CASE REPORT

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Pancreatic Neuroendocrine Tumor Mimicking Intraductal Papillary Mucinous Neoplasm: Case Report

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ABSTRACT Pancreatic neuroendocrine tumors (PanNETs) are rare pancreatic tumors. They usually exhibit parenchymal growing, however some cases can exhibit intraductal growing. PanNET with intraductal growth may cause intraductal papillary mucinous neoplasm (IPMN)-like clinic scenario by presenting as cystic formations secondary to duct obstruction. In our case, a 69-year- old man with a history of abdominal pain and nausea underwent a computed tomography scan that showed dilated pancreatic duct and cystic lesion which was 8 cm originating from the pancreas. Imaging and laboratory findings were considered to be consistent with an IPMN so the patient underwent distal pancreatectomy and splenectomy. However, the pathological examination of the surgical specimen showed a millimeter-sized PanNET located in pancreatic tail mimicking the IPMN by obstructing the pancreatic duct.

Keywords: Gastroenteropancreatic neuroendocrine tumor; pancreatic intraductal neoplasms

Pancreatic neuroendocrine tumors (PanNETs) are rare pancreatic tumors that are thought to originate from pancreatic islet cells. The incidence of PanNETs are approximately 0.5 cases per 100,000 and present 1-2% of all pancreatic neoplasms.[1,2] Although they generally show slow growth, they have a high potential for nodal and distant organ metastasis.[3] Surgical therapies are the first-line treatments for PanNET patients.[1]

Intraductal papillary mucinous neoplasms (IPMNs) are described as cystic dilatation of pancreatic ducts in which an intraductal proliferation of neoplastic mucin-producing cells is usually constructed in papillary patterns.[4] They are considered as premalignant lesions and surgical resection is the primary treatment method in many cases.[5] In some cases, pancreatic tumors with extensive ductus involvement may cause a clinical scenario like IPMN that termed pseudo-IPMN.[6]

PanNET with intraductal growth is an unusual clinical condition and may cause IPMN-like clinic scenario by cystic formations secondary to duct obstruction.[2] Here, we report a case of pseudo-IPMN, which is a very rare presentation of PanNET.

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A 69-year-old male patient was hospitalized with abdominal pain and nausea/vomiting. On physical examination, no pathological findings were found, biochemistry parameters were evaluated as normal. Abdominal computed tomography (CT) was reported as a 9x6 cm cystic lesion on the pancreas body (Figure 1A, B). Magnetic resonance images (MRI) revealed a cystic lesion which was 8 cm originating from the pancreas. The lesion was localized in the bursa omentalis and extended towards the splenic hilum as a grape bunch with solid areas (Figure 1C, D). Fine needle aspiration (FNA) was performed from a 70x58 mm cystic lesion in the pancreatic body on endoscopic ultrasonography (EUS). CA 19-9 level was measured as 150,000 U/ml, carcinoembryonic

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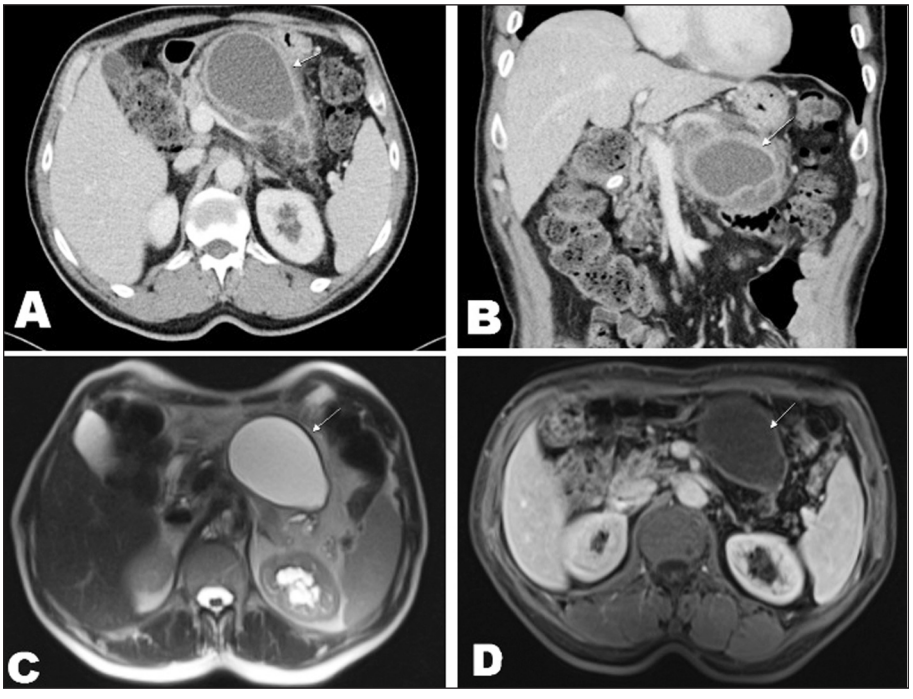

FIGURE 1: Axial contrast-enhanced computed tomography scan (A) and coronal contrast-enhanced computed tomography scan (B) show cystic mass with septations and thick wall in the corpus of pancreas (yellow arrow). Axial T2-weighted fat-saturated magnetic resonance imaging scan (C) and axial contrast-enhanced magnetic resonance imaging scan (D) show cystic lesion in the corpus of pancreas (yellow arrow).

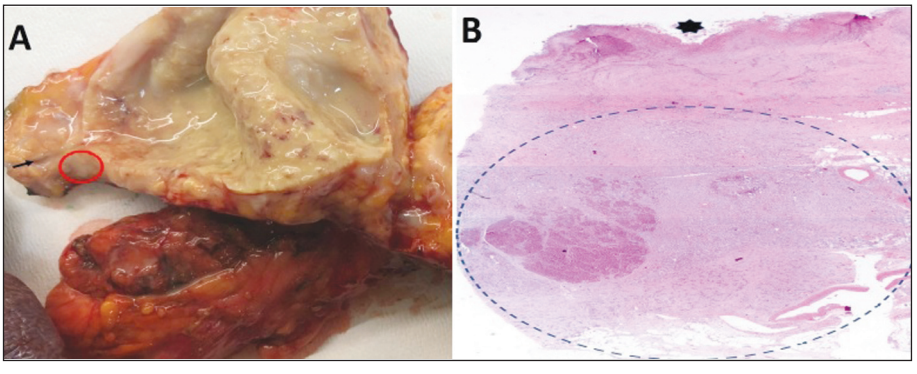

FIGURE 2: A) The area in the red circle shows pancreatic neuroendocrine tumor (PanNET) and the black arrow shows duct, obstructed by PanNET. B) The area in the circle shows PanNET and the black star shows duct, obstructed by PanNET (H&E stain, x10).

antigen (CEA) was 97.15 ng/mL and amylase level was 19,490 U/L in cyst fluid sampling. The cytological evaluation was reported as non-diagnostic. Serum carbohydrate antigen 19-9 (CA 19-9) level was measured as 86.6 U/mL and CEA level was measured 97.15 ng/mL. The patient was discussed at the multidisciplinary oncology-surgery council and resection decision was made with the pre-diagnosis of malignant IPMN. Distal pancreatectomy and splenectomy were performed. The histopathological diagnosis of the patient was reported as PanNET with intraductal growth, which was of 8 mm diameter and located in the pancreatic tail. Immunohistochemical staining with chromogranin A, synaptophysin, somatostatin receptor 2 was positive in the tumor mass. However, the tumor was not stained with any of the specific hormone markers. The tumor has a Ki-67 labeling index of less than 2%. According to the 2017 World Health Organization Classification, a definitive diagnosis of well-differentiated, non-functional low grade neuroendocrine tumor of the pancreas was made. There was a pseudocystic degeneration area which was 10 cm of diameter adjacent to the tumor, lined with benign mucinous epithelium without dys-

plasia. In fact, it was considered that PanNET obstructed the lateral branch duct of the pancreatic tail and caused cystic degeneration, which mimicked the IPMN (Figure 2A, B). No pathological involvement was detected in Gallium 68 positron emission tomography/CT in the postoperative period. The patient was asymptomatic at the outpatient control at the 3rd, 6th, and 12th postoperative months, no recurrence was found in the abdominal MRI at the 12th month. Informed consent was obtained from the patient for the case report.

DISCUSSION

IPMNs can be examined under three main types as IPMN-main duct type, IPMN-branch duct type and IPMN-mixed type. Surgical resection is the primary treatment method in IPMN-main duct type due to the high risk of malignancy. However, surgical resection or follow-up can be preferred in IPMN-branch duct type by evaluating the patient's symptoms, biochemical, and radiological parameters.[5] High-risk signs are important for surgical decision-making during follow-up. High-risk signs can be listed as cyst size, mural nodules, presence of solid components, high serum level of CA 19-9, dilated pancreatic duct, amount of annual growth, new-onset diabetes mellitus, and positive cytology.[5] In our case, cyst size greater than 3 cm, solid component and high serum CA 19-9 were the determinants in surgical decision making with the provisional diagnosis of malignant IPMN. However, the pathological examination of the surgical specimen has shown that the millimeter sized PanNET mimicked the IPMN by pancreatic duct obstruction.

PanNETs are generally described as well-circumscribed and parenchymal-growing tumors. PanNETs with intraductal growth are uncommon. Previous studies showed that lesions may radiologically and clinically mimic IPMNs and represent a challenge in the preoperative differential diagnosis of pancreas cystic lesions. In the review by Manuel-Vazquez et al., intraductal growing panNETs are reported as in our case are very rare, and 20 cases have been reported.[2] In 13 of 20 patients, the preoperative diagnosis was not specified, 5 of the 20 patients were operated with the provisional diagnosis of IPMN, and 2 were operated with the suspicion of malignancy.[7-14]

The final diagnosis was completed on surgical resection specimens in our case. Nevertheless, preoperative diagnosis of PanNETs may be a clinical necessity in metastatic or local advanced tumors. It has been shown that metastatic or local advanced PanNETs benefited from aggressive surgical approach conversely to malignant IPMNs. EUS and FNA may be helpful for diagnosing and grading millimeter sized PanNETs.[1] However, as in our case, even though FNA is a method for diagnosing intraductal materials, it may not be diagnostic due to insufficient sample size.

In conclusion, preoperative accurate diagnosis of intraductal growing PanNETs are difficult. PanNETs should be kept in mind in the differential diagnosis of intraductal pancreatic lesions since they may have potential benefits for the management and follow-up of these patients.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Ahmet Akmercan;* ***Design:*** *Ahmet Akmercan, Tevfik Kıvılcım Uprak;* ***Control/Supervision:*** *Cumhur Yeğen, Pelin Bağcı, Ali Emre Atıcı;* ***Data Collection and/or Processing:*** *Ahmet Akmercan, Pelin Bağcı;* ***Analysis and/or Interpretation:*** *Tevfik Kıvılcım Uprak, Ahmet Akmercan, Ali Emre Atıcı;* ***Literature Review:*** *Ahmet Akmercan, Tevfik Kıvılcım Uprak;* ***Writing the Article: Critical Review:*** *Ahmet Akmercan, Tevfik Kıvılcım Uprak;* ***References and Fundings:*** *Cumhur Yeğen, Ali Emre Atıcı.*

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