Case Report

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Synchronous pancreatic neuroendocrine tumor and pancreatic cyst: a case report

Hao Hua[1], Xun Ran[1], Yi-gang Chen[1],Chao-quan Hu[1], Fu-tang Li1, Shi Zuo1, Cheng-yi Sun1, Chao Yu1

1.Department of Hepatic-Biliary-Pancreatic Surgery, The Affiliate Hospital of Guizhou Medical University, Guiyang, Guizhou, China

Corresponding author

Chao Yu, No. 28, Guiyi road, Yunyan distract, Guiyang city, Guizhou Province, 550004, People's Republic of China.
E-mail: yuchao2002@gmc.edu.cn.
Tel: 86-851-6771326

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Abstract

Synchronous pancreatic neuroendocrine tumor (pNET) and pancreatic cyst are very rare. Herein, we report a case of pNET combined with a pancreatic cyst in a 49-year-old female. The patient first presented with a complaint of persistent pain in the right upper abdomen. As a follow-up, CT scan was performed that showed a solid pNET of 2-cm in diameter and a pancreatic cyst of 5-cm in the tail. Then, the subsequent distal pancreatomy with splenectomy was performed. The tumor histologically diagnosed with a cystic pancreatic neuroendocrine tumor. Therefore, it is critical to get a deep insight into this seldom tumor. Surgery remains the standard option of treatment for pNET.

Keywords

pancreatic cyst, pancreatic neuroendocrine tumor, case report

1. Introduction

Pancreatic neuroendocrine tumors(pNETs), a subgroup of neuroendocrine neoplasms, are different from pancreatic adenocarcinoma due to the distinctive biological behavior[1], accounting for 1%~2% of primary pancreatic tumors. From the perspective of tumor secretion function and hormone-related symptoms, these tumors can be classified as "functional tumors" and "non-functional tumors". Whereases, based on the genetic characteristics of tumors, they are divided into "sporadic tumors" and "genetically related tumors". pNETs are mostly solid, and solid cystic changes may also occur[2]. Pancreatic cysts are usual in pancreatic diseases and the morbidity increases with age[3], mostly detected accidently depended on noninvasive imaging[4]. Pancreatic cysts are divided into several types. Some types may have malignant potential, while others are benign, and even cysts with malignant potential rarely develop into cancer[5]. Although various solitary pancreatic tumors are common, pNETs have been less depicted in association with a pancreatic cyst. However, most cystic pNETs combined with pancreatic cyst showed no clinical symptoms of abnormal hormone secretion, and their imaging manifestations were easily mixed up with other pancreatic cystic tumors, making accurate preoperative diagnosis challenging. Given the high heterogeneity and infrequency of this type of tumor, the clinical pathway, the treatment methods, and the combination of protocols are complex and diverse. Meanwhile, with the rapid progress of clinical and basic research, the standardized diagnosis and treatment of this disease are still being improved. Previously, there were a few reported about this disease. Herein, we report a case of pNET combined with a pancreatic cyst to improve the clinician's knowledge of this rare disease.

2.Case report

A 49-year-old female presented to the department of Hepatic-Biliary-Pancreatic Surgery with pain in the right upper abdomen over the last three months, affecting the waist and back. She denied fever or jaundice. Her past medical history included hypertension for three years, and she regularly takes loradipine. On an examination, she showed slight pain in the epigastrium. Laboratory findings, including aspartate aminotransferase, alanine aminotransferase, amylase, carcinoembryonic antigen, carbohydrate antigen 19-9, and α-fetoprotein protein, were within normal ranges. An abdominal ultrasound found a 2-cm solid tumor in the pancreatic body and a 5-cm cystic lesion in the pancreatic tail end. She underwent an enhanced computed tomography (CT), which revealed the possibility of benign pNET associatied with pancreatic cyst (figure 1). On follow-up MRI abdomen, there are nodules of blood supply in the pancreatic body and a cystic lesion in the tail, with obstructive pancreatic atrophy and dilatation of the main pancreatic duct, similar to the CT scan result. Then, distal pancreatomy with splenectomy was performed assisted by Leonardo's robot (figure 2). The final diagnosis of pNET G1 and synchronous pancreatic cystic change in the pancreatic tail was confirmed by histopathology (figure 3). No lymph node metastases were detected. The patient received no adjuvant therapy. A progression-free survival, as shown by CT, was obtained for two months.

3.Discussion

The concurrent occurrence of various kinds of tumors in pancreatic is clinically uncommon[6]. The case presented here underlined that a pancreatic cyst, a common benign lesion, can be associated with inapparent simultaneous pNETs. This kind

of disease tends to occur in middle-aged people and is slightly more common in women[7]. Moreover, the patient in our case is a middle-aged woman, which is similar to the literature report. The pancreatic cyst is more likely to occur in the tail of the pancreas[8]. Nineteen cases of cystic and solid pNETs were reported, mostly located in the body of the pancreatic body[9]. In our case, the pNET was located in the body of the pancreatic, and the pancreatic cyst occurred in the tail of the pancreatic, which is in well agreement with the result in the previous literature.

The understanding of the pathological mechanism of cystic degeneration is also critical for diagnosing pNETs combined with a pancreatic cyst from cyst pNETs. In cyst pNETs, liquefaction and infarction were the main reasons for the formation of cystic lesions, and the degeneration of the tumor is accompanied by the formation of the surrounding fibrous capsule, which blocks the blood supply of the tumor, leading to internal ischemia and necrosis of the tumor[10]. However, in the current case, the pancreatic cyst was caused by cystic dilation of the distal pancreatic duct or acinar due to obstruction of the pancreatic duct by the solid pNET without necrosis confirmed by pathological diagnosis. The inner wall of the retention cyst is a monolayer of cubic or flat epithelial cells with clear cystic fluid.

Traditional imaging methods, including ultrasound, enhanced computed tomography (CT), and magnetic resonance imaging (MRI) are commonly used to diagnose pNETs and pancreatic cyst[11]. However, these methods have limited efficacy in the localization and staging of pNETs and the classification of a pancreatic cyst. Most pNETs express somatostatin receptors, so radionuclide labeled somatostatin analogs can be applied for tumor imaging[12]. Given the large proportion of cystic tumors, it is easy to be confused with other pancreatic cystic tumors, such as cystic pNETs, resulting in misdiagnosis. We should take some entities that can simulate it into consideration. Endoscopic ultrasound (EUS) and EUS-fine-needle aspiration (FNA) are beneficial for preoperative diagnosis of suspicious pNETs[13, 14]. Previous studies also elucidated that EUS-FNA with cyst fluid analysis can accurately diagnose individual sorts of pancreatic cysts and guide us to make a therapeutic decision in patients with pNETs and pancreatic cyst[15, 16]. Therefore, EUS—FNA should be considered for further diagnosis in patients whose clinical manifestations and CT or MRI are challenging to diagnose and cannot exclude this simultaneous disease. In the present case, the tumor was detected by imaging techniques as masses and the diagnosis is mainly based on CT and MRI. Accurate pathological diagnosis is the cornerstone of the clinical decision of pNETs and pancreatic cyst. The report of our case revealed that the tumor is non-functional and solid. The immunohistochemical result of current patient showed that, there were no microscopic satellite lesions and surrounded invasion and lymphatic metastasis. Features and the tumors presented as solitary masses with no extrahepatic involvement.

Compared with solitary pancreatic tumors, it is critical to perform surgical resection on multiple tumors, even when they are benign. Surgery is the main treatment for this simultaneous disease and radical surgical resection is the only choice for long-term survival. The main surgical strategy is distal pancreatectomy according to the location and nature of tumors[17]. Paiella et al. showed that cystic lesions were mostly located in the tail of the pancreas and solid lesions were likely to locate in the head and body of the oancreas[18]. Given the location of the tumor in the tail and body of the pancreas, we conducted resection of the body and tail of pancreas and splenectomy assisted by Leonardo's robot. There were no postoperative complications occurred, and the recovery was well. Due to the degree of proliferation of tumor cells, pNETs can be classified into three levels, including G1, G2, and G3, and the lower the grade, the less invasive it is[19]. Tumor size is considered to be a key factor in assessing the biological behavior and risk of recurrence of pNETs, and tumors with length longer than 2.0cm are more aggressive[20]. In our case, the degree of proliferation of pNET is G1 and its size is just 2.0cm. Considering the relatively mild biological behavior of pNETs and pancreatic cysts, there are no guidelines to recommend postoperative adjuvant therapy for these patients undergoing radical surgery. Thus, the prognosis is optimistic compared with other malignant pancreatic tumors.

To sum up, pNETs combined with pancreatic cysts are scarce. Being aware of pNETs and pancreatic cysts can improve the management and diagnosis of this entity and surgery is the main effective strategy for treatment. In the future, more clinical and basic researches will be required to explore the characteristic of this type of disease, which can optimize their therapeutic procedures and promote the establishment of standardized diagnosis and treatment strategies.

Data availability

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

AUTHOR CONTRIBUTIONS

HH prepared and wrote this article. CYG, LFT and HCQ were involved in managing the patient besides preparing the intraoperative pictures. YC wrote and revised the manuscript as well as acted as the corresponding author. RX wrote and reviewed this manuscript. YC was the main surgeon and SCY and ZS was involved directly in managing the patient. All authors contributed to the article and approved the submitted version

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Statement of Ethic

The patient consented to the anonymous publication of her data, which is gratefully acknowledged by the authors. Ethical approval was not required because this manuscript only reports a case

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Disclosure statement

The authors declare no conflicts of interest

Figures

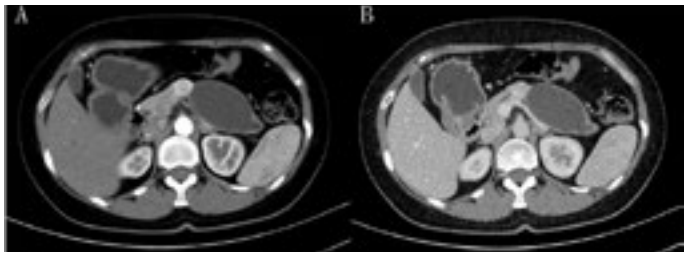

Figure 1: Examples of contrast-enhanced computed tomography (CT) images of the patient with pNET and pancreatic cyst (A and B). CT imaging demonstrates a well-defined mass of 2cm in size centered in the tail of the pancreas with enhancement and a 5cm cystic change lesion in the tail of the pancreas with low density.

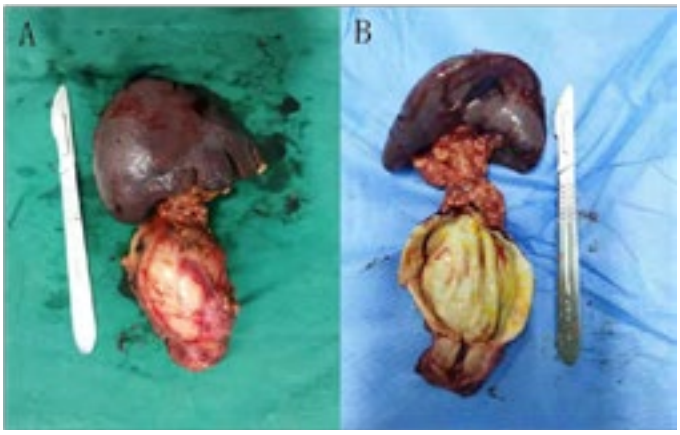

Figure 2: Macroscopic findings of the resected specimen (A). The section of the mass in the body of the pancreas showed a pale and solid nodule with clear boundary. The section of the pancreatic tail is characterized by single cystic change and smooth capsule wall (B)

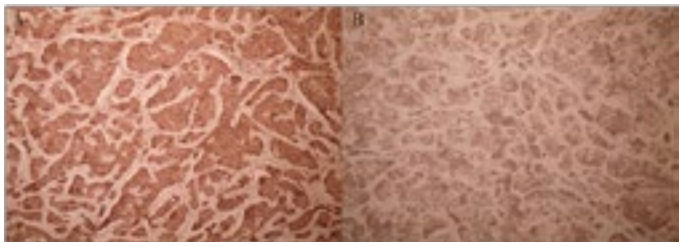

Figure 3: Pathological images showed cellular smears of classic neuroendocrine cells and pancreatic retention cyst (A and B).

References

1. Dasari A, Shen C, Halperin D, Zhao B, Zhou S, Xu Y, Shih T, Yao JC: Trends in the Incidence, Prevalence, and Survival Outcomes in Patients With Neuroendocrine Tumors in the United States. JAMA Oncol 2017, 3(10):1335-1342.

2. Toshima F, Inoue D, Komori T, Yoshida K, Yoneda N, Minami T, Matsui O, Ikeda H, Gabata T: Is the combination of MR and CT findings useful in determining the tumor grade of pancreatic neuroendocrine tumors? Jpn J Radiol 2017, 35(5):242-253.

3. Laffan TA, Horton KM, Klein AP, Berlanstein B, Siegelman SS, Kawamoto S, Johnson PT, Fishman EK, Hruban RH: Prevalence of unsuspected pancreatic cysts on MDCT. AJR Am J Roentgenol 2008, 191(3):802-807.

4. Zaheer A, Pokharel SS, Wolfgang C, Fishman EK, Horton KM: Incidentally detected cystic lesions of the pancreas on CT: review of literature and management suggestions. Abdom Imaging 2013, 38(2):331-341.

5. Simeone DM: SSAT/AGA/ASGE state of the art conference on cystic neoplasms of the pancreas. J Gastrointest Surg 2008, 12(8):1475-1477.

6. Posniak HV, Olson MC, Demos TC: Coexistent adenocarcinoma and microcystic adenoma of the pancreas. Clin Imaging 1991, 15(3):220-222.

7. Ridtitid W, Halawi H, DeWitt JM, Sherman S, LeBlanc J, McHenry L, Coté GA, Al-Haddad MA: Cystic pancreatic neuroendocrine tumors: outcomes of preoperative endosonography-guided fine needle aspiration, and recurrence during long-term follow-up. Endoscopy 2015, 47(7):617-625.

8. Reid MD, Choi H-J, Memis B, Krasinskas AM, Jang K-T, Akkas G, Maithel SK, Sarmiento JM, Kooby DA, Basturk O et al: Serous Neoplasms of the Pancreas: A Clinicopathologic Analysis of 193 Cases and Literature Review With New Insights on Macrocystic and Solid Variants and Critical Reappraisal of So-called "Serous Cystadenocarcinoma". The American journal of surgical pathol-

ogy 2015, 39(12):1597-1610.

9. Yoon WJ, Daglilar ES, Pitman MB, Brugge WR: Cystic pancreatic neuroendocrine tumors: endoscopic ultrasound and fine-needle aspiration characteristics. Endoscopy 2013, 45(3):189-194.

10. Kajiwara M, Gotohda N, Konishi M, Nakagohri T, Takahashi S, Kojima M, Hasebe T, Kinoshita T: Cystic endocrine tumor of the pancreas with an atypical multilocular appearance. J Hepatobiliary Pancreat Surg 2007, 14(6):586-589.

11. de Jong K, Nio CY, Mearadji B, Phoa SS, Engelbrecht MR, Dijkgraaf MG, Bruno MJ, Fockens P: Disappointing interobserver agreement among radiologists for a classifying diagnosis of pancreatic cysts using magnetic resonance imaging. Pancreas 2012, 41(2):278-282.

12. Ito T, Igarashi H, Jensen RT: Pancreatic neuroendocrine tumors: clinical features, diagnosis and medical treatment: advances. Best Pract Res Clin Gastroenterol 2012, 26(6):737-753.

13. Figueiredo FAF, Giovannini M, Monges G, Bories E, Pesenti C, Caillol F, Delpero JR: EUS-FNA predicts 5-year survival in pancreatic endocrine tumors. GastrointestEndosc 2009, 70(5):907-914.

14. Jani N, Khalid A, Kaushik N, Brody D, Bauer K, Schoedel K, Ohori NP, Moser AJ, Lee K, McGrath K: EUS-guided FNA diagnosis of pancreatic endocrine tumors: new trends identified. GastrointestEndosc 2008, 67(1):44-50.

15. Singhi AD, Zeh HJ, Brand RE, Nikiforova MN, Chennat JS, Fasanella KE, Khalid A, Papachristou GI, Slivka A, Hogg M et al: American Gastroenterological Association guidelines are inaccurate in detecting pancreatic cysts with advanced neoplasia: a clinicopathologic study of 225 patients with supporting molecular data. GastrointestEndosc 2016, 83(6).

16. Springer S, Wang Y, Dal Molin M, Masica DL, Jiao Y, Kinde I, Blackford A, Raman SP, Wolfgang CL, Tomita T et al: A combination of molecular markers and clinical features improve the classification of pancreatic cysts. Gastroenterology 2015, 149(6):1501-1510.

17. Ramanathan R, Al Abbas AI, Mason T, Wolfe LG, Kaplan BJ: Age-related risks of complications after distal pancreatectomy for neuroendocrine and cystic neoplasms. HPB (Oxford) 2019, 21(7):810-817.

18. Canellas R, Burk KS, Parakh A, Sahani DV: Prediction of Pancreatic Neuroendocrine Tumor Grade Based on CT Features and Texture Analysis. AJR Am J Roentgenol 2018, 210(2):341-346.

19. Mpilla GB, Philip PA, El-Rayes B, Azmi AS: Pancreatic neuroendocrine tumors: Therapeutic challenges and research limitations. World J Gastroenterol 2020, 26(28):4036-4054.

20. Abdullah JM, Mutum SS, Nasuha NA, Biswal BM, Ariff AR: Intramedullary spindle cell hemangioendothelioma of the thoracic spinal cord--case report. Neurol Med Chir (Tokyo) 2002, 42(6):259-263.