Diagnosis of a Solid Pseudopapillary Neoplasm Using EUS-FNA

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Abstract

A woman in her 50s was found to have a pancreatic mass on abdominal ultrasound. The tumor measured 40 mm in diameter and included a cystic lesion and calcification. In this case, we suspected a diagnosis of solid pseudopapillary neoplasm (SPN) due to the findings observed on various images. However, we were unable to exclude the possibility that the lesion was a neuroendocrine tumor. Therefore, we performed endoscopic ultrasound (EUS)-guided fine-needle aspiration (EUS-FNA). In addition, in order to confirm the diagnosis of SPN, we performed minimized resection (segmental pancreatectomy). Obtaining a definitive preoperative diagnosis of SPN using EUS-FNA can guide the surgical approach.

Key words: solid pseudopapillary neoplasm, EUS-FNA, minimized resection

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Introduction

Solid pseudopapillary neoplasm (SPN) was first described by Frantz in 1959 (1). SPN of the pancreas is a rare neoplasm with a reported frequency of between 0.17% and 2.7% of all nonendocrine tumors of the pancreas (2). It is usually observed in young women and is generally asymptomatic. The lesions are typically large (2.5-10 cm) and encapsulated and frequently contain varying amounts of necrosis, hemorrhage, calcification and cystic changes (3). Because SPN is rarely aggressive and has a low-grade malignant potential with an excellent prognosis after complete resection, it should be differentiated from other, more aggressive tumors, such as adenocarcinoma and endocrine tumors (4). SPN is an ideal pancreatic tumor for treatment with minimized resection (5). Therefore, making an accurate preoperative diagnosis is very important. Endoscopic ultrasound (EUS)-guided fine-needle aspiration (EUS-FNA) cytology has recently been established as a modality for diagnosing pancreatic mass-related lesions. We herein present a case of SPN that was diagnosed before surgery using EUS-FNA. We were able to perform minimized resection (segmental pancreatectomy) in this case. Obtaining a definitive preoperative diagnosis of SPN using EUS-FNA can guide the surgical approach.

Case Report

A woman in her 50s was referred to our hospital. She had no symptoms; however, she was found to have a pancreatic mass on abdominal ultrasound (US) during a complete medical checkup. US showed a low echoic mass measuring 44 mm in diameter with calcification and a cyst that projected forward in the pancreatic body. Abdominal cystography computed tomography also revealed an increasingly enhanced mass measuring 40 mm in diameter with calcification and a cyst that projected forward in the pancreatic body (Fig. 1). T1-weighted magnetic resonance (MR) images demonstrated low signal intensity in the body of the pan-
Figure 1. Abdominal plain computed tomography showed a mass measuring 40 mm in diameter with calcification that projected forward in the pancreatic body (arrowhead) (A). The tumor contained a cyst (arrow), and the solid lesions of the tumor were increasingly enhanced on cystography CT (B).

Figure 2. T1-weighted magnetic resonance (MR) images showed low signal intensity in the body of the pancreas (A). T2-weighted MR images showed high signal intensity in the body of the pancreas (B). A fat-suppressed T1-weighted image partially showed very high intensity, and bleeding in the cyst was suspected (arrow) (C).

creas. T2-weighted MR images showed high signal intensity in the body of the pancreas. A fat-suppressed T1-weighted image partially showed very high intensity, and bleeding in the tumor was suspected (Fig. 2). EUS (GF-UE 260; Olympus Co., Tokyo, Japan) demonstrated the same findings as abdominal US. On contrast-enhanced harmonic EUS (CH-EUS) (using sonazoid) (SSD-alpha 10 Ultra-sound System; Aloka Co., Ltd., Tokyo, Japan), the tumor was increasingly enhanced, although it was hypovascular compared with the surrounding pancreatic parenchyma (Fig. 3). We first suspected a diagnosis of SPN based on the imaging findings. However, neuroendocrine tumor (NET) with cystic degeneration was also suspected as a secondary differential diagnosis, and we performed EUS-FNA to obtain a definitive diagnosis. FNA was performed via the transgastric approach with linear EUS (GF-UCT240; Olympus Co.) and two passes were made with a 22-gauge needle (EchoTip Ultra, ECHO-22; Cook Endoscopy, Winston-Salem, NC, USA). The solid lesions of the tumor were punctured. No abnormal signs or symptoms were noticed during or after the procedure. Smears were created at the bedside in the endoscopy suite. The cytological findings were hypercellular and characteristically showed branching papillary arrangements composed of delicate fibrovascular cores with at-
Figure 3. EUS showed a low echoic mass (arrowhead) with calcification (arrow) (A). On contrast-enhanced harmonic EUS (CH-EUS) (using sonazoid), the tumor was increasingly enhanced (thin arrow), although it was hypovascular compared with the surrounding pancreatic parenchyma (thick arrow) (B).

Figure 4. EUS-FNA was performed via the transgastric approach and two passes were made with a 22-gauge needle (A). The smears exhibited hypercellular findings and characteristically showed branching papillary arrangements composed of delicate fibrovascular cores with attached monotonous cuboidal neoplastic cells (Giemsa stain, ×200) (B).

tached monotonous cuboidal neoplastic cells (Fig. 4). These findings were strongly suggestive of SPN. We diagnosed the tumor as SPN and performed minimized resection (segmental pancreatectomy). An examination of the resected specimen revealed a solid tumor extending to the body of the pancreas. The tumor contained calcified, hemorrhagic and necrotic areas (Fig. 5). The histopathological findings included degenerative necrosis and the presence of cells with round nuclei that exhibited pseudopapillary growth. The tumor cells demonstrated immunopositivity for CD10, the progesterone receptor and β-catenin (Fig. 6). The final pathological diagnosis was SPN of the pancreatic body. No vascular invasion or infiltrative growth was observed. The tumor margin was negative. Approximately one year and six months has passed since the operation, and we have followed up on the patient periodically.

Discussion

SPN is an exceedingly rare pancreatic tumor with a reported frequency of less than 1% of all pancreatic diseases (2). Most of the reports of SPN are in the form of small case series, with only a few reports involving more than 50 cases identified in the literature (6). SPN predominantly occurs in adolescent girls and young women, with a reported frequency of 87% to 90% of all cases (mean age: 25 to 35 years) (7). The striking predilection for SPN to occur in women suggests a hormonal influence in the pathogenesis of this tumor (8). Cases occurring in the first decade of life are rare, and less than 10% of SPN cases have been reported in patients older than 40 years of age (9). The occurrence of SPN in men is rare, accounting for 7% of all cases (10). The present case occurred in an asymptomatic woman whose age was slightly higher than the typical age of patients with SPN. Most SPN lesions are generally large, with a mean diameter of 10.3 cm, and approximately 72% arise in the body or tail of the pancreas, or less frequently in the head (9). The lesion observed in this case was comparatively large (44 mm in diameter) with a location in the body of the pancreas. In addition, SPNs are generally encapsu-
An examination of the resected specimen showed a solid tumor extending to the body of the pancreas (A). The tumor contained calcified, hemorrhagic and necrotic areas (B).

Histopathological findings. (A) Note the degenerative necrosis in the tumor tissue and tumor cells with round nuclei that exhibited pseudopapillary growth (Hematoxylin and Eosin staining, ×200). (B-D) Immunohistochemically, the tumor cells were positive for CD10 (B), the progesterone receptor (C) and β-catenin (D) (×400).

The tumor characteristics observed on imaging reveal cystography effects; however, these effects are weak compared with those of NET (11). T1-weighted MR images demonstrate low signal intensity, while T2-weighted MR images demonstrate high signal intensity. Fat-suppressed T1-weighted images sometimes partially show very high intensity due to bleeding in the tumor (12). Reports of the use of CE-EUS to evaluate SPNs are limited, and the characteristic findings observed on this modality remain unknown. It has been reported that CE-EUS can be used to visualize the blood flow inside the tumor; however, in this case, the tu-
morb was hypovascular compared with the surrounding pancreatic parenchyma (13).

Based on these findings, this case was first considered to involve SPN, although the patient’s age was slightly higher than the typical age of patients with SPN. In addition, none of the findings were specific for SPN, and NET with cystic degeneration was suspected as a secondary diagnosis. Therefore, we performed EUS-FNA to obtain a definitive diagnosis.

It has been reported that pancreatic NET and SPN frequently pose diagnostic challenges (14). Making the histologic differential diagnosis between SPN and NET is very important because SPN is associated with a much better prognosis than NET, with only 10% to 15% of cases recurring or metastasizing. More than 95% of patients with SPN are cured with complete surgical resection alone (15). In addition, it has been reported that a minimized resection of SPN can achieve favorable curative effects due to the nonaggressive behavior of the tumor, the presence of a dense capsule and the excellent prognosis. At the same time, it is necessary to limit the removal of tissue in order to preserve as much normal pancreatic tissue as possible and maintain the functional structure of the pancreas, as these tumors are usually found in young women, and it is important to consider their postoperative quality of life (16, 17). Obtaining an accurate preoperative diagnosis of SPN enables the use of minimized resection. Therefore, the significance of making a preoperative pathological diagnosis is substantial.

However, definitive preoperative diagnoses are made in only a minority of cases of SPN. Since Bondeson et al. first made a correct diagnosis of SPN on preoperative FNA (18), 57 cases of SPN have been diagnosed based on cytologic findings on percutaneous FNA (18, 19). In the largest study, which incorporated 718 cases from the literature, only 52 patients (7%) received a confirmed preoperative diagnosis of SPN based on the findings of FNA (20). A smaller study reported only a 25% success rate with CT-guided FNA with respect to the diagnosis of SPN (21).

Recently, EUS-FNA has become a useful diagnostic and staging tool for assessing patients with pancreatic tumors. The sensitivity and specificity of EUS-FNA for diagnosing pancreatic neoplasms has been reported to be 91% and 94%, respectively (22). The overall complication rate of EUS-FNA is reported to be <1% in large centers. In one study, among 1,034 patients who underwent pancreatic EUS-FNA, the complications included 10 (0.96%) cases of hemorrhage, two (0.19%) cases of acute pancreatitis and one (0.09%) case of duodenal perforation (23). It has been reported that EUS-FNA is a useful and safe method for evaluating pancreatic masses, with a high feasibility rate even when the lesions are small (24).

Since Nadler et al. first made a correct diagnosis of SPN using EUS-FNA in 2002 (25), additional cases of SPN have been diagnosed based on this method (26, 27). Song et al. summarized the EUS-FNA cytologic features observed in 43 cases of SPN described in the English literature. They reported that FNA cytomorphologic features are highly characteristic and distinct from those of other cystic or solid tumors of the pancreas (26). In addition, the diagnostic accuracy of EUS-FNA for assessing SPN lesions was 75% in another study (28). In that study, minimized resection was performed in 29% of the patients. Furthermore, it has been reported that diagnosing SPN preoperatively using EUS-FNA is feasible.

We experienced a case of pancreatic SPN. NET with cystic degeneration was considered as a differential diagnosis; therefore, we performed EUS-FNA to obtain a definitive diagnosis and were able to perform minimized resection. In conclusion, obtaining a definitive preoperative diagnosis of SPN using EUS-FNA can guide the surgical approach.

The authors state that they have no Conflict of Interest (COI).

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