Small solid pseudopapillary tumor of the pancreas in a 32-year-old man; Report of a Case.

A Case of Small SPT in a young man

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Abstract

Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm that mainly occurs in young women. We report a case of small SPT arising from the head of the pancreas in an asymptomatic 32 year-old man, plus a literature review of this tumor. A 32-year-old man was admitted to our department at Kumamoto University Hospital for evaluating of a pancreatic mass. The tumor was with central necrosis, which was poorly perfused on contrast-enhanced CT and high intensity on T2-weighted MRI. Histology revealed that the lesion was a solid pseudopapillary tumor of the pancreas with the characteristic pseudopapilla formation and central degeneration. However, capsule formation was not eminent. The tumor was positive for CD56, CD10, alpha-1-antitrypsin, alpha-1-antich*y*motrypsin, beta-catenin, progesterone receptor. However, pancreatic hormones, chromogranin-A, CEA, CA19-9 were negative. We diagnosed SPT based on these histological findings. Small-sized solid pseudopapillary tumor of the pancreas is increasingly recognized with recent advances in CT and MRI. We should also consider SPT even if in a male case, when the tumor contains necrosis-suspected area which is poorly perfused on contrast-enhanced CT and high intensity on T2-wighted MRI.

Keyword

Solid pseudopapillary tumor (SPT), pancreatic tumor, pancreatic neoplasm

Introduction

Solid pseudopapillary tumor (SPT) of the pancreas is a rare neoplasm of low malignant potential[1, 2]. It was first described by Frantz in 1959[3] and has been recognized with increasing frequency in recent year. Various terms have been used to describe this neoplasm, including solid and cystic tumor, Frantz tumor, papillary epithelial neoplasm, solid-cystic papillary epithelial tumor, and papillary cystic tumor. In 1996, the World Health Organization renamed it as solid pseudopapillary tumor[2]. It accounted for 0.2%-2.7% of primary pancreatic neoplasms, with slow growth and favorable prognosis. SPT affects predominantly young women. Grossly, this tumor is a large, well-encapsulated mass that usually demonstrates variable degrees of internal hemorrhage, cystic degeneration and calcification. The tumor is regarded as of low-grade malignant potential, because follow-up of numerous cases has shown that most patients had long-term survival after surgical resection[4]. As for the treatment for SPT, surgical resection that included simple enucleating, distal pancreatectomy with or without splenic preservation and pancreaticoduodenectomy, chemotherapy and radiotherapy were reported[1, 4, 5]. Chemotherapy is successful in SPT with multiple hepatic metastases, and radiotherapy has been suggested in cases of unresectable SPT[5]. In this report, we present a male case of small SPT with clinical information, CT and MRI findings, and literature review.

Case Report

A 32-year-old man admitted to our department at Kumamoto University

Hospital for evaluation and treatment of a pancreatic mass. He had neither abdominal pain nor vomiting. Ultrasonography (US) showed a hypoechoic and poorly-circumscribed 15 mm mass at the pancreatic head. Plain CT scan did not show any tumor (Figure 1a). Contrast-enhanced CT scan showed hypodese mass within the head of the pancreas (Figure 1b). The peripheral part was gradually enhanced and became isodense with surrounding pancreatic parenchyma. In the pancreatic phase, a small central low density area became to be eminent (Figure 1c, d). A magnetic resonance imaging (MRI) revealed a clearly-demarcated 15 mm mass in the head of the pancreas (Figure 2). T1-weighted MR images demonstrated areas of low signal intensity. Small high signal intensity areas on T2-weighted images seen in central may correspond to the small low density area on CT, suggesting degeneration into cystic component. Endoscopic retrograde cholangiopancreatography (ERCP) showed normal pancreatic duct. Although plasma insulin levels were slightly higher than normal, selective arterial secretagogue injection (SASI) test was negative with no significant increase in plasma insulin after selective stimulation. The diagnosis was difficult because the mass demonstrated rare radiological features. Nonfunctioning islet tumor, solid pseudopapillary tumor, and pancreatic ductal carcinoma were the suspected diagnosis. The patient underwent subtotal stomach-preserving pancreatoduodenectomy with lymph node dissection and had an uneventful postoperative course. Cut section showed a solid 15 mm tumor and without obvious cystic component (Figure 3a). However, there was a scarce area suspecting degeneration in histology

(Figure 3b, c). The surrounding part showed pseudopapilla formation, which is characteristic for SPT. Capsule formation was not eminent. The tumor was positive for Vimentin, CD56, CD10, alpha-1-antitrypsin, alpha-1-antichymotrypsin, beta-catenin, progesterone receptor (Figure 3d). However, pancreatic hormones, chromogranin-A, CEA, CA19-9 Estrogen receptor were negative. These immunohistochemical results are summarized in Table 1. We diagnosed SPT based on these histological findings.

Discussion

A male case of small SPT of the pancreas was reported. SPT of the pancreas is increasingly recognized. A large well-encapsulated mass with internal hemorrhage, cystic degeneration and calcification seen in a young woman is diagnostic[6, 7, 8]. In the large reviews in the English literature by Papavramidis[5] that included 718 well-documented cases of this neoplasm, 626 (91%) of the patients were female with a mean age of 22 years (ranging from 2 to 85 years). Okino et al. [9] summarized 173 cases reported in Japanese literatures. Average age was 34.3 years in men and 28.9 years in women, and that the men-women ratio was 1:10. As for the size of the tumor, Papavramidis and Okino reported that mean diameter of the tumor was 6.1 cm and 7.8 cm. Typical SPT is also reported to be encapsulated[2, 5]. Comparing with the reported cases, our case is not typical in all aspects above. However, we should know that the imaging findings of SPT are sometimes not typical and that small tumor size and male gender cannot

contradict a possibility of this neoplasm. Imaging studies of solid pseudopapillary neoplasm of the pancreas demonstrate variable degrees of hemorrhagic degeneration. Although recent advances in CT and MRI have significantly improved pancreatic imaging, preoperative diagnosis of SPT is still difficult especially for small-sized cases. The problematic differential diagnosis of small-sized and non-cystic SPT on CT or MRI includes nonfunctioning islet tumor and pancreatic ductal carcinoma. Nonfunctioning islet tumor appears cystic degeneration, calcification, internal hemorrhage and develop a liver metastasis[10,11]. Pancreatic ductal carcinoma shows hypodese/hypointense mass in the pancreas on CT or MRI. However, in approximately 10% of the cases, the mass is isodense on contrast-enhanced CT[12]. We should make the diagnosis of pancreatic solid tumors considering small-sized and non-cystic SPT. The tumor in our case, which was small and without any eminent cystic component macroscopically in the cut section, was an atypical case. Uchimi et al.[13] reported that SPT without cystic component tended to be seen in men, smaller in size, and asymptomatic. Degeneration in SPT may occur in the rather late stage. However the presence of a high signal intensity area on T2-weighted MRI and a poorly perfused area on enhanced CT, which corresponds to the degenerated area on histology, might be a key finding for correct preoperative diagnosis. Pettinato et al. [14] reported that characteristic cytological features of SPT, which was the presence of branching papillary fronds composed of central thin fibrovascular stalks, by fine-needle aspiration. And islet tumor, pancreatic ductal carcinoma could be

distinguished from SPT from their cytological features. Cytologic examination is well worth performing preoperatively if it is performed in safety because we can select a limited operation. On the other hand, Okada et al. [15] reported that a pancreatobiliary fistula caused by an intraductal papillary-mucinous pancreatic neoplasm (IPMN), manifesting as obstructive jaundice, and the implantation of a papillary-mucinous neoplasm in the mucosa of the common bile duct via a pancreatobiliary fistula. We should consider the implantation of the IPMN when selecting a limited operation, and determining the surgical margin of the bile duct. Moreover, we should know that fine-needle aspiration and percutaneous transhepatic biliary drainage (PTBD) carry a risk of a site implantation.

The histogenesis and natural history of SPT in man remain controversial. Our case was positive for CD56, CD10, alpha-1-antitrypsin, alpha-1-antichymotrypsin, beta-catenin, progesterone receptor. However, pancreatic hormones, chromogranin-A, CEA, CA19-9, Estrogen receptor were negative. The accumulation and analysis of small SPT in men are necessary for further clarification of its nature. In conclusion, we should also consider SPT even if in a male case, when the tumor contains necrosis-suspected area which is poorly perfused on contrast-enhanced CT and high intensity on T2-wighted MRI.

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Figure Legends

Figure 1. Computed tomography

(a) Plain scan. The mass was unclear.

(b) In arterial-phase imaging CT, the mass was hypodense.

(c) In pancreatic-phase imaging CT, central area of the mass became to be hypodense.

(d) Coronal reconstruction showed a slightly hypovascular and poorly demarcated tumor in the pancreatic head, in which the eminent poorly perfused area is present in the center.

Figure 2. Magnetic resonance imaging (MRI)

(a) T1-weighted axial MR image showed a small hypointensity mass in the pancreatic head.

(b) T2-weighted axial MR image showed the mass as slight hyperintensity tumor with an eminent central high intensity.

(c) T2-weighted coronal MR image showed almost the same findings as in the axial image.

Figure 3. Macroscopic appearance, Microscopic appearance (H&E stain), and immunohistochemical staining

(a) Gross appearance showed a solid area with 1.5 cm in diameter.

(b) Histology showed degenerating component in the center (×2.5).

(c) Typical appearance with prominent myxoid change of the collagen surrounding the stalks of the pseudopapillae (×400).

(d) Immunohistochemical staining (×400). The tumor cells showed positive reaction for alpha-1-antitrypsin (Left), alpha-1-antichymotrypsin (Middle), beta-catenin (Right).

Table 1. Results of immunohistochemical staining