Clinical Case Report

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Concurrent ductal adenocarcinoma, pseudocyst, and neuroendocrine tumor of the pancreas

A case report

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Abstract

Introduction: Pancreatic pseudocyst is one of the most common cystic lesions. It always occurs following pancreatitis and is rarely found in combination with pancreatic adenocarcinoma. The coexistence of exocrine and neuroendocrine tumors of the pancreas is also infrequent. We herein report a case of simultaneous occurrence of a pancreatic ductal adenocarcinoma (PDAC), pseudocyst, and neuroendocrine tumor (NET), showing a "side-by-side pattern."

Patient concern: A 74-year-old man was hospitalized for epigastric pain and poor appetite. He had no history of pancreatitis, alcohol consumption, or smoking.

Diagnosis and intervention: Abdominal enhanced computed tomography and magnetic resonance imaging revealed a 15 × 8 cm cystic lesion with poor enhancement located in the tail of the pancreas. The distal aspect of the main pancreatic duct was dilated. The pancreatic parenchyma adjacent to the cystic lesion showed slightly heterogeneous enhancement on computed tomography and magnetic resonance imaging. Laboratory examination showed an elevated carbohydrate antigen 19–9 serum level. The patient was preoperatively diagnosed with intraductal papillary mucinous neoplasm and subsequently underwent laparotomy. During the operation, a hard white tumor measuring about 4 × 3 cm was palpated adjacent to the cystic lesion on the duodenal side, and a 0.6-cm nodule was simultaneously found in the pancreatic tail. Therefore, total pancreatectomy and splenectomy were performed. Histopathological examination showed that the tumor was PDAC with an adjacent pseudocyst, and the small nodule was suggestive of a NET.

Outcomes: The patient survived without recurrence or metastasis in the follow-up visit 10 months after the operation and adjuvant chemotherapy.

Conclusions: The concomitant occurrence of a PDAC, pseudocyst, and NET has not been previously reported. We suggest that if a pancreatic cyst is found, the coincidental occurrence of a malignant tumor should be considered, especially if the carbohydrate antigen 19–9 level is increased. Additionally, dilation of the pancreatic duct may be a diagnostic clue. Furthermore, the simultaneous occurrence of pancreatic endocrine and exocrine tumors is very uncommon. Preoperative diagnosis becomes difficult because of the lack of specific symptoms and radiological features.

Abbreviations: CA19-9 = carbohydrate antigen 19-9, CT = computed tomography, IPMN = intraductal papillary mucinous neoplasm, MPD = main pancreatic duct, MRI = magnetic resonance imaging, NET = neuroendocrine tumor, PDAC = pancreatic ductal adenocarcinoma.

Keywords: adenocarcinoma, neuroendocrine tumor, pancreas, pseudocyst

1. Introduction

The accurate preoperative diagnosis of pancreatic cysts is a clinical challenge.[1] Pancreatic pseudocyst is one of the most common cystic lesions. Approximately 75% to 90% of pseudocysts occur following pancreatitis and trauma, and they are rarely found secondary to neoplasms.[2] In the present report,

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All data generated or analyzed during this study are included in this published article [and its supplementary information files].

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we describe a pseudocyst that was accompanied by a pancreatic ductal adenocarcinoma (PDAC).

PDAC is one of the most common malignant tumors worldwide, while neuroendocrine tumors (NETs) are relatively rare. NETs represent approximately 1% to 2% of all pancreatic neoplasms.[3,4] The incidence of these combined neoplasms reportedly ranges only from 0.06% to 0.20% among all pancreatic tumors.[5] Additionally, the combination of PDAC and NET is extremely rare.[6,7] We herein report a highly unusual case of the coexistence of a PDAC, pseudocyst, and NET, showing a "side-by-side pattern."

2. Clinical data

This study was approved by the clinical research ethics committee of our hospital. The patient provided informed consent for publication of the case. A 74-year-old man was hospitalized for epigastric pain and poor appetite. He had no history of pancreatitis, alcohol consumption, or smoking. Laboratory examination revealed a high carbohydrate antigen 19-9 (CA19-9) serum level (187.2 U/ml; reference range, 0–37 U/ml). The pancreatic amylase blood level was slightly elevated (204 IU/L; reference range, 0–135 IU/L). The immunoglobulin G4 and carcinoembryonic antigen levels were within the normal limit.

Abdominal enhanced computed tomography (CT) examination revealed a multilocular cystic lesion of about 15×8 cm in the tail of the pancreas (Fig. 1A). The pancreatic parenchyma adjacent to the cystic lesion showed slightly heterogeneous enhancement on CT (Fig. 1A), and the distal aspect of the main pancreatic duct (MPD) was dilated (Fig. 1B). Additionally, a small nodule measuring 0.6 cm in diameter with severe enhancement in the arterial phase was located in the tail of the pancreas (Fig. 1C). Unfortunately, we did not preoperatively investigate the nodule because it was too small to be detected. Magnetic resonance imaging (MRI) revealed similar findings. An abnormal lesion in the body of the pancreas showed hypointensity on T1-weighted imaging and slight hyperintensity on T2-weighted imaging (Fig. 1D). However, no obvious high-intensity signal was seen on diffusion-weighted imaging. Slight heterogeneous enhancement of the lesion was observed on contrast MRI (Fig. 1E). A multilocular cyst without enhancement was found beside the lesion (Fig. 1E). However, the small nodule observed on CT was not found on MRI. One possible reason may have been that the MRI slice was too thick (5 mm) to detect the small lesion. The magnetic resonance cholangiopancreatography findings are shown in Fig. 1F. A diagnosis of intraductal papillary mucinous neoplasm (IPMN) was made preoperatively according to the septum of the cystic lesion and the dilation of the MPD. Based on the imaging findings and the elevated CA19-9 serum level, the patient underwent laparotomy. During the operation, a hard white tumor of about 4×3 cm was palpated adjacent to the cystic lesion on the duodenal side, and a 0.6-cm nodule was incidentally found in the pancreatic tail. Therefore, total pancreatectomy and splenectomy were carried out. Neither liver metastasis nor peritoneal seeding was found.

Histopathological examination revealed a ductal cell carcinoma in the body of the pancreas with an adjacent pseudocyst. The small nodule in the tail of the pancreas was suggestive of a grade 1 NET. Immunohistochemical analysis of the endocrine cells revealed the following: synaptophysin (+), chromogranin A (+), cytokeratin (+), and Ki-67 proliferation index of 2%. In the area of the ductal carcinoma, immunochemical staining for cytokeratin 19, CA19-9, CD31 and carcinoembryonic antigen was positive. The Ki-67 proliferation index was 15%. After surgery, the patient received gemcitabine-based adjuvant treatment. Currently, the patient has no signs of relapse or metastasis 10 months after surgery.

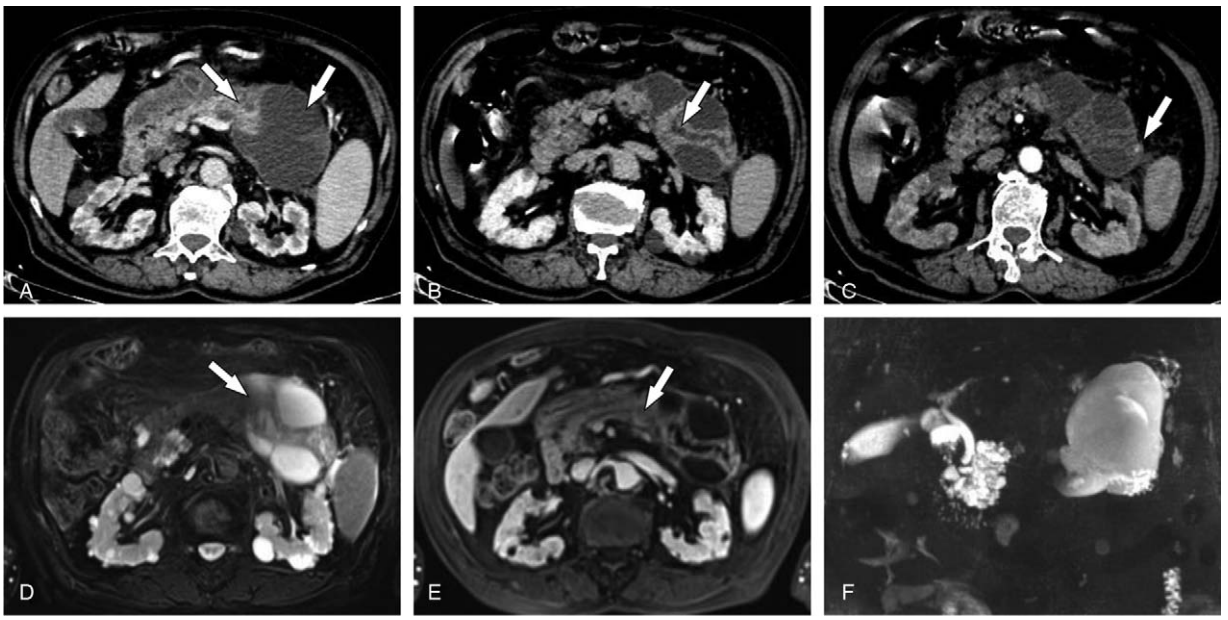

Figure 1. Abdominal CT, MRI, and MRCP images. CT showed (A) a massive multilocular cyst in the tail of the pancreas with (B) dilation of the MPD. (A) A slightly heterogeneously enhanced lesion was found in the body of the pancreas. (C) A small nodule measuring 0.6 cm in diameter with severe enhancement in the arterial phase was located in the tail of pancreas (arrow). MRI showed (D) a hyperintense lesion on T2-weighted imaging in the body of the pancreas with (E) slightly heterogeneous enhancement. (F) MRCP revealed a cyst in the pancreatic tail. The distal aspect of the MPD was dilated. CT = computed tomography, MPD = main pancreatic duct, MRCP = magnetic resonance cholangiopancreatography, MRI = magnetic resonance imaging.

3. Discussion

We have herein reported the first case of concomitant occurrence of a PDAC, pseudocyst, and NET. The preoperative work-up was consistent with IPMN as evidenced by the multilocular cyst and dilation of the MPD. Pathological examination revealed a massive pseudocyst secondary to a PDAC of the pancreatic body, accompanied by a small NET in the tail of the pancreas, showing a "side-by-side pattern."

The association between pseudocyst and pancreatic carcinoma has been previously reported but is exceedingly rare.[2,8–10] Fujiwara[8] described a pancreatic adenocarcinoma of 6 mm in diameter that caused acute pancreatitis and a pseudocyst. Kimura et al[10] reported 2 cases of pancreatic carcinoma accompanied by a pseudocyst. In one case, the tumor was too small to be detected. The other case was misdiagnosed as a cystadenocarcinoma because of the septum and protuberant nature of the cyst. Most pancreatic carcinomas are believed to arise in the ductal system. As the carcinoma grows, the pancreatic duct becomes obstructed from the high intraductal pressure, causing proximal dilatation and rarely pseudocyst formation.[9,10] Pseudocysts are usually unilocular and almost always located outside of the pancreas.[11] In our case, the pseudocyst was multilocular and located inside of the pancreas, accompanied by distal dilation of the MPD. It was misinterpreted as an IPMN because we failed to accurately assess the adjacent pancreatic parenchyma of the pseudocyst. The mass lesion suggestive of pancreatic cancer was regarded as part of the IPMN. In fact, an elevated CA19-9 level is a strong indicator of malignancy. Although the CA19-9 level increases even in patients with chronic pancreatitis, our patient had no evidence of such an inflammatory condition.

Cases of concomitant or collision tumors in the pancreas have rarely been reported. Such rare cases include the association of IPMN and NET,[3,12–14] solid pseudopapillary neoplasm and NET,[15] pancreatic and periampullary collision cancers,[16] and mixed adenoneuroendocrine carcinoma.[17] In most cases, the coexistence of these tumors is incidentally discovered through the postoperative pathology. Only 2 reported cases of collision tumors involved a PDAC and a NET.[6,7] In one case, the preoperative diagnosis was a main-duct IPMN. The pathologic examination revealed a PDAC involving the whole pancreas and a NET located in the head of the pancreas.[6] In the other case, the preoperative diagnosis was consistent with a typical NET. Gross pathologic examination revealed neuroendocrine cells with ductal carcinoma without a transition zone.[7] In the present case, the PDAC and NET were not collision tumors; they were discontinuous and independent of each other. We did not observe the NET in the tail of the pancreas preoperatively because it was too small to be found on CT or MRI, and the patient had no symptoms of hormone production such as hypoglycemia, gastrointestinal ulceration, or diarrhea.

In conclusion, we have herein reported a new case of adenocarcinoma coexisting with a pseudocyst and NET, a very rare association that was misinterpreted as an IPMN. The mechanism of their concomitant evolvement is not clear. We suggest that if a pancreatic cyst is found, the coincident occurrence of a malignant tumor should be considered, especially when the CA19–9 level is increased. Additionally, dilation of the MPD may be a diagnostic clue. Furthermore, the simultaneous occurrence of pancreatic endocrine and exocrine tumors is very uncommon. When present, the preoperative diagnosis becomes difficult because of the lack of specific symptoms and radiological features.

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Author contributions

Writing – original draft: Yu Liu.

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