Case Report

Neuroendocrine tumor within main pancreatic duct: a case report

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Abstract
Background: Pancreatic neuroendocrine tumor (NET) is a well-known, but uncommon disease that manifests as a heterogeneous, intense, round mass on enhanced computed tomography (CT). Herein, we report a case of pancreatic NET with unusual CT manifestation. A 66-year-old female was referred to our hospital for treatment of intermittent back pain. CT revealed a diffusely swollen pancreas from the head to tail. Imaging enhancement was weak except for a small area at the pancreatic head, and the lumen of the main pancreatic duct seemed narrowed. The entire pancreas showed accumulation of fluoro-D-glucose (FDG) on positron emission tomography (PET), suggesting autoimmune pancreatitis; however, endoscopic ultrasound-fine needle aspiration revealed atypical cells with round nuclei in a rosette formation, resulting in a diagnosis of non-functioning NET. Pathological examination revealed tumor growth within the main pancreatic duct with large areas of necrosis, and an area of viable cells at the head of the pancreas. In conclusion, CT imaging depicted an NET within the main pancreatic duct as a diffusely enlarged pancreas mimicking autoimmune pancreatitis.

Keywords: Pancreatitis, neuroendocrine tumor, pancreatic tumor

Introduction

Pancreatic neuroendocrine tumor (NET) is an uncommon disease comprising 1-2% of pancreatic malignancies1. Radical surgery for pancreatic NET offers a 5-year survival rate of 45-92%1, highlighting the importance of accurate diagnosis and appropriate referring of a patient for surgery. Herein, we describe a case of pancreatic NET with unusual finding on enhanced computed tomography (CT).

Clinical data

A 66-year-old female was referred to our hospital for treatment of intermittent back pain and enlarged pancreas. Her past medical history included stones of the common bile duct and gall bladder treated 20 years previously. She was on medication for type II diabetes mellitus and hypertension. Her blood tests revealed normal levels of various tumor markers, IgG4, and pancreas-related hormones including gastrin, insulin, and vasoactive intestinal peptide, but slightly elevated hemoglobin A1c.

Enhanced CT revealed a diffusely enlarged pancreas from the head to tail (Fig. 1A & B). The main pancreatic duct (MPD) appeared slightly and diffusely narrowed, and a 1-cm area in the head of the pancreas was strongly enhanced (Fig. 1B). There was no obvious enlargement of lymph nodes around the pancreas, but positron emission tomography (PET)-CT showed accumulation of fluoro-D-glucose (FDG) throughout the pancreas (Fig. 1C & D).

Endoscopic retrograde cholangiopancreatography (ERCP) and endoscopic ultrasound-fine needle aspiration (EUS-FNA) confirmed a pancreatic tumor with extensive intraductal growth, comprising atypical cells with small round nuclei (Fig. 2A & B). The tumor was tentatively diagnosed as non-functioning NET, and total pancreatectomy with regional lymph node dissection was performed.

Gross examination of the resected specimen revealed tumor filling the main pancreatic duct from the tail to the head of the pancreas (Fig. 3). Histologically, the tumor was composed of atypical cells with round nuclei arranged within small nests and cords, and immunopositivity for synaptophysin and chromogranin A. Most of the tumor was necrotic, with only a sheet of NET and pancreatic parenchymal cells observed around the outer

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Fig. 1  Enhanced computed tomography and FDG-PET-CT
(A) Abnormally enlarged pancreas with sausage-like appearance is shown. The lumen of the main pancreatic duct (arrowhead) can be identified in this image. The present case showed neither a dotted enhancement nor capsule-like rim. (B) The pancreatic head showed strongly enhanced lesion at the pancreatic phase, which could be retrospectively diagnosed as enhanced tumor. (C & D) Strong accumulation of FDG is visible throughout the pancreas, suggesting the diffuse involvement of pancreatic tissue.

Fig. 2  ERCP and cytology
(A) ERCP demonstrated a mass lesion within the main pancreatic duct (MPD). Visualizing the MPD at the pancreatic body and tail was hampered due to the history of EST and occupying lesion. (B) The EUS-FNA showed atypical cells with small round nuclei that are formed into nests.
edge. A tumor area with viable cells was observed at the head of the pancreas, which was compatible with the enhanced CT findings. No lymph node metastasis was identified. The patient has shown no sign of recurrence for two years post-discharge.

**Discussion**

Pancreatic NETs are rare, with an incidence of 0.4 to 1 per 100,000 people\(^2\). Chetty et al.\(^3\) noted that such tumors are morphologically distinctive and that diagnostic problems are scarce despite their rarity, with one of the unusual features of pancreatic NET being tumor growth predominantly within the main pancreatic duct. The few cases reported in the literature have been divided into two categories\(^3\): those showing intraductal tumor accompanied by a pancreatic parenchymal component, and those comprising purely intraductal pancreatic NET
(without a parenchymal component). In the former case, the NET arising in the vicinity of the main pancreatic duct could grow into and along the duct, while the latter type might theoretically have derived from totipotential stem cells located in the epithelium of major ducts. In the present study, the majority of the tumor was necrotic and determining the pathway of tumor occurrence was difficult.

Nanno et al. analyzed the relationship between ductular involvement and prognosis for well-differentiated NET. In their study, MPD involvement was defined as stenosis of the duct in the tumor mass and up-stream dilatation. Most importantly, the authors revealed that stricture of the MPD was an independent factor associated with nodal metastasis at the time of surgery and subsequent tumor recurrence during follow-up, with a 5-year recurrence-free rate of only 41%. In contrast, the present case involved the entire MPD, although the duct was not trapped within the tumor. Thus, unlike the NET described by Nanno et al., our patient’s tumor was not related to lymph node metastasis, lymphatic invasion, or perineural invasion. Whether such exclusivity of NET growth within the MPD is associated with the better prognosis remains to be clarified in the future. In the present case, regional lymph node dissection was performed due to large tumor size, although the preoperative CT did not show enlarged lymph nodes. As the result, no lymph node metastasis was identified. Again, whether regional lymph node dissection can be omitted in a case similar to ours has to be determined in the future.

Difficulties of distinguishing autoimmune pancreatitis from pancreatic malignancies on CT have been reported previously, particularly in cases of focal autoimmune pancreatitis, which shows partial pancreatic enlargement and abnormal enhancement that mimics pancreatic adenocarcinoma or hypovascular NET. In addition, negative cytology results do not exclude the possibility of pancreatic tumors, resulting in unnecessary pancreatic resections. In contrast to previous studies, distinguishing NET from autoimmune pancreatitis was quite difficult in the present study. Instead of the typical CT presentation of homogenous and strongly enhanced round tumor within the pancreatic parenchyma, the present case showed diffuse enlargement of the pancreas with relatively heterogeneous weak enhancement. We attributed this atypical finding on CT to ischemia of the tumor within the MPD. We also assume that the tumor was not necrotic when imaged by CT and FDG-PET-CT because enhancement and accumulation of FDG was observed, respectively. This conclusion is partially supported by the fact that viable cells were obtained from pancreatic body on EUS-FNA. In contrast, only a small part of the tumor at the head of the pancreas showed strong enhancement, and this could be considered a diagnostic feature in the present study.

In conclusion, we report a patient with pancreatic NET growing predominantly within the MPD that radio logically mimicked autoimmune pancreatitis. Long-term follow up and accumulation of this rare type of NET is required to elucidate the clinical course.

Statement of Authors Contribution

All authors have participated sufficiently in the work to take public responsibility for appropriate portions of the content. MM, TN, TK, AK, and MH have participated in the conception and design, acquisition, analysis, and interpretation of data; revising it critically and ensuring the accuracy and integrity of manuscript. HM, MK, and KH have participated in drafting, acquisition, analysis, and interpretation of data; revising it critically and ensuring the accuracy and integrity of manuscript. All authors have participated in the final version approval of manuscript.

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