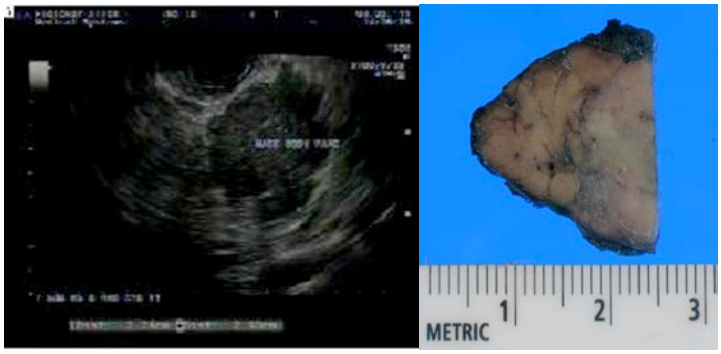
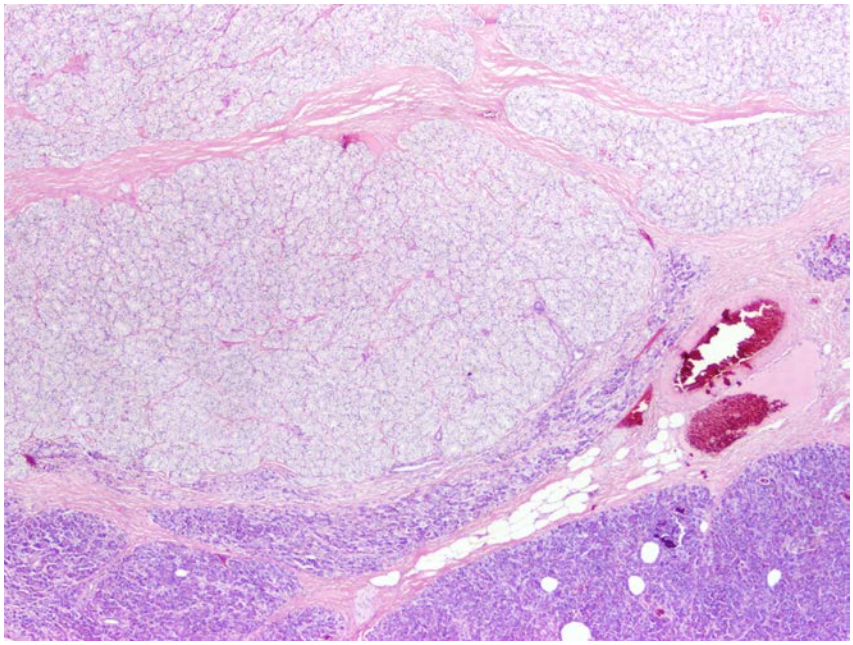
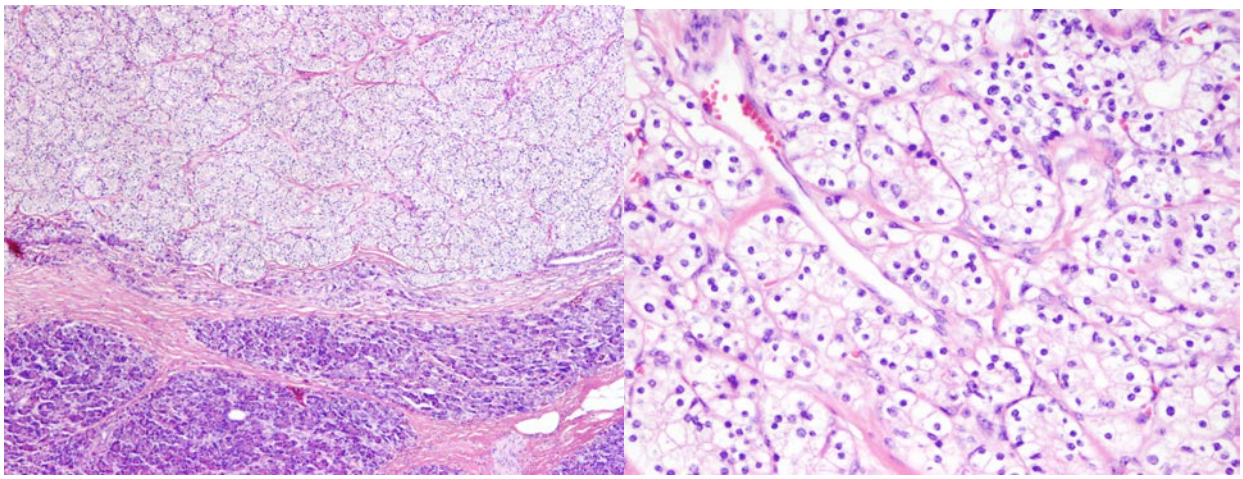
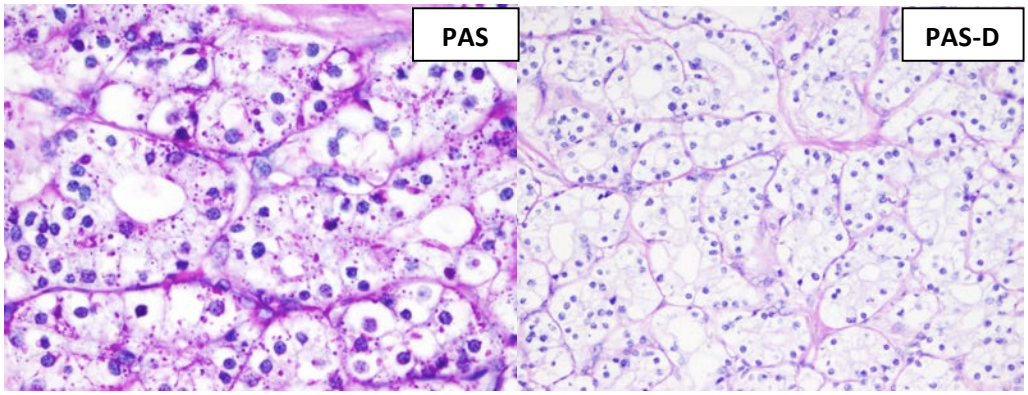
Case:

A 62 year-old man with a history of hyperlipidemia presents with mild abdominal pain and was found to have elevated pancreatic enzymes (amylase 187U/L and lipase 1691 U/L). His family history was significant for thyroid cancer in a sister and pancreatic and prostate cancer in his father. CT scan revealed a 3.0 cm solid, well-circumscribed pancreatic mass at the junction of the body and tail compressing the splenic vein. Subsequent endoscopic ultrasound revealed a 2.7 x 2.5 cm hypoechoic mass in the distal pancreatic body with no dilation of the bile duct. Both imaging reports favored a pancreatic neuroendocrine neoplasm. FNA was non-diagnostic. A distal pancreatectomy was performed. Gross examination revealed a 2.4cm well demarcated mass. Representative histologic sections and selected special stains are shown below as well as a table summarizing the immunohistochemical profile.









Antibody (Clone)	Result
Beta Catenin (14/Beta-Catenin, BD Biosciences)	Positive; membranous pattern
CD10 (56c6, Lab Vision)	Negative
CD56 (56C04, Lab Vision)	Negative
Chromogranin (LK2H10, Hybritech)	Negative
Synaptophysin (Snp88, Biogenex)	Negative
PAX-2 (polyclonal, Invitrogen)	Negative
PAX-8 (MRQ-50 Ventana)	Negative
SMA (1A4 Sigma)	Negative

What is your diagnosis?

a. Metastatic renal cell carcinoma
b. Well differentiated PEN, clear cell variant
c. Solid serous adenoma of the pancreas
d. Solid-pseudopapillary neoplasm, clear cell variant
e. Acinar cell carcinoma

Answer: (C) Solid Serous Adenoma of the Pancreas

Discussion:

Pancreatic serous cystadenomas are a relatively uncommon neoplasm composed of glycogen-rich epithelial cells thought to derive from a centroacinar cell origin. The most common subtype of this neoplasm, the microcystic serous cystadenoma has a characteristic appearance on both imaging and on gross organ examination and therefore rarely creates a diagnostic dilemma.

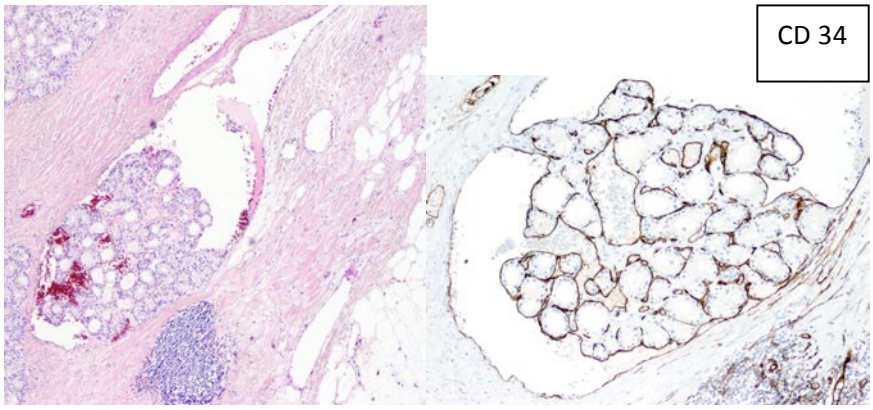
Microcystic serous cystadenoma : characteristic central stellate scar and sponge-like appearance (compare with case study patient above)

However, as its name implies, the solid serous adenoma (SSA) of the pancreas has a solid configuration and therefore losses its characteristic appearance. In addition, it is the rarest variant of the already uncommonly encountered pancreatic serous cystadenoma and therefore may not enter into a differential diagnosis. Given their solid, well-circumscribed, and hypervascular appearance on CT scan and hypoechoic appearance on ultrasound, they are usually diagnosed preoperatively as pancreatic neuroendocrine neoplasms. Microscopically, SSAs have a well-demarcated border, exhibit a solid growth pattern, and are composed of small nests and trabeculae of clear neoplastic cells with small uniform nuclei. Intermixed within the tumor there are thick collagenous fibrous bands. Atypia and mitoses are absent. On histologic examination, diagnostic considerations hinge on its clear cell morphology and include metastatic RCC, pancreatic ductal or NET with clear cell features, PEComa and solid-pseudopapillary neoplasm with clear cell features.

Immunohistochemical and special stains are essential in delineating SSAs from other clear cell neoplasms. As in the classic microcystic serous cystadenoma, the neoplastic cells of SSAs have abundant intracytoplasmic glycogen by PAS stain. Pretreatment with diastase removes glycogen leaving the cytoplasm clear**.** As seen in the summary table above all neuroendocrine markers and RCC markers were negative. The presence of normal membranous staining with β-catenin without nuclear accumulation distinguishes our case from a solid-pseudopapillary neoplasm with clear cell features. PEComas of the pancreas are quite rare and would be expected to show immunoreactivity with SMA.

The solid serous adenoma was first described in 1996 by Perez-Ordonez et al. and is undoubtedly the rarest subtype of the serous neoplasms of the pancreas. All reported cases to date have demonstrated a benign course; however, rare cases of local recurrence and metastasis in the broader category of serous cystadenoma/serous

cystadenocarcinoma of the pancreas have been reported. Interestingly, our case demonstrated foci of venous invasion; however, the patient remains disease-free at a clinical follow-up of two years.



References

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