Synchronous Solid Pseudopapillary Tumor and Nonfunctioning Neuroendocrine Tumor in the Pancreas

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Solid pseudopapillary tumor (SPT) is known to be cured in more than 95% of patients after surgery. However, it is reported that 7% of this tumor can recur after resection, and continued surveillance is needed after surgery for SPT. In this case report, a pancreatic mass which had been detected during a routine health checkup was followed up and surgical resection (distal pancreatectomy) was performed due to an increase in size. Postoperative pathology revealed synchronous SPT and neuroendocrine tumor (NET). At twelve months after surgery, a recurrence of the mass in the head of the remnant pancreas was noted and further surgical resection (total pancreatectomy) was performed. Postoperative pathology confirmed the diagnosis of malignant SPT. To date, there are only a few reports of the synchronous SPT and NET, and there are no reports in Korea. Therefore, we report a case of the synchronous SPT and nonfunctional NET diagnosed after surgery.

Keywords: Pancreatic neoplasms; Solid pseudopapillary tumor; Neuroendocrine tumors; Multiple primary neoplasm; Recurrence

INTRODUCTION

Solid pseudopapillary tumor (SPT) is known to be cured in more than 95% of patients after surgery, with complete resection in most cases.¹ However, it is reported that 7% of this tumor can recur after resection.¹

To date, there are only a few reports of synchronous SPT and neuroendocrine tumor (NET)²,³ and there are no reports in Korea. Therefore, we aimed to present a case of synchronous SPT and nonfunctioning NET with review of previous literatures. The Institutional Review Board of Boramae Medical Center approved this study (IRB No. 20-2024-15).
CASE

A 76-year-old man visited to our institution for a pancreatic mass found on an abdominal ultrasonography and computed tomography (CT) scan as part of routine health checkup at other hospital. A 1 cm-sized hypoechoic mass in the body of the pancreas was observed on an abdominal ultrasonography (Fig. 1A, and this lesion was observed as a 1 cm-sized high attenuating mass on an abdominal CT scan (Fig. 1B). There was no abdominal pain, weight loss, diarrhea, sweating, or facial flushing in this patient.

In initial examination in our institution, serum carbohydrate antigen 19-9 level was 20 U/mL, and abdominal magnetic resonance imaging showed a 1 cm-sized mass which showed high signal intensity in T2-weighted imaging, iso signal intensity in T1-weighted imaging, and subtle restriction in diffusion-weighted imaging. These findings are compatible with those of pancreatic NET (Fig. 2). With the of suspicion of pancreatic NET, we decided to perform regular surveillance with abdominal CT scan.

Abdominal CT at 4 and 8 months showed no change in the pancreatic mass. However, abdominal CT at 12 months showed an increase in the size of the mass to 2.4 cm with edema, inflammation, and multiple pseudocysts in the tail of the pancreas (Fig. 3). Subsequently, distal pancreatectomy and cholecystectomy were performed. At the time of surgery, severe adhesions due to inflammation were found in the pancreatic body and tail, and an approximately 5 cm-sized mass was observed in the area. Postoperative pathology revealed severe inflammation and multiple pseudocysts with pancreatic ductal destruction, accompanied by scattered tumor nests (Fig. 4). In addition, tumor cells showed trabecular arrangements and salt and pepper chromatin pattern was noted (Fig. 5A). Cell division level was less than 1 per 10 high power field. Immunohistochemical staining

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**Fig. 1.** Cross-sectional image findings. (A) Transabdominal ultrasound done at another hospital showed a 1 cm-sized hypoechoic mass in the pancreatic body. (B) Contrast-enhanced abdominal computed tomography demonstrated a 1 cm-sized enhancing mass in the pancreatic body and multiple gallstones.

**Fig. 2.** Magnetic resonance imaging of the abdomen. In the pancreatic body, there was a 1 cm-sized mass which showed high signal intensity in T2-weighted imaging, iso signal intensity in T1-weighted imaging, and subtle restriction in diffusion-weighted imaging. These findings are compatible with those of pancreatic neuroendocrine tumor.

**Fig. 3.** Contrast-enhanced abdominal computed tomography done 12 months later. The pancreatic body mass has increased in size to 2.4 cm. There were edema, inflammation, and multiple pseudocysts in the tail of the pancreas.
showed CD56 and synaptophysin positivity, chromogranin negativity, and Ki-67 less than 1% positivity (Fig. 5B). Therefore, the diagnosis of grade 1 pancreatic NET could be made.

Abdominal CT scan performed one week after surgery still showed a 2.2 cm-sized, high attenuating mass in the body of the pancreas and second surgery was performed to resect the residual mass. Microscopic examination of the resected mass revealed a tumor with papillary growth and with cystic degeneration, voluminous cytoplasm, and hyaline globules. Immunohistochemical staining showed CD10 positivity, but Ki-67, synaptophysin, and chromogranin negativity. The diagnosis of pancreatic SPT could be made at postoperative pathology. In addition, there were multiple tumor nests around the SPT, which were diagnosed as residual NETs (Fig. 6).

There was no tumor recurrence at abdominal CT scan performed 6 months after the second surgery. However, abdominal CT scan performed 12 months after the second surgery showed a high attenuating mass in the head of the pancreas. This mass in the head of the pancreas had not been observed in abdominal CT scans performed before surgery. Follow-up with abdominal CT scan was performed at 3-month intervals. During the follow-up, the pancreatic head mass increased consistently (Fig. 7A), and 18F-fluorodeoxyglucose positron emission

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**Fig. 4.** Microscopic findings after initial distal pancreatectomy under lower power. There was severe inflammation with pancreatic ductal destruction which was accompanied by scattered tumor nests (red arrows) (Hematoxylin and eosin stain, x50).

**Fig. 5.** Microscopic findings after distal pancreatectomy under higher power and with immunohistochemical staining. (A) Tumor cells showed trabecular arrangements. Salt and pepper chromatin pattern was noted (Hematoxylin and eosin stain, x400). (B) Immunohistochemical staining were positive for CD56 and synaptophysin, but negative for chromogranin.

**Fig. 6.** Microscopic examination of the resected mass after second resection. (A) Tumor cells showed papillary growth pattern (Hematoxylin and eosin stain, x12). (B) Tumor cells showed cystic degeneration (Hematoxylin and eosin stain, x100). Also, the tumor cells contain voluminous cytoplasm and hyaline globule (Hematoxylin and eosin stain, x400). (C) Multiple tumor cell nests of neuroendocrine tumor are noted around the solid pseudopapillary tumor (Hematoxylin and eosin stain, x12).

**Fig. 7.** Image findings done 24 months after second surgery. (A) Contrast-enhanced computed tomography of the abdomen showed a 2.7 cm-sized high attenuating mass in the head of the pancreas. (B) 18F-fluorodeoxyglucose positron emission tomography demonstrated a hypermetabolic lesion in the remnant pancreatic head.
tomography performed 24 months after surgery showed hypermetabolic uptake in the remnant pancreatic head (Fig. 7B). Serum carbohydrate antigen 19-9 level was 380 U/mL. Therefore, additional pancreaticoduodenectomy resulting in total pancreatectomy was performed, and postoperative pathology revealed a malignant SPT.

After the third surgery, the patient was discharged without any complications, and underwent regular surveillance with abdominal CT scan. There was no evidence of tumor recurrence on subsequent abdominal CT scans up to 18 months after the third surgery.

**DISCUSSION**

SPT of the pancreas is a low-grade malignancy with histopathologic findings of alternating solid, pseudopapillary, or cystic components. Most SPT (90%) occur in female, and the frequency of the SPT is 5% among pancreatic cystic tumors, and 2% of pancreatic exocrine tumors. All SPTs are considered as low-grade malignant tumors, and according to previous studies, the risk of malignancy in the SPTs is known to be 4.8 to 76.6%.

The diagnosis of SPT can be made with microscopic histopathology after surgery in most cases. In some cases, however, immunohistochemical staining required to differentiate from acinar tumors, pancreatoblastoma, NET, and adenocarcinoma. In most cases, immunohistochemical staining is sufficient for differentiation of these tumors.

Although various types of immunohistochemical staining have been performed to determine the origin of the SPT, the exact origin of the SPT was not determined to date. The SPTs are positive for α1-antitrypsin and α1-antichymotrypsin, indicating an exocrine origin, and some SPTs are also positive for neuron-specific enolase and synaptophysin, indicating an endocrine origin. They have a low mitotic index and are stained poorly for Ki-67, but are mostly positive for vimentin staining. Other markers of endocrine origin and pancreatic enzymes, as well as epithelial membrane antigen and chromogranin A, are also absent in the SPT.

To date, only a few studies have reported the cases of synchronous pancreatic SPT and NET. One study reported a case of a 16-year-old male patient with hypoglycemic episodes and multiple endocrine neoplasia type 1, who was diagnosed with SPT and functional NET (insulinoma) after surgery. In a study with 20 patients who underwent surgical resection for pancreatic masses and was diagnosed as SPT on postoperative pathology, immunohistochemical staining showed vimentin and CD10 positivity in all patients, and a synchronous 2 cm-sized nonfunctional NET was observed around a 4 cm-sized SPT in one patient, a 63-year-old woman.

Although we did not performed tests for a functionality of NET in this case, we believe that it was a non-functional NET because the patient had no symptoms of endocrine dysfunction such as abdominal pain, weight loss, diarrhea, sweating, and hot flashes, and the patient was relatively old (over 70 years). In addition, due to severe inflammation and adhesions in the pancreas, it was difficult to accurately identify the extent of the mass at the time of the initial surgery, so it was not possible to resect all of the pancreatic masses at the initial surgery and additional secondary surgery was required for complete resection of the tumor. Therefore, when performing surgical resection of pancreatic masses, it is important to keep in mind that multiple synchronous masses can occur within the pancreas. And, in case of difficulty to accurately evaluate the extent of the pancreatic mass with CT or MR due to inflammation around the mass, it might be better to consider endoscopic ultrasonography before surgery to accurately evaluate the extent of the mass.

SPT is known to be cured in more than 95% of patients after surgery, with complete resection in most cases. However, about 7% of cases have been reported to recur after resection. In this case, SPT recurred 12 months after surgery, and this recurred mass had not been observed in imaging studies performed before surgery. So, additional surgery was required to treat the recurred SPT. Therefore, continued surveillance might be essential after surgery for SPT.
요 약

고형가유두상종양은 수술 후 95% 이상의 환자에서 완전 절제가 이루어지며 대부분 완치되는 것으로 알려져 있다. 하지만, 절제 후 재발하는 경우가 7% 정도로 보고되고 있어 수술 후에도 지속적인 추적 관찰이 필요하다. 또한,췌장 내에서 다발성으로 종괴가 발생할 수 있으므로,췌장 종괴에 대한 수술적 절제를 시행할 경우 수술 전에 반드시 영상 검사와 내시경 초음파 등을 통한 총분한 평가가 선행되어야 한다. 본 증례 보고에서는 간질 검진으로 발견된 썰장 종괴에 대해 추적 관찰을 시행하다가 크기가 증가해서 수술적 절제(췌장과부절제술)를 시행하였다. 수술 후 병리 결과에 의해 고형가유두상종양과 신경내분비종양이 동시에 진단되었다. 수술 12개월 후 남아 있는 웅장 두부에 종괴가 재발한 소견을 보여 추가적인 수술적 절제(전췌장절제술)을 시행 받았고 수술 후 병리 결과로는 악성 고형가유두상종양으로 진단되었다. 현재까지 고형가유두상종양과 신경내분비종양이 동시에 발생하는 증례에 대한 보고가 많지 않고 국내에서는 아직 보고가 없다. 이에 수술 후 고형가유두상종양과 비기능성 신경내분비종양이 진단된 증례를 보고하는 바이다.

국문 색인: 썰장 종양; 고형가유두상종양; 신경내분비종양; 다발성 원발 종양; 재발

Conflicts of Interest

The authors have no conflicts to disclose.

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