonance cholangiopancreatography (MRCP) may demonstrate a relationship with main pancreatic duct.

Immunoistochemical findings reported from our specimen were typical enough for the diagnosis of SPT: indeed, almost all solid-pseudopapillary neoplasms strongly and diffusely express vimentin, CD10, neuron-specific enolase, progesterone receptors and about 75% express cyclina D1 too.

Several cystic lesions occur in the pancreatic parenchyma: a high rate of diagnostic overlap is frequently observed; MDCT and MRI are used to make a differential diagnosis because they are extremely useful in the identification of cystic lesions [7]. The characterization of a cystic lesion requires an accurate study of intralesional components: presence of hemorrhage, fluid level, calcifications, septa, internal nodule and degree of enhancement are all important features in better defining the nature of the lesion [11]. Pseudopapillary solid tumor of the pancreas may have typical and atypical manifestations [6]. Typical appearance consists of an encapsulated mass with varying cystic and solid components caused by hemorrhagic degeneration; calcification and slow enhancement of intralesional components are reported too [6,12]. In the case we report here, we found a classic cystic-solid lesion; in this case we feel that diagnosis is better suggested by MRI rather than by MDCT. "Our pre- and post-contrast MDCT images could suggest the presence of internal papillary components only due to the presence of weak enhancement, whereas MRI clearly demonstrates the solid papillary content thanks to its excellent contrast resolution" (Figure 3).

In atypical appearance SPT should be differentiated from ductal adenocarcinoma. It is important to remember that adenocarcinoma is more frequently observed in male adults and that at the time of presentation it is small in size. The main pancreatic duct obstruction is occasionally found due to the large size of the solid pseudopapillary mass.

Pancreatic pseudocysts can present peripheral calcifications and hemorrhagic areas [13]; in these cases it may be difficult to make a differential diagnosis [14]. However, since a pseudocyst occurs after an episode of pancreatitis, an accurate clinical history of the patient is very useful in making a differential diagnosis. The presence of occasional internal debris may simulate a solid component, especially if there is no fluid level; a change in the patient's position is helpful in identifying debris because its internal disposition is gravity-dependent.

Solid pseudopapillary tumors could be indistinguishable from non-functioning islet cell tumors; these lesions are similar "because they are well-defined and have a cystic appearance, limited by a continuous capsule" [6]. In our experience, the clinical presentation of these tumors is very similar, because a non-functioning islet cell tumor is generally detected when it reaches a large size.

In conclusion, preoperative diagnosis of SPT remains difficult owing to its heterogeneous appearance; the rate of misdiagnosis reported in a work by Yang et al. [15] is very high (38.5%); fine-needle aspiration in well-defined tumors is indicated for unresectable lesions, because the risk of seeding of neoplastic cells with any other form of biopsy is present [15]. Radiologists should keep in mind that pancreatic lesions with cystic-solid appearance in young females are compatible with SPT, and that a differential diagnosis is important to begin appropriate clinical management.

TEACHING POINT

Solid pseudopapillary tumors occur most frequently in young women, with greatest incidence in the second and third decade, and a female to male ratio of 10:1. Typical appearance consists of an encapsulated mass with varying cystic and solid components caused by hemorrhagic degeneration; calcification and slow enhancement of intralesional components are reported too [6,11]. Atypical manifestations such as metastases (to liver and peritoneum), main pancreatic duct obstruction, capsular invasion or parenchymal infiltration may occur. Multidetector CT and MRI are useful in the identification of cystic lesions and in the characterization of intralesional components and thus for a formation of a good differential diagnosis.

REFERENCES

1. Shaikh S, Arya S, Ramadwar M, Barreto SG, Shukla PJ, Shrikhande SV. Three cases of unusual solid pseudopapillary tumors. Can radiology and histology aid decision-making?. JOP 2008; 9(2): 150-159. PMID: 18326922
2. Bostanoglu S, Otan E, Akturan S, Hamamci EO, Bostanoglu A, Gokce A, Albayrak L. Frantz's tumor (solid pseudopapillary tumor) of the pancreas. A case report. JOP 2009; 10(2): 209-211. PMID: 19287121
3. Zinner MJ, Shurbaji MS, Cameron JL. Solid and papillary epithelial neoplasms of the pancreas. Surgery 1990; 108(3), 475-480. PMID: 2396191
4. Volmer CM Jr, Dixon E, Grant DR. Management of solid pseudopapillary tumor of the pancreas with liver metastases. HPB (Oxford) 2003; 5(4), 264-267. PMID: 18333000.
5. Ulasan S, Bal N, Kizilkilic O, Bolat F, Yildirim S, Yildirim T, Niron EA. Case report: solid-pseudopapillary tumour of the pancreas associated with dorsal agenesis. Br J Radiol 2005; 78(929), 441-443. PMID: 15845940.
6. Choi JY, Kim MJ, Kim JH, et al. Solid pseudopapillary tumor of the pancreas: typical and atypical manifestations. AJR Am J Roentgenol 2006; 187(2), W178-186. PMID: 16861508.
7. Sahani DV, Kadavigere R, Saokar A, Fernandez-del Castillo C, Brugge WR, Hahn PF. Cystic pancreatic lesion: a simple imaging-based classification system for guiding management. Radiographics 2005; 25(6), 1471-1484. PMID: 16284129.
8. Frias Vilaça A, Rodrigues P, Scigliano H, Pinto J, Reis A. Solid Pseudopapillary Tumor of the Pancreas: a rare and probably misdiagnosed neoplasm. Radiology Case. 2011 Jul;5(7):24-34.
9. Coleman KM, Doherty MC, Bigler SA. Solid-pseudopapillary tumor of the pancreas. Radiographics 2003; 23(6), 1644-1648. PMID: 14615569.
10. Kang CM, Kim KS, Choi JS, Kim H, Lee WJ, Kim BR. Solid pseudopapillary tumor of the pancreas suggesting malignant potential. Pancreas 2006; 32(3), 276-280. PMID: 16628083.
11. Kalb B, Sarmiento JM, Kooby DA, Adsay NV, Martin DR. MR imaging of cystic lesions of the pancreas. Radiographics 2009; 29(6), 1749-1765. PMID: 19959519.
12. Buetow PC, Buck JL, Pantongrag-Brown L, Beck KG, Ros PR, Adair CF. Solid and papillary epithelial neoplasm of the pancreas: imaging-pathologic correlation on 56 cases. Radiology 1996; 199(3), 707-711. PMID: 8637992.
13. Kehagias D, Smyrniotis V, Gouliamos A, Vlahos L. Cystic pancreatic neoplasms: computed tomography and magnetic resonance imaging findings. Int J Pancreatol 2000; 28(3), 223-230. PMID: 11373061.
14. Aydiner F, Erinanç H, Savaş B, Erden E, Karayalçin K. Solid pseudopapillary tumor of the pancreas: emphasis on differential diagnosis from aggressive tumors of the pancreas. Turk J Gastroenterol 2006; 17(3), 219-222. PMID: 16941259.
15. Yang F, Jin C, Long J, et al. Solid pseudopapillary tumor of the pancreas: a case series of 26 consecutive patients. Am J Surg 2009; 198(2), 210-215. PMID: 19268906.
16. Geers C, Moulin P, Gigot JF, Weynand B, Deprez P, Rahier J, Sempoux C. Solid and pseudopapillary tumor of the pancreas - review and new insights into pathogenesis. Am J Surg Pathol 2006; 30(10): 1243-1249. PMID: 17001154.

FIGURES

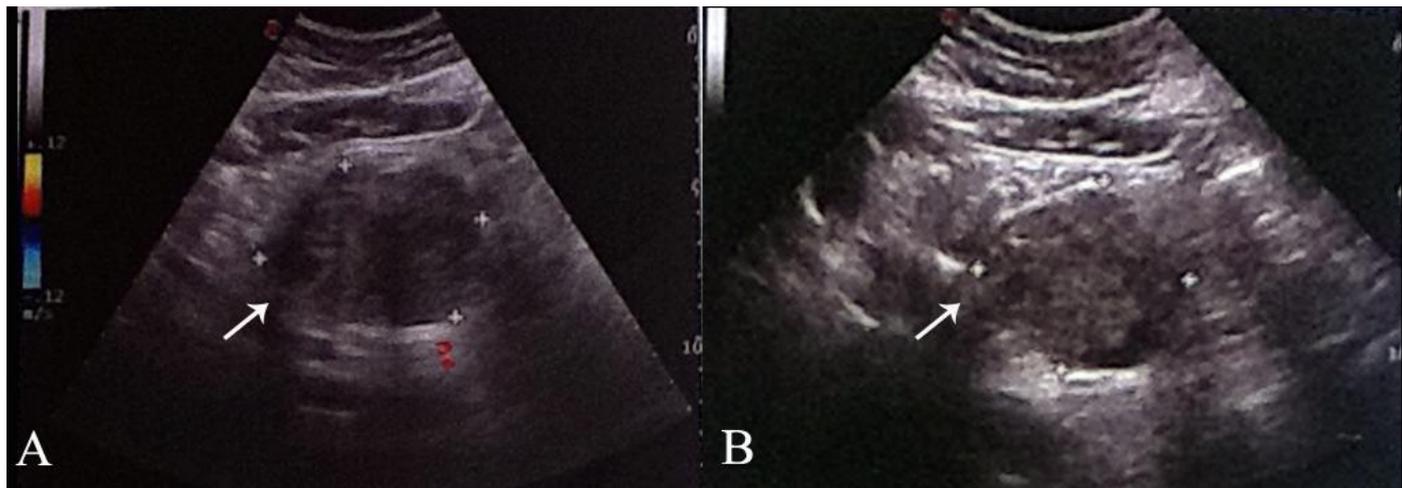


Figure 1: A thirty-year-old female with a typical solid pseudopapillary tumor. Ultrasonography was performed using a 3.5 Mhz convex transducer; color-Doppler (figure A) and grayscale (figure B) images were acquired (kindly provided by patient; also courtesy of Dr. Giuseppe Di Bella of "Centro di Epatogastroenterologia e nutrizione", Giarre). Images A and B show a lobulated mass (white arrow) centered on the body of the pancreas, with round-shaped cystic appearance; the mass is well defined and contains multiple echoic intralesional areas - due to the presence of solid components.



Figure 2: A thirty-year-old female with a typical solid pseudopapillary tumor. Computed tomography - unenhanced (2a) and enhanced images (2b and 2c, respectively arterial and portal phases), obtained by a multidetector scanner (Protocol: 140 Kv, 250 milliamperes, slice thickness = 2.5 mm, contrast medium Iomeprolo 400 mg/ml, total dosage of contrast 120 ml). The images show a large mass (white arrow) centered on pancreatic parenchyma; in figure 2a the mass appears slightly hypodense. After contrast administration (figure 2b and figure 2c) there is a slow fill-in enhancement, suggesting the presence of intralesional solid components. In the venous phase - figure 2c - a cystic area (asterisk) appears in the large mass.

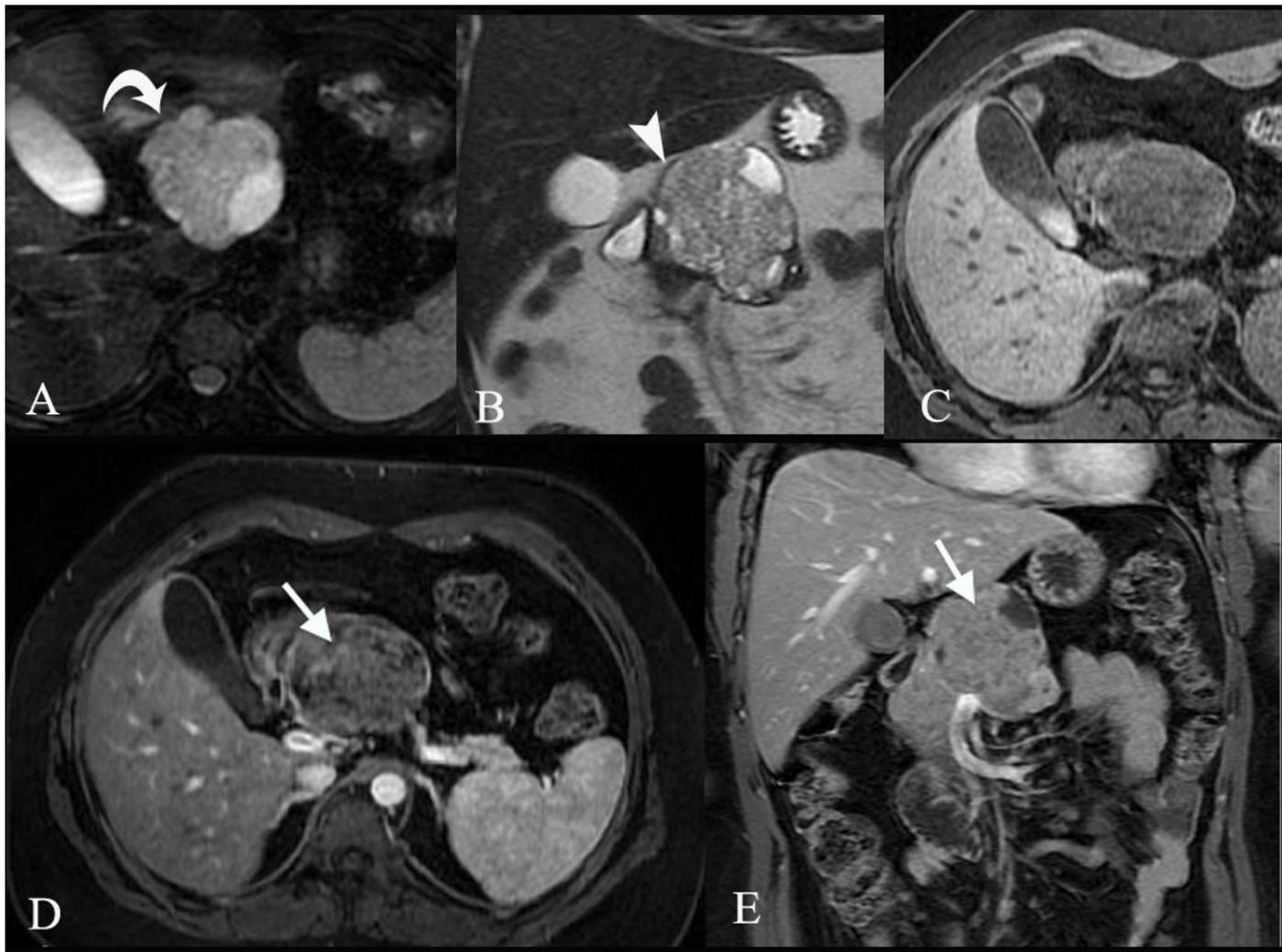


Figure 3: A thirty-year-old female with a typical solid pseudopapillary tumor. Fig. 3 A. Axial fast spin echo image with fat saturation - TR= 2100 msec, TE= 98.5 msec, thickness 6 mm, spacing 1 mm - obtained by a 1.5 Tesla GE Signa HDxt scanner. MRI shows the presence of the pancreatic mass (white curved arrow in Fig. 3a) and clearly demonstrates the solid intralesional components. Fig. 3b. Coronal T2-weighted single-shot spin echo image - TR= 715 msec, TE= 92.5 msec, thickness 5 mm, spacing 1 mm - obtained by a 1.5 Tesla GE Signa HDxt scanner. The image shows a lobulated pancreatic mass, which contains multiple internal slightly hypointense solid areas. The lesion is limited by a continuous capsule - which appears hypointense on image (white arrowhead). Axial unenhanced image (Fig. 3c), axial enhanced image (Fig. 3d) and coronal enhanced image (Fig. 3e), obtained by a T1-weighted 3D fast spoiled gradient echo image - TR = 4.3 msec, TE = 2.1 msec, thickness = 3 mm - using a 1.5 Tesla GE Signa HDxt scanner. After contrast administration (Gadobenate dimeglumine 0.5 M, at a dosage of 0.1 mmol/kg) MRI clearly demonstrates the solid content thanks to its excellent contrast resolution; there is a slow fill-in enhancement (white arrow), due to the presence of intralesional solid components.

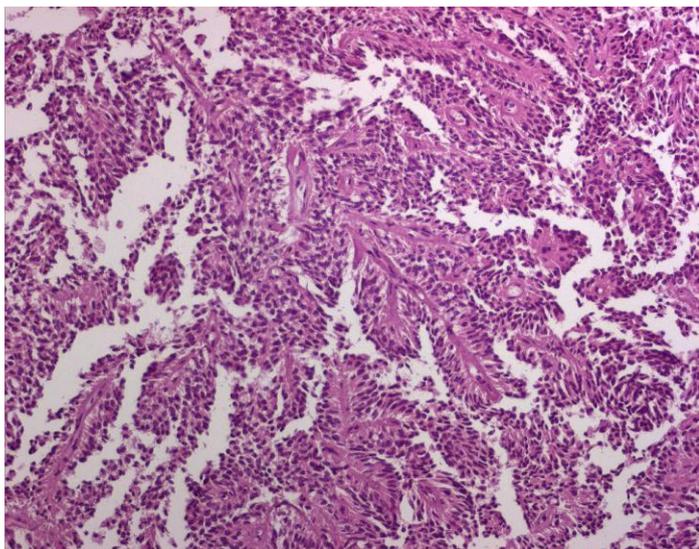


Figure 4 (left): A thirty-year-old female with a typical solid pseudopapillary tumor. Solid-pseudopapillary neoplasm microscopic findings on our biopsy specimen (Haematoxylin eosin, 20x high power field): prominent pseudopapillary growth pattern; pseudopapillae are formed when neoplastic cells drop away, leaving a variable number of cells surrounding delicate capillary-sized blood vessels.

SUMMARY TABLE

Etiology	<ul style="list-style-type: none"> Not yet established; A pancreatic origin is unlikely; many Authors believe SPT might originate from the genital ridge-related cells - incorporated into the pancreatic parenchyma during organogenesis; this may in fact justify the presence of estrogenic receptors [9] Galectin-3 - involved in the carcinogenesis of pancreatic ductal adenocarcinoma - has not been investigated in SPT; progesterone receptors are frequently reported in SPT, whereas the presence of estrogen receptors is unclear [16]
Incidence	Rare neoplasm [7]; 0.13-2.7% of all exocrine pancreatic neoplasms [1]
Gender Ratio	Female to male ratio of 10:1 [4]; Asian and African women mainly affected
Age Predilection	Greatest incidence in the second and third decade
Risk Factors	Female; young age
Treatment	Treatment of solid-pseudopapillary tumor is surgical excision
Prognosis	<ul style="list-style-type: none"> The tumor has a low grade of malignancy and a good prognosis; however, atypical manifestations – metastases (involvement of liver and peritoneum), main pancreatic duct obstruction, capsular invasion or parenchymal infiltration – are also described in literature [6] Even if aggressive, the tumor maintains a good prognosis for the patient and a long life-expectancy has been reported in cases of diffuse metastases [10]
Findings on imaging	<ul style="list-style-type: none"> Typical appearance of a well-defined encapsulated mass composed of a mixture of cystic and solid components MDCT shows a large inhomogeneous mass limited by a capsule; after contrast administration there is a progressive and gradual intralesional “fill-in enhancement” in the portal and venous diagnostic phases MRI identifies the presence of the capsule and the intratumoral hemorrhage better than MDCT [12] MRCP could demonstrate a relationship with the main pancreatic duct In atypical appearance SPT of the pancreas may present as a tumor with metastases in the liver or a ductal obstruction

Table 1: Summary table of Solid Pseudopapillary Tumor (SPT)

LESION	US	CT	MR	Pattern of contrast enhancement	PET
SPT	heterogeneous mass, containing multiple internal anechoic and echoic areas	unenanced images show a homogeneously hypodense mass, cystic lesion	encapsulated mass with cystic and solid components; MR T1: heterogeneous signal MR T2: heterogeneous signal, lesion containing solid papillary areas;	post-contrast acquisitions revealed - on MDCT and MR images - a weak intralesional enhancement ("progressive fill-in enhancement")	flurodeoxyglucose uptake in the solid portion; cystic areas and calcification do not show tracer accumulation
Adenocarcinoma	heterogeneous mass, infiltrative pattern	heterogeneous and infiltrative lesion	MR T1: hypointense, ill-defined lesion MR T2: heterogeneously hyperintense lesion, ill defined MR DWI: restricted diffusion	irregular contrast enhancement, hypovascular lesion	intense tracer accumulation; heterogeneous uptake may be due to the presence of necrosis
Pancreatic pseudocyst	often well encapsulated; anechoic or ipoechoic pattern in lesions containing transudate fluid	well encapsulated mass, hypodense; possibilities of hemorrhagic material or intralesional debris	MR T1: hypointense, signal intensity based on intralesional content (possibilities of internal debris) MR T2: hyperintense or heterogeneous signal intensity	generally absent or mild enhancement of the wall	increased flurodeoxyglucose uptake in recent pseudocysts
Non-functioning islet cell tumors (cystic pattern)	cystic lesion limited by a continuous capsule; anechoic areas (cystic spaces), hypoechoic and hyperechoic areas (solid portions)	well defined cystic lesion, heterogeneous appearance (solid and cystic areas)	encapsulated cystic mass; MR T1: hypointense lesion or heterogeneous signal intensity; MR T2: heterogeneous lesion, containing solid hypointense areas;	enhancement of solid areas; hypervascular intracystic components	increased flurodeoxyglucose uptake
Serous cystadenoma	lobulated mass, inhomogeneous hypoechoic pattern	lobulated mass, slightly hypodense on unenhanced images; possibility of a central scar, with calcifications	polycystic, oligocystic or honeycomb pattern; MR T1: hypointense; MR T2: multiple hyperintense cysts - ranging from a few mm up to 2 cm; MR DWI: no restricted diffusion	weak enhancement of wall; late enhancement in the central scar	no increased uptake
Mucinous cystadenoma	mass with smooth contours; usually hypoechoic cysts exceeding 2 cm in diameter	hypodense cystic lesions; possibility of peripheral calcifications	MR T1: hypointense; MR T2: cystic spaces show high signal intensity MR DWI: no restricted diffusion	enhancement of wall or septa	no increased uptake
IPMNs	small hypoechoic cystic lesions	small hypodense cystic lesions	MR T1: hypointense cysts MR T2: hyperintense cysts MR DWI: generally no restricted diffusion	generally absent; possibility of enhancement of intralesional nodules or septa	no increased uptake

Table 2: Differential table of Solid Pseudopapillary Tumor (SPT)

ABBREVIATIONS

MDCT = MultiDetector Computed Tomography
MRCP = Magnetic Resonance Cholangiopancreatography
MRI = Magnetic Resonance Imaging
SPT = Solid Pseudopapillary Tumor
TE = Echo Time
TR = Ripetition Time
US = Ultrasonography

KEYWORDS

Pancreas; Pancreatic Neoplasms; Carcinoma, Papillary/diagnosis; Magnetic Resonance Imaging; Tomography, X-Ray Computed; Gruber-Frantz tumor; Frantz-Gruber tumor

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/823

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiolopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad



www.EduRad.org