

Case Report

Computed tomography findings of pancreatic peripheral primitive neuroectodermal tumor in an elderly man: a case report

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Abstract: Peripheral primitive neuroectodermal tumors (pPNETs) with pancreatic origins are extremely rare. This study reported a case of pancreatic pPNET in a 64-year-old man. Plain abdominal computed tomography (CT) identified a large, round-like, poorly defined solid mass with a large hypointense area in the pancreatic tail. Contrast-enhanced CT revealed that the tumor parenchyma had medium heterogeneous contrast enhancement with a large non-enhanced area of cystic degeneration and necrosis during pancreatic substance phase and progressive contrast enhancement during portal phase. The tumor infiltrated the splenic artery and vein. Patient's advanced age complicates the preoperative diagnosis of this malignancy. Therefore, we proposed that pPNET should be considered in the differential diagnosis of masses arising in the pancreas, particularly in those with wide local invasion.

Keywords: Peripheral primitive neuroectodermal tumors, pancreas, computed tomography

Introduction

Peripheral primitive neuroectodermal tumors (pPNETs) are small round cell tumors that arise from primitive neuroepithelial stem cells. pPNETs with a pancreatic origin are extremely rare, and their clinical and imaging features have not been adequately studied. To our knowledge, only 26 cases of pancreatic pPNETs have been reported in the literature [1-20]. These cases exclusively involved children, adolescents, and middle-aged patients, that is, patients aged 2-37 years old. Herein, we report a case of pancreatic pPNET in a 64-year-old man to provide additional information and improve the diagnostic specificity of this disease.

Case report

A 64-year-old man presented to the Affiliated Yantai Yuhuangding Hospital of Qingdao University in June 2016 with a 2-month history of persistent pain in the left abdomen. The pain was moderate in intensity and can radiate to

the back and become aggravated after eating. It was accompanied by nausea, abdominal distention, anorexia, and emaciation. The clinical examination of the left abdomen was indicative of tenderness and rebound tenderness.

Plain abdominal computed tomography (CT) revealed a large, round-like, 7.7 cm × 6.1 cm × 7.1 cm, poorly defined solid mass with a large hypointense area in the pancreatic tail (**Figure 1A**). Upon contrast-enhanced CT imaging, the tumor parenchyma showed medium heterogeneous contrast enhancement with a large non-enhanced area of cystic degeneration and necrosis during pancreatic substance phase (**Figure 1B**) and progressive contrast enhancement during portal phase (**Figure 1C**). The tumor infiltrated the splenic artery and vein. Clinical examination and imaging showed no evidence of any other metastatic or primary lesions in the body. CT findings were suggestive of a malignant tumor of the pancreatic tail. The tumor was initially considered as neuroendocrine carcinoma, which primarily occurs in the adult population.

CT findings of pancreatic pPNET

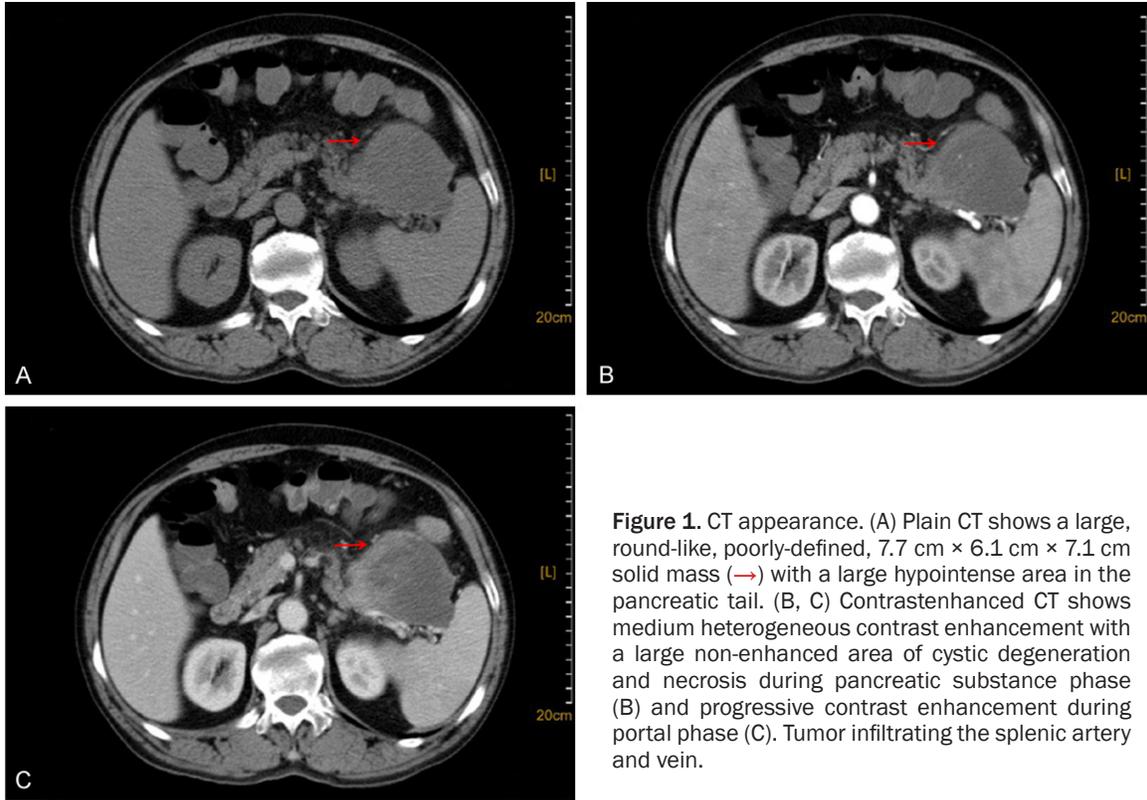


Figure 1. CT appearance. (A) Plain CT shows a large, round-like, poorly-defined, 7.7 cm × 6.1 cm × 7.1 cm solid mass (→) with a large hypointense area in the pancreatic tail. (B, C) Contrast-enhanced CT shows medium heterogeneous contrast enhancement with a large non-enhanced area of cystic degeneration and necrosis during pancreatic substance phase (B) and progressive contrast enhancement during portal phase (C). Tumor infiltrating the splenic artery and vein.

The patient showed low hemoglobin levels of 108 g/L and elevated neuron-specific enolase (NSE) and tumor-specific growth factor levels of 25.9 and 75.8 U/ml, respectively.

Laparotomy revealed that the tumor originated from the pancreatic tail and had invaded into the splenic artery and vein. An enbloc-resection of the tumor was accomplished through left pancreatic resection and splenectomy.

Gross examination showed that the tumor measured 8 cm × 7 cm × 7.5 cm in size, was surrounded by an edematous capsule, and appeared cystic with alternating necrotic areas.

Microscopic examination revealed that the tumor cells were distributed in the form of sheets with abundant interstitial sinuses and the presence of hemorrhaging and blood paste formation (**Figure 2A**). Under high magnification, the tumor cells appeared epithelial or fusiform and showed unclear boundaries and abundant eosinophilic granules in the cytoplasm. Nuclear atypia was pronounced, and the nuclear fission phase was visible (**Figure 2B**). Immunohistochemical staining revealed that the tumor cells were strongly positive for

CD99 (**Figure 3**), VIM, INI-1, and CD34. The positive rate of KI67 was approximately 50%.

Cytogenetic analysis through fluorescence in situ hybridization confirmed t(11;22)(q24;q12) translocation.

The results of histology, immunohistochemical staining, and cytogenetic analysis were indicative of a PNET.

No chemotherapy or radiotherapy was performed because of the refusal of the patient and his family. The patient died of disease recurrence approximately 15 months later.

The present study was approved by the ethics committee of the Affiliated Yantai Yuhuangding Hospital of Qingdao University (Yantai, China). Written informed consent was obtained from the patient.

Discussion

PNETs are small round cell tumors that originate from primitive neuroepithelial stem cells. These tumors are members of Ewing's sarcoma family, with common morphological, histological, immunohistochemical, and ultrastructural

CT findings of pancreatic pNET

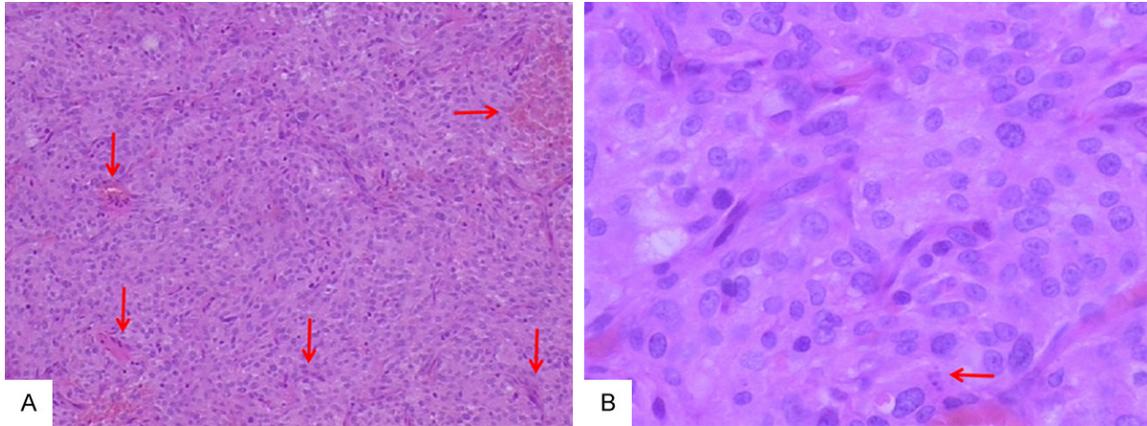


Figure 2. Pathological features. A. Microscopic examination reveals that the tumor cells are distributed in the form of sheets with abundant interstitial sinuses (↓) and the presence of hemorrhaging and blood paste formation (→) (hematoxylin and eosin stain, × 100). B. Under high magnification, the tumor cells appear epithelial or fusiform and show unclear boundaries and abundant eosinophilic granules in the cytoplasm. Nuclear atypia is pronounced, and the nuclear fission phase is visible (←) (hematoxylin and eosin stain, × 400).

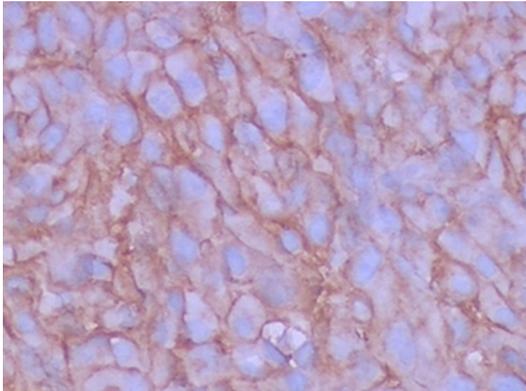


Figure 3. Immunohistochemical feature. Immunohistochemical staining reveals that the tumor cells are strongly positive for CD99.

indications of neuroectodermal differentiation, and identical chromosomal translocations at t(11;22)(q24;q12) [21].

PNETs can be further classified as central PNET and peripheral PNET (pPNET). pPNETs that occur outside the central and sympathetic nervous systems account for approximately 1% of all sarcomas [22]. pPNETs predominantly occur in the soft tissues of the extremities and are commonly found in patients less than 35 years of age with a slight predominance among males; the 5-year survival rate for this malignancy is approximately 50% [23].

Although pPNETs seldom arise in organs, sporadic cases with origins in the kidney, urinary bladder, lung, uterus, gall bladder, and vagina

have been reported. pPNETs with pancreatic origins are extremely rare. Lüttges reported that only 2 out of 600 cases of primary pancreatic neoplasma (0.3%) were diagnosed as pancreatic PNETs [2]. The ages of the reported patients ranged from 2 years to 37 years with a mean age of 19 years [1-20]. The present case was a 65-year-old man who was considerably older than the previous patients with PNETs.

pPNET is a locally invasive tumor that grows aggressively and exhibits an infiltrative and expansive pattern [24]. The clinical presentation of pancreatic pPNETs is nonspecific, and its onset appears insidious with abdominal pain as the most common symptom, accompanied by jaundice, nausea, anemia, and endocrine disorders [1-20].

This tumor commonly occurs in the pancreatic head (68%). Compared with cases of invasive growths, such as ductal carcinoma, only 29% of cases of pancreatic PNET exhibited supervened obstructive jaundice because of the expansive growth of the tumor [19].

Histologically, pPNET comprises small monomorphic round cells with small nuclei and scant cytoplasm. These cells are typically arranged tightly in cords, nests, or clusters to form rosettes and pseudorosettes (Homer-Wright daisy groups). PNETs express the products of the MIC2 gene, which include CD99, O13, and 12E74, on the X chromosome and may be positive for NSE, vimentin, or cytokeratin expression. PNETs also exhibit a typical chromosome

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translocation involving the EWS gene locus t(11;22)(q24;q12). pPNET can be accurately diagnosed based on these histological features and immune cell changes.

Preoperative imaging examinations, including abdominal CT/MRI scan, are useful in identifying and diagnosing the origin and extension of a pPNET. However, these imaging modalities fail to provide accurate diagnoses due to the lack of specificity and inadequate sensitivity. Tan et al. [25] reported that the further diagnosis of pPNET should be suggested in young men when the following imaging criteria are met: presence of a single large ill-defined solid mass with a small area of necrosis, negligible extent of calcification or hemorrhage, and local invasion of adjacent structures. The diagnosis for pPNET should be considered, especially for patients with tumors that are substantially enhanced and isointense on T1WI and T2WI. The tumor observed in our patient presented as a large, round-like, and poorly defined solid mass in the pancreatic tail with a large necrosis area and medium heterogeneous contrast enhancement. The tumor infiltrated the splenic artery and vein. The imaging performance of the tumor was consistent with Tan's diagnostic criteria. Nevertheless, the tumor was misdiagnosed as a neuroendocrine tumor because the patient's age was out of the typical age range of patients with PNET.

Pancreatic pPNETs among the elderly should be distinguished from other neoplasms, such as neuroendocrine tumors, lymphomas, and metastatic tumors. Similarities between the imaging appearances of pPNETs and those of other malignancies complicate the preoperative diagnosis based on radiological criteria. Diagnosis is confirmed only after the histological analysis of the tumor.

The current standard treatment for pPNETs is complete surgical resection with an adequate margin. Surgical resection increases disease control and survival rates [26], but performing this approach in pancreatic pPNETs may occasionally be difficult because of the unresectable major vessels adjacent to the pancreas. In the present case, the tumor had infiltrated the splenic vasculature, and splenectomy was performed. Chemotherapy and radiotherapy have an important role in controlling pPNET, though their therapeutic effects are sometimes unsatisfactory.

In conclusion, we presented a case of pancreatic pPNET in an elderly man. Patient's advanced age complicates the preoperative diagnosis of this malignancy. Despite its rarity, pPNET should be considered in differential diagnosis of masses in the pancreas, particularly those with wide local invasion.

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Disclosure of conflict of interest

None.

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