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Case Report
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Cystic Pancreatic Neuroendocrine Tumor: A Challenging Diagnosis

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Abstract

Background: Cystic Pancreatic Neuroendocrine Tumors (PNET) is rare and their diagnosis might be challenging. The aim of this report is to present a case of cystic PNET and discuss the radiological characteristics of these tumors and the possible differential diagnoses.

Case Report: A 57-year-old woman with nonspecific abdominal pain underwent Magnetic Resonance Imaging (MRI) showing a nodular lesion of the pancreatic tail, with a maximum diameter of 18 mm and main pancreatic duct dilatation. She repeated a magnetic resonance after 6 months showing increased cyst size (18 mm vs. 23 mm). Main Pancreatic Duct IPMN (MPD-IPMN) was considered the most probable diagnosis so she underwent distal pancreatectomy with spleen and splenic vessels preservation. Final histological exam showed a cystic neuroendocrine tumor. Cystic neuroendocrine tumor represents a challenging preoperative diagnosis. Most patients are asymptomatic or have nonspecific symptoms (abdominal pain, diarrhea, weight loss and asthenia). Computed tomography and magnetic resonance are not always diagnostic as these tumors may simulate other more frequent pancreatic cystic lesions.

Conclusion: The present case underlines the clinical, radiological and histological characteristics of cystic PNETs, discussing the possible differential diagnoses, including pancreatic cyst, pancreatic pseudocyst, MPD-IPMN, solitary pseudopapillary tumor and mucinous cystic tumor.

Keywords: Pancreas; Neuroendocrine pancreatic tumor; Pancreatectomy; Spleen preservation

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Abbreviations

PNET: Cystic Pancreatic Neuroendocrine Tumors; MRI: Magnetic Resonance Imaging; MPD-IPMN: Main Pancreatic Duct-Intraductal Papillary Mucinous Neoplasm; DWI: Diffusion Weighted Imaging; EUS: Echo endoscopy; CT: Computed Tomography; FNA: Fine Needle Aspiration

Case Presentation

A 57-year-old woman with an unremarkable past medical history was referred for abdominal pain localized in the upper quadrants with posterior irradiation. A Magnetic Resonance Imaging (MRI) performed 6 months before had shown a cystic pancreatic lesion of 18 mm of maximum diameter with Main Pancreatic Duct (MPD) dilatation. At our institution, she underwent MRI showing a nodular lesion of the pancreatic tail, with maximum diameter of 23 mm, iso-hypointense in T1 sequences, hyperintense with dishomogeneity in T2, with prevalent fluid content, a thin wall and incomplete septa. No signal restriction in Diffusion Weighted Imaging (DWI) sequences was observed. Enhancement of the cystic wall was detected. No communication between the lesion and the MPD was detected but the MPD was dilated at the level of the pancreatic tail. Small cystic pancreatic lesions, probably IPMN branch type, were observed in the head and tail (Figure 1). The volume of the lesion has increased comparing with the previous exam. The differential diagnosis included pancreatic cyst (but they usually have non-enhancing walls), pancreatic pseudocyst (but no history of pancreatitis was reported), MPD-IPMN, mucinous cystic tumor, neuroendocrine cystic tumor. MPD-IPMN was considered the most probable diagnosis. Even if the communication with the MPD was not visible, it was suspected considering such a MPD dilatation. Echo Endoscopy (EUS) demonstrated a pancreatic tail cystic lesion of approximately 25 mm. Due to the suspicion of MPD-IPMN, upfront surgery was scheduled after multidisciplinary evaluation and discussion

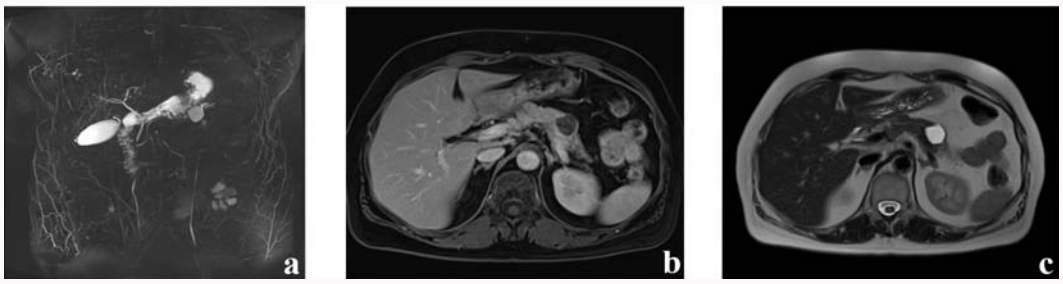

Figure 1: Magnetic resonance imaging with multiplanar in/out T1, T2 and T2fs, DWI, 3D cholangiopancreatography, T1fs sequences pre- and post-intravenous contrast injection with dynamic study and pancreatic protocol, showing a nodular lesion of the pancreatic tail, with maximum diameter of 23 mm.

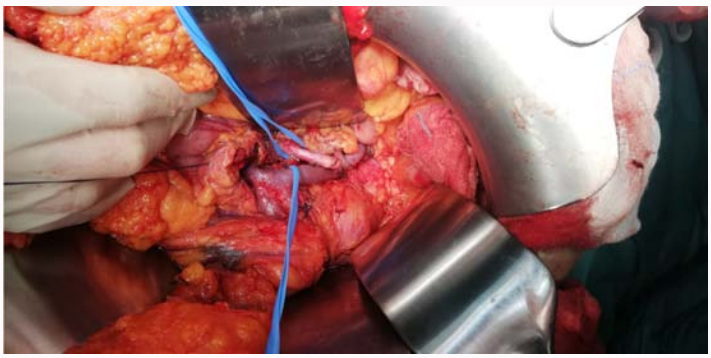

Figure 2: Intraoperative field after distal pancreatectomy with spleen and splenic vessels preservation.

Figure 3: Macroscopic (a) and histologic (b) aspect of a cystic neuroendocrine tumor of the pancreas.

with the patient. Distal pancreatectomy with preservation of the splenic vessels and the spleen was performed (Figure 2). Frozen section analysis of the pancreatic resection margin was negative. Final histological exam showed a cystic neuroendocrine tumor (chromogranin A+, synaptophysin +, insulin -), absence of mitosis, Ki67 of 2%, R0 (Figure 3).

Discussion and Conclusion

Cystic neuroendocrine tumor is a rare disease and represents a challenging preoperative diagnosis. Cystic Pancreatic Neuroendocrine Tumors (PNET) is infrequent tumors representing approximately 10% of all PNETs [1]. Most of them are non-secreting tumors [2]. The preoperative differential diagnosis includes mucinous cystic neoplasms, serous cystic neoplasms, solid pseudopapillary neoplasms, non-neoplastic cysts, IPMNs. Clinical suspicion of PNET may arise when a clinical syndrome due to excess hormone secretion is present. Non-specific symptoms may include abdominal pain, diarrhea, weight loss, asthenia [3]. Computed Tomography (CT) and MRI allows for the identification of the pancreatic mass, its relationship with the contiguous structures and the presence of distant metastases [4]. MRI cholangiopancreatography permits optimal visualization of the MPD. In our case, MRI showed a 23 mm lesion with ectasia of the MPD and suspect communication between the lesion and the MPD. The dilatation of the MPD was considered a high-risk stigma, and the increase in size during time was also taken into account [5]. EUS confirmed the findings of MRI. Unfortunately, Fine Needle Aspiration (FNA) was not performed. A diagnosis of MPD-IPMN was considered the most probable and the patient was informed about the potential risk of malignancy. The options of surveillance or surgery were extensively discussed with the patient and finally the decision of distal pancreatectomy with spleen preservation was undertaken. The postoperative period was complicated by grade B pancreatic fistula.

The present case underlines the importance of considering cystic neuroendocrine tumors in the differential diagnosis of pancreatic cystic lesions. In this case, MPD-IPMN was the most probable diagnosis according to preoperative exams. Other authors reported preoperative diagnosis of solid pseudopapillary neoplasm [6] or mucinous cystic neoplasm [2]. If a solid tumor component is accessible

to FNA, it may allow a preoperative cytopathological diagnosis [7]. FNA is considered a safe procedure even if rarely complications may occur [8]. The challenging diagnosis of cystic PNET affects patients' treatment. In our case, the arguments in favor of surgery were the fact that MPD-IMPN with MPD dilatation was suspected and its increase in size. The location of the tumor also had a role in patient's counseling and in the therapeutic decision, as distal pancreatectomy is less harmful than pancreaticoduodenectomy. Finally, patients' characteristics and will were also taken into account. We based our reflections on the available preoperative data and on the guidelines on the management of IPMNs [9]. A preoperative diagnosis of PNET could have differently oriented the discussion about the treatment. In case of small PNETs (<2 cm), a conservative management including observation or enucleation may be proposed [10].

In conclusion, preoperative diagnosis of cystic PNET is a real challenge, and it is very important to share preoperative images of such complex cases. If available, FNA may allow an accurate preoperative diagnosis based on the cytopathological exam.

Author Contributions

MP wrote the paper. CF provided the cases. ER and IM provided the MRI images and interpretation of the data. EP provided histopathological images and interpretation of the data. NP and MC reviewed and edited the manuscript. All authors read and approved the manuscript.

References

1. Noda K, Kuroki T, Yamashita M, Hirayama T, Natsuda K, Kobayashi S, et al. Nondegenerated cystic neuroendocrine tumor of the pancreas: A case report. Surg Case Rep. 2020;6(1):154.
2. Kumar T, Gupta B, Das P, Madhusudhan KS. The rare case of a cystic pancreatic neuroendocrine tumor. Autopsy Case Rep. 2020;10(3):e2020171.
3. Pezzilli R, Buscarini E, Pollini T, Bonamini D, Marchegiani G, Crippa S, et al. Epidemiology, clinical features and diagnostic work-up of cystic neoplasms of the pancreas: Interim analysis of the prospective PANCY survey. Dig Liver Dis. 2020;52(5):547-54.
4. Dromain C, Déandréis D, Scoazec J-Y, Goere D, Ducreux M, Baudin E, et al. Imaging of neuroendocrine tumors of the pancreas. Diagn Interv Imaging. 2016;97(12):1241-57.
5. Hecht EM, Khatri G, Morgan D, Kang S, Bhosale PR, Francis IR, et al. Intraductal Papillary Mucinous Neoplasm (IPMN) of the pancreas: Recommendations for standardized imaging and reporting from the Society of Abdominal Radiology IPMN disease focused panel. Abdom Radiol (NY). 2021;46(4):1586-606.
6. Limaiem F, Mestiri H, Mejri S, Lahmar A, Mzabi S. Solid pseudopapillary neoplasm of the pancreas in two male patients: Gender does not matter. Pan Afr Med J. 2017.
7. Ishii T, Katanuma A, Toyonaga H, Chikugo K, Nasuno H, Kin T, et al. Role of endoscopic ultrasound in the diagnosis of pancreatic neuroendocrine neoplasms. Diagnostics (Basel). 2021;11(2):316.
8. Virgilio E, Mercantini P, Ferri M, Cunsolo G, Tarantino G, Cavallini M, et al. Is EUS-FNA of solid-pseudopapillary neoplasms of the pancreas as a preoperative procedure really necessary and free of acceptable risks? Pancreatology. 2014;14(6):536-8.
9. Tanaka M, Fernández-del Castillo C, Kamisawa T, Jang JY, Levy P, Ohtsuka T, et al. Revisions of international consensus Fukuoka guidelines for the management of IPMN of the pancreas. Pancreatology. 2017;17(5):738-53.
10. Howe JR, Merchant NB, Conrad C, Keutgen XM, Hallet J, Drebin JA, et al. The North American Neuroendocrine Tumor Society Consensus Paper on the surgical management of pancreatic neuroendocrine tumors. Pancreas. 2020;49(1):1-33.