Solid and Papillary Epithelial Neoplasm of the Pancreas: A Case Report

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Solid and papillary epithelial neoplasm of the pancreas is a rare low-grade malignant tumor occurring chiefly in young women. We describe the clinical and radiologic presentation of the tumor in a 29-year-old woman and discuss the histogenesis, imaging appearance, and differential diagnosis.

Key words: Pancreas, Neoplasms; Computed tomography (CT)

CASE REPORT

A 29-year-old woman with hyperthyroidism under medical treatment was admitted because of a seven-day history of pruritic skin rash and fever of 38°C. Laboratory findings were noncontributory except for leukocytosis of 22,000/mm³ with a shift to the left and mild liver function impairment. During investigation of the source of fever, an abdominal sonogram incidentally showed a large, well-circumscribed oval mass of complex echogenicity in the tail of the pancreas. She denied a history of oral contraceptive use, alcoholism, biliary tract disease, pancreatitis, or abdominal trauma.

A CT scan (Fig. 1) revealed a sharply defined, thick-walled lesion, displacing the splenic vein anteriorly. The lesion had cystic and solid portions without definite internal septation. Contrast material-enhanced CT showed enhancement only in the solid portion.

At surgery, there was an 11x8x5 cm tumor in the tail of the pancreas adhering to the spleen. She was treated with distal pancreatectomy and splenectomy. The cut surface of the tumor showed a solid area with hemorrhagic necrosis and a cystic area filled with yellowish-red fluid in a thick fibrous capsule (Fig. 2).

Microscopically, the tumor was a SPEN composed of multiple long, thin papillary structures and solid areas. The former were composed of layers of tall cells situated on fibrovascular stalks (Fig. 3) and the
latter of sheets of small, round uniform cells with minimal mitotic activity. No evidence of recurrence is found during follow up for 14 months.

**DISCUSSION**

The histogenesis of pancreatic SPEN is controversial. Eccentric nuclei, numerous mitochondria, sparse endoplastic reticulum, and very few secretory granules are suggestive of terminal pancreatic duct cell origin [3,11]. However, features of acinar origin may be demonstrated. For instance, secretory granules or large zymogen-like granules of various sizes are found in some cells. The closely packed cells form small intercellular spaces resembling rudimentary acini [4].

Preoperative recognition is important because excision of this low-grade malignant tumor leads to an excellent prognosis. The appearance of the SPEN on the CT scan is variable [5-8, 10]. A thick capsule is usually present. The internal architecture of the tumor varies, including solid muscle density mass, mixed solid and cystic components and a thick-walled cyst, depending on the degree of hemorrhagic necrosis. The CT number (Hounsfield unit) of the cystic area is higher than that of water and ranges from +40 HU to +50 HU. There is no discernible septum within the tumor. Calcification is not uncommon and is more often seen in the capsule. MR imaging also demonstrates the internal architecture of the tumor. The fibrous capsule and intratumoral hemorrhage are more easily perceived on MRI than on CT, but calcification is more obvious on CT. Definitive diagnosis depends on the histological examination.

The differential diagnosis includes microcystic adenoma, mucinous cystic neoplasm, islet cell tumor, pancreatic duct cell adenocarcinoma, pancreatoblastoma, and pancreatic pseudocyst. The first four rarely occur in patients younger than 30 years of age. Microcystic adenoma [12] is composed of innumerable cysts smaller than 2 cm in diameter, which may have a characteristic honeycomb appearance. Mucinous cystic neoplasms [13], including cystadeno- ma and cystadenocarcinoma, contain large unilocular or multilocular cysts and may look like a SPEN. Multilocularity and thin septations in mucinous cystic neoplasms, however, usually enable differentiation, since they are not seen in SPEN.

Nonfunctioning islet cell tumors [14] may be indistinguishable from SPEN. Although islet cell tumors occur in a slightly older age group and do not have a female predominance, they may appear as a well-demarcated large pancreatic mass with or without cystic components, internal hemorrhage, calcifications and liver metastasis. When dynamic imaging is performed, islet cell tumors are usually hyperdense on arterial-dominant phase, compared with the hypodensity of SPEN. Pancreatic adenocarcinoma, the most common primary pancreatic malignancy, usually does not grow as large as SPEN. Calcification is quite unusual and cystic degeneration is extremely uncommon in these neoplasms. Pancreatoblastoma [15] is an extremely rare disease, usually discovered in childhood. Cystic areas due to liquefaction necrosis may be present without evidence of hemorrhage, a clue to the diagnosis.

*Figure 1.* A 29-year-old woman with a solid and papillary epithelial neoplasm of the pancreas. **a.** Non-enhanced CT scan shows a well-defined, cystic lesion without evidence of calcification or internal septation in the tail of the pancreas. **b.** Following a bolus injection of contrast material, the lesion reveals enhancement only of the solid portion.
In the majority of cases, it is not possible to provide a specific preoperative diagnosis because of the substantial overlap of imaging features of these tumors. However, in the appropriate clinical setting, a reasonable estimate of the likelihood of a particular diagnosis can be made on the basis of imaging findings.

In conclusion, when the CT shows a pancreatic tumor with characteristic findings, including a sharply demarcated large mass, solid and cystic portions with a CT number higher than that of water, and no definite internal septation in a young female patient, we believe that SPEN should be the primary diagnostic consideration.

REFERENCES

胰腺的固體及乳頭狀上皮性腫瘤：病例報告

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胰腺的固體及乳頭狀上皮性腫瘤是一種主要發生於年輕女性，罕見的低度惡性腫瘤。我們敘述一個29歲女性病患的臨床和影像表現，並討論組織學來源、影像特徵與鑑別診斷。

關鍵詞：胰腺腫瘤，電腦斷層掃描