REPORTE DE CASO

Pancreatic neuroendocrine tumor masquerading as intra-ductal papillary mucinous neoplasm: a case report

Tumor neuroendocrino pancreático que simula una neoplasia papilar mucinosa intraductal: reporte de un caso

Pritesh Kumar N.[1], **Yash Kumar Jain**[1], **Hirdaya H. Nag**[1]

[1] Department of Surgical Gastroenterology, GB Pant Institute of Postgraduate Medical Education and Research, New Delhi, India.

Received: 19/02/2026
Peer-reviewed
Accepted: 30/07/2026
Online: 24/09/2026

Author contribution
PKN: conception and design of study, analysis and/or interpretation of data, drafting the manuscript. YKJ: acquisition of data, drafting the manuscript. HHN: conception and design of study, analysis and/or interpretation of data, revising the manuscript critically for important intellectual content, supervision and leadership.

Conflict of interest
None.

Funding
None.

Cite as
Kumar N P, Kumar Jain Y, HN H. Pancreatic neuroendocrine tumor masquerading as intra-ductal papillary mucinous neoplasm: a case report. Rev Gastroenterol Peru. 2026;46(3). doi: 10.47892/rgp.2026.463.2218.

ABSTRACT
Pancreatic cysts are being commonly diagnosed owing to the increased utilization of radiological modalities for the evaluation of abdominal complaints. Cysts can represent an underlying benign, pre-malignant or malignant diagnosis, and making an accurate diagnosis is often challenging. Here we report a case of a pancreatic cystic lesion, provisionally diagnosed as intra-ductal papillary mucinous neoplasm (IPMN) based on pre-operative and intra-operative findings, which on final histopathology turned out to be a pancreatic neuroendocrine tumor.
Keywords: *Pancreatic cyst; Pancreatic Neoplasm; Pancreatic Intraductal Neoplasms (source: MeSH NLM).*

RESUMEN
Los quistes pancreáticos se diagnostican cada vez con mayor frecuencia debido al uso creciente de las técnicas de imagen para la evaluación de pacientes con síntomas abdominales. Estas lesiones pueden corresponder a entidades benignas, premalignas o malignas, por lo que establecer un diagnóstico preciso continúa siendo un desafío. Presentamos el caso de una lesión quística pancreática que fue diagnosticada inicialmente como una neoplasia papilar mucinosa intraductal (NPMI), tanto por los hallazgos preoperatorios como intraoperatorios. Sin embargo, el estudio histopatológico definitivo demostró que se trataba de un tumor neuroendocrino pancreático.
Palabras clave: *Quiste Pancreático; Neoplasias Pancreáticas; Neoplasias Intraductales Pancreáticas (fuente: DeCS Bireme).*

INTRODUCTION

The term "cystic lesions of the pancreas" encompasses a wide variety of diagnoses that range from benign and indolent lesions such as a simple cyst to aggressive malignant tumors with cystic degeneration [1]. Amongst the neoplastic entities, pancreatic cystic neoplasms are most common, accounting for 10-15% of all pancreatic cysts, and are further sub-divided into specific types based on their morphological and pathological characteristics [2]. One such cystic neoplasm is the intraductal papillary mucinous neoplasm (IPMN) which has a varied clinical presentation ranging from incidental diagnosis on imaging, to infiltrative malignancy and metastatic disease [3].

On the other hand, the pancreas is the most common gastrointestinal site for neuroendocrine tumor (NET) which represents approximately 5% of all pancreatic neoplasms [4,5]. These tumors may be hormonally inert or may secrete an array of active substances, each associated with a clinical syndrome. Pancreatic NETs (PNET) usually appear as solid space occupying lesions (SOL) which are hypervascular, a feature that is generally picked up on contrast-enhanced imaging. However, a subset of the tumors may have atypical appearance posing a preoperative diagnostic dilemma in some patients [6]. Here we report one such case of PNET which was misdiagnosed as IPMN preoperatively and intraoperatively.

Correspondencia:
Hirdaya H. Nag
E-mail: hirdayanag@gmail.com

CASE REPORT

Patient was a 50-year-old female presenting with an 8-month history of mild upper abdominal pain, occasionally radiating to the back, without any history of weight loss, anorexia, steatorrhoea, vomiting, jaundice, abdominal distension, or symptoms suggestive of any hormonal excess. She had no significant family history, nor did she have any history of alcohol or other substance abuse. On examination, she was vitally stable, with essentially normal general physical examination. Abdomen was soft, non-tender, with no organomegaly, palpable lump or free fluid. The remainder of the systemic examination was also normal. Her baseline blood tests, including serum amylase and lipase were within normal limits. CEA (Carcinoembryonic antigen) and CA 19-9 (Carbohydrate antigen 19-9) were 1.89 ng/dl and 7.73 IU/ml, respectively.

Ultrasonography revealed a cystic SOL in the neck and body of pancreas measuring 4.5 x 4.0 cm, without any internal septations, and associated with dilated main pancreatic duct (MPD). Contrast-enhanced computed tomography (CECT) showed a well-defined non-enhancing hypodense lesion (23HU) in neck and proximal body of pancreas measuring 45 x 46 x 49 mm without soft-tissue component and was associated with dilatation of proximal MPD and side branches with resultant atrophy of pancreatic body and tail region, having suspicious communication with the MPD (Figure 1). Magnetic resonance cholangiopancreatography (MRCP) also revealed similar findings suggestive of a well-defined T2 hyperintense cyst in neck and proximal body which appeared to be communicating with the tortuously dilated MPD (Figure 2). Endoscopic ultrasound (EUS) showed which also showed that the anechoic cystic lesion was communicating with the dilated MPD, had no solid component, and was associated with parenchymal atrophy in body. Cyst fluid analysis reports were as follows: Amylase 14.993 IU/L, CEA 2.8 ng/mL, CA19.9: >1000 IU/mL. String sign test for mucin was positive. Cytological evaluation was non-diagnostic.

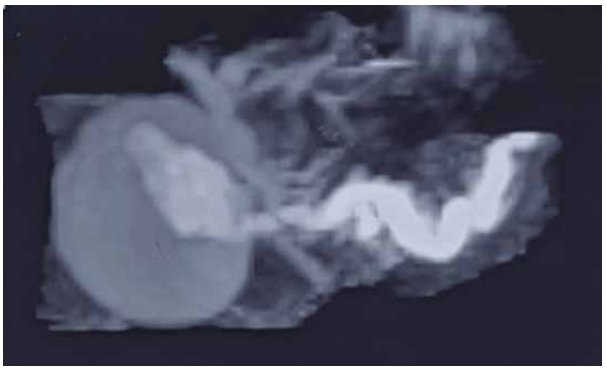

Figure 2. MRCP showing T2 hyperintense cyst in neck and proximal body of pancreas, which appears to be communicating with the tortuously dilated MPD.

A working diagnosis of IPMN (main duct type) was made, and the patient was planned for surgery. At laparotomy, a well-defined cystic lesion was identified in neck and proximal body of pancreas measuring ~5 x 5 cm, protruding anteriorly and superiorly (Figure 3). Head of pancreas was grossly normal, while body and tail were mildly atrophic. There were no post- inflammatory peri-pancreatic adhesions. Regional lymph nodes were not enlarged. Splenic artery and vein were free from the lesion. Spleen-preserving distal pancreatectomy was done by dividing the pancreas at the level of neck using Echelon™ 60 stapler with gold reload (Ethicon Endo-surgery, USA) (Figure 4). Frozen section analysis of resection margin did not show any dysplasia or malignancy. A drain was placed in lesser sac and abdomen was closed in layers. Postoperatively, she developed Grade A pancreatic fistula, which was managed conservatively. Drain was removed on 5th day, and she was discharged on the 6th day.

Gross pathology showed a unilocular cyst with wall thickness of 2-3 mm, communicating with MPD, with small papillary projections and mucinous fluid within the cyst

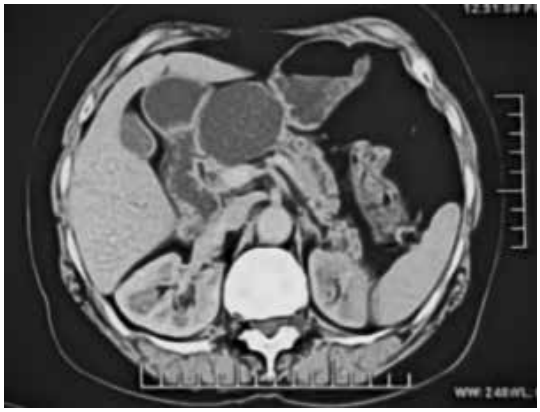

Figure 1. CECT showing well-defined non enhancing hypodense lesion in neck and proximal body of pancreas, with dilatation of MPD.

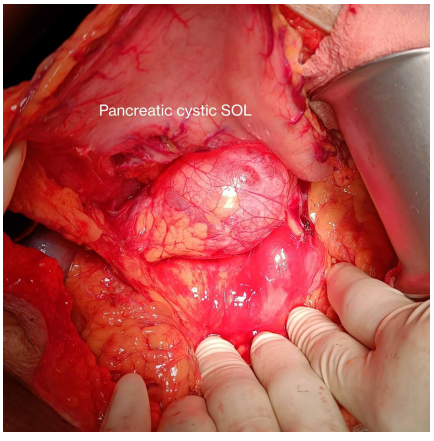

Figure 3. Intra-operative photograph showing cystic pancreatic lesion.

https://doi.org/10.47892/rgp.2026.463.2218

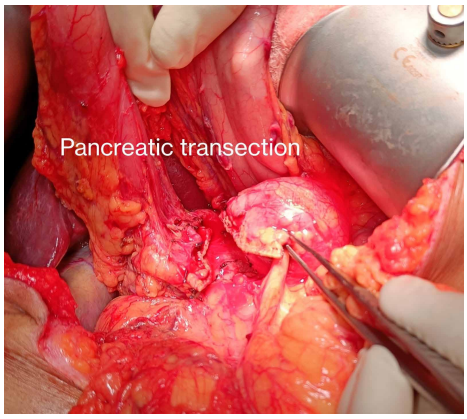

Figure 4. Pancreatic transection proximal to the cystic lesion.

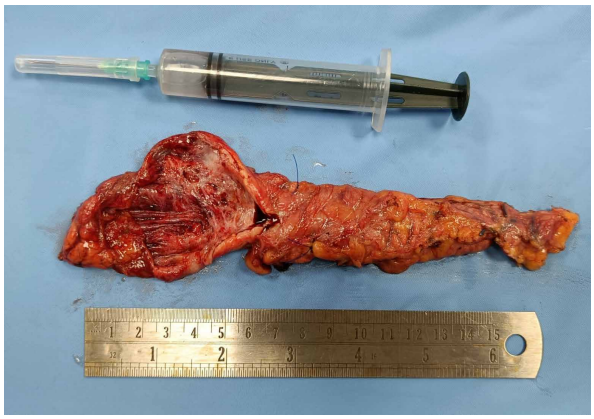

Figure 5. Resected specimen showing unilocular cyst opening into MPD; aspirate yielded mucinous fluid.

(Figure 5). Histopathological examination revealed a tumor in the proximal body measuring 2.1 x 0.5 x 0.5 cm, lying 0.5 cm away from the resection margin, with tumor cells arranged in trabeculae and nests, showing amphophilic cytoplasm, high nuclear-to-cytoplasmic ratio, round nucleus with stippled chromatin and inconspicuous nucleoli (Figure 6A). The cells were immunopositive for synaptophysin, chromogranin and INSM1, with MIB-1 index <1% and Ki-67 labelling index <1% (Figure 6 C-F). There was no lymphovascular or perineural invasion. Sections from cyst wall showed fibrous tissue and histiocytes, with no epithelial lining suggestive of a pseudocyst, while sections from rest of pancreatic parenchyma showed features of chronic pancreatitis. There were no features of IPMN in the entire specimen. The final diagnosis was a well-differentiated PNET (2017 World Health Organization Classification, TNM stage pT2pN0).

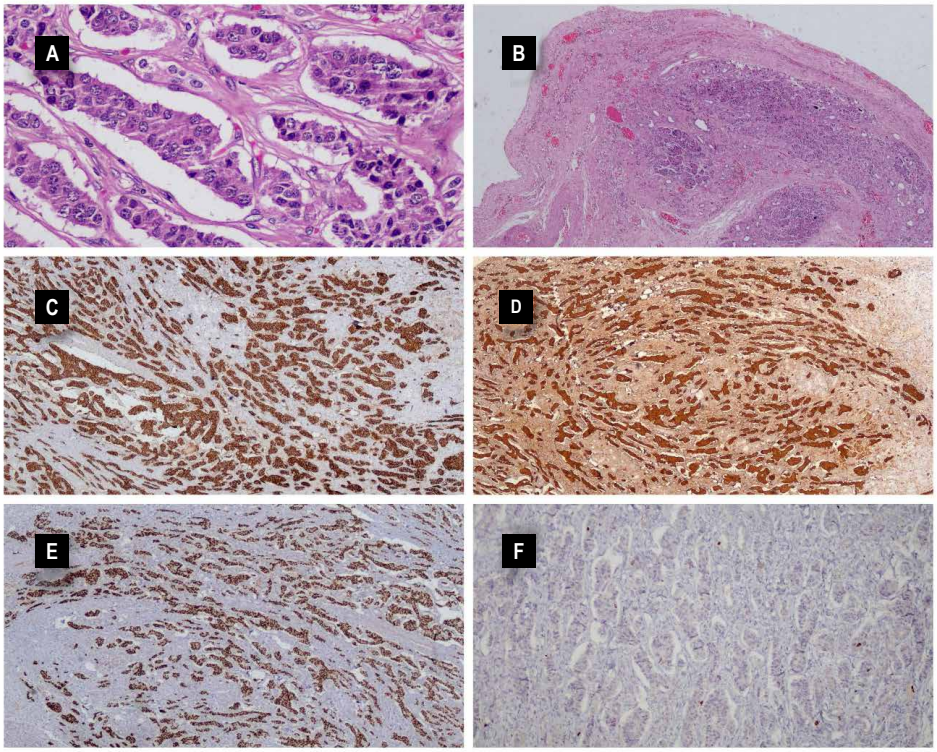

Figure 6. Photomicrographs of representative histopathological and immunohistochemical findings. **Panel A:** H&E staining (40X) showing tumor cells arranged in trabeculae and nests, with amphophilic cytoplasm, high nuclear-to-cytoplasmic ratio, round nucleus and inconspicuous nucleoli. **Panel B:** H&E staining showing cyst wall and pancreatic parenchyma. Panels C through F showing immunohistochemical staining: The cells are immune-positive for synaptophysin **(C)**, chromogranin **(D)** and INSM1 **(E)**, with Ki-67 labelling index <1% **(F)**.

Table 1. Differential diagnoses of pancreatic cystic lesions with MPD dilatation.

Lesion	Characteristic features
Chronic pancreatitis with pseudocyst	Unilocular cyst without solid components or wall calcification, Atrophic parenchyma, peripancreatic fat-infiltration, ductal and parenchymal calcification, cyst fluid high in amylase
Main duct IPMN	Cystic dilatation (uni- or multi-focal) of the MPD caused by papillary protrusions lined by mucin-secreting epithelium with variable degree of dysplasia. Cyst fluid contains mucin, high levels of amylase, and CEA
MCN with malignant degeneration	Macrocystic with thick wall septations, peripheral calcifications, typically does not involve MPD; however, with malignant degeneration, MPD involvement can cause ductal dilatation.
Tumors of ductal origin (PDAC)	Usually ill-defined and heterogenous; however, large masses with extensive necrosis may appear cystic. MPD dilatation is due to direct involvement by cancerous cells.
Tumors of parenchymal origin	These tumors may cause MPD dilatation by extrinsic compression either due to their large size (eg: SPEN), or proximity to the duct (eg: PNET, metastasis). Intra-tumoral necrosis is often the cause for cystic appearance on imaging; however, ductal obstruction may produce pseudocysts.
Benign lesions (eg: Hydatid cyst)	MPD dilatation by extrinsic compression, or fistulization; radiological appearance depends on the stage of the disease; cyst fluid analysis may reveal the scolices; to be suspected in endemic areas, especially in patients with positive serology.

CEA= Carcinoembryonic Antigen, **IPMN= I**ntra-ductal Papillary Mucinous Neoplasm, **MCN=** Mucinous Cystic Neoplasm, **MPD=** Main Pancreatic Duct, **PDAC=** Pancreatic Ductal Adenocarcinoma, **PNET=** Pancreatic Neuroendocrine Tumor, **SPEN=** Solid Pseudopapillary Epithelial Neoplasm.

The case was discussed in a multi-disciplinary tumor board meeting, following which we obtained serum chromogranin level which was normal, and 68-Gallium DOTATATE scan, which showed no residual tumor in remnant pancreas or anywhere elsewhere in the body. She received no adjuvant therapy. At 1-year follow up she was asymptomatic, having no signs of endocrine or exocrine pancreatic insufficiency, with no recurrent lesion on CECT scan.

DISCUSSION

With the increasing usage of radiological investigations, cystic lesions of the pancreas are being diagnosed more commonly [1]. A wide range of differential diagnoses needs to be considered and differentiating them from one another can be a diagnostic challenge pre-operatively. Patients may have vague abdominal symptoms or be totally asymptomatic. Some patients may have a history of acute pancreatitis in whom the diagnosis of pseudocyst should also be considered. Presence of weight loss, jaundice, new onset diabetes should raise the suspicion of an underlying malignancy. Good-quality cross-sectional imaging is paramount. IPMN shows cystic lesion communicating with the pancreatic duct, which itself may show multifocal dilatations, with or without side branch dilatation [7]. Intra-cystic or intra-ductal nodules may be present. MRCP is helpful in visualization of ductal communication, as was the case in our patient. Resection is indicated in main duct and mixed type IPMNs, as well as select cases of branch duct type as they have a propensity to degenerate into malignancy [3].

Pancreatic NETs are typically solid, hypervascular lesions that show marked enhancement on contrast-enhanced imaging. However, no such lesion was identified on CECT scan of our patient. Peripheral contrast enhancement in a cystic PNET may be seen only transiently, and this remains a potential drawback of CECT in this scenario. It is also advisable to obtain baseline serum chromogranin levels and a 68-Gallium DOTATATE scan if PNET is being suspected. The former is of prognostic significance, while the latter helps to assess for multifocal disease and distant metastatic lesions. EUS has an established role in the evaluation of patients with cystic neoplasms, providing an opportunity to obtain cyst fluid for diagnosis and to perform FNAC (fine needle aspiration cytology) from suspicious areas [8]. Newer developments such as the use of elastography, contrast-enhanced EUS, and contrast enhanced harmonic (CH-EUS) have further improved the utility of EUS in decision-making in PNET [9]. It is well known that intra-operative ultrasound (IOUS) is a highly sensitive modality to identify small NETs, locate multiple tumors, define tumor extent and its relation to vascular structures [10]. However, this facility was not available at our centre.

There are many reports in which conditions such as chronic pancreatitis, autoimmune pancreatitis, serous cystadenoma, mucinous cystadenoma, PNET have masqueraded as IPMN [11-15]. Radiological appearance can be variable, and this worsens the diagnostic dilemma. While our patient had a well-defined unilocular cystic lesion with no mural nodules, Kirchgesner *et al* reported a multilocular cystic lesion in the head of the pancreas communicating with the MPD and associated with endoluminal solid nodules [16]. Peixoto *et al* also reported a regular walled cyst in the body–tail of the pancreas, while Chetty *et al* found a solid-cystic tumor situated wholly within the main pancreatic duct, presenting as an obstructing intraluminal polypoid lesion [17,18].

While PNETs are predominantly solid, cystic variants account for 7-17% of all resected PNETs [19]. In a study by Kawamoto *et al*, only 3.4 % of PNETs were purely cystic with an extremely thin wall mimicking cystic neoplasm [20]. These cystic PNETs are more commonly located in the

 https://doi.org/10.47892/rgp.2026.463.2218

body or tail of the pancreas, are hormonally inert, have lower Ki-67 index, and likely to have a less aggressive course [21]. Whether cystic PNET represents a distinct entity or merely represent cystic degeneration of a solid PNET is a matter of contention, although evidence from multiple studies favours the former hypothesis [19,22,23]. PNET may have an intraductal growth causing ductal obstruction and subsequent ductal dilatation producing an IPMN-like picture, as was the scenario in our case. In one systematic review, intra-ductally growing pattern in PNETs was reported to be of rare occurrence, having been demonstrated in only 20 cases, five of whom had provisional diagnosis of IPMN, with two cases of suspected malignancy [24]. Surgical resection remains the standard of care until strong evidence emerges favouring active surveillance [25]. Table 1 summarizes the characteristic features of pancreatic cystic lesions with MPD dilatation.

In conclusion, despite advances in various imaging modalities, accurate diagnosis of a pancreatic cystic lesion remains elusive. Although the diagnosis of cystic PNET is rare, it should be considered within the differential diagnosis of complex cystic lesions of the pancreas. IOUS, if available, should be used to identify any missed lesion in the substance of pancreas that may be the cause for ductal obstruction, and cystic dilatation.

REFERENCES

1. Scheiman JM. Cystic lesion of the pancreas. Gastroenterology. 2005;128(2):463-9. doi: 10.1053/j.gastro.2004.12.010.
2. Jana T, Shroff J, Bhutani MS. Pancreatic cystic neoplasms: Review of current knowledge, diagnostic challenges, and management options. J Carcinog. 2015;14:3. doi: 10.4103/1477-3163.153285.
3. 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):738-753. doi: 10.1016/j.pan.2017.07.007.
4. Figueiredo FA, Giovannini M, Monges G, Charfi S, Bories E, Pesenti C, *et al*. Pancreatic endocrine tumors: a large single-center experience. Pancreas. 2009;38(8):936-40. doi: 10.1097/MPA.0b013e3181b365db.
5. Verbeke CS. Endocrine tumours of the pancreas. Histopathology. 2010;56(6):669-82. doi: 10.1111/j.1365-2559.2010.03490.x.
6. Lewis RB, Lattin GE Jr, Paal E. Pancreatic endocrine tumors: radiologic-clinicopathologic correlation. Radiographics. 2010;30(6):1445-64. doi: 10.1148/rg.306105523.
7. Vege SS, Ziring B, Jain R, Moayyedi P; Clinical Guidelines Committee; American Gastroenterology Association. American gastroenterological association institute guideline on the diagnosis and management of asymptomatic neoplastic pancreatic cysts. Gastroenterology. 2015;148(4):819-22; quize12-3. doi: 10.1053/j.gastro.2015.01.015.
8. Rangwani S, Juakiem W, Krishna SG, El-Dika S. Role of Endoscopic Ultrasound in the Evaluation of Pancreatic Cystic Neoplasms: A Concise Review. Diagnostics (Basel). 2023;13(4):705. doi: 10.3390/diagnostics13040705.
9. Franchellucci G, Andreozzi M, Carrara S, De Luca L, Auriemma F, Paduano D, *et al*. Contrast Enhanced EUS for Predicting Solid Pancreatic Neuroendocrine Tumor Grade and Aggressiveness. Diagnostics (Basel). 2023;13(2):239. doi: 10.3390/diagnostics13020239.
10. Weinstein S, Morgan T, Poder L, Shin L, Jeffrey RB, Aslam R, *et al*. Value of Intraoperative Sonography in Pancreatic Surgery. J Ultrasound Med. 2015;34(7):1307-18. doi: 10.7863/ultra.34.7.1307.
11. Nakaji S, Hirata N, Fujii H, Iwaki K, Shiratori T, Kobayashi M, *et al*. A case of focal autoimmune pancreatitis (AIP) mimicking an intraductal papillary mucinous neoplasm (IPMN). Clin J Gastroenterol. 2013;6(4):329-33. doi: 10.1007/s12328-013-0392-z.
12. Mizuta N, Noda H, Kakizawa N, Toyama N, Konishi F. Chronic pancreatitis mimicking intraductal papillary mucinous neoplasm of the pancreas: Report of two cases. Jichi Med Univ J. 32;(2009):111-117.
13. Jee KN. Mass forming chronic pancreatitis mimicking pancreatic cystic neoplasm: A case report. World J Gastroenterol. 2018;24(2):297. doi: 10.3748/wjg.v24.i2.297.
14. Park M, Lee J, Kim Y, Yi KS, Cho BS, Choi C-H, *et al*. Pancreatic serous cystic neoplasm mimicking intraductal papillary mucinous neoplasm: Two case reports and literature review. Medicine (Baltimore). 2023;102:5(e32820). doi: 10.1097/MD.0000000000032820.
15. Akmercan A, Uprak TK, Bagci P, Atici AE. Pancreatic Neuroendocrine Tumor Mimicking Intraductal Papillary Mucinous Neoplasm: Case Report. Turkiye Klinikleri J Case Rep. 2021;29(3):105-8. doi: 10.5336/caserep.2021-81972.
16. Kirchgesner J, Colignon N, Balladur P, Fléjou JF, Svrcek M. Pancreatic neuroendocrine tumor mimicking a malignant intraductal papillary mucinous neoplasm. Pancreatology. 2016;16(2):290-2. doi: 10.1016/j.pan.2016.02.009.
17. Peixoto A, Pereira P, Lopes S, Macedo G. Pancreatic neuroendocrine tumour simulating an intraductal papillary mucinous neoplasm. Dig Liver Dis. 2015;47(3):256. doi: 10.1016/j.dld.2014.10.015.
18. Chetty R, El-Shinnawyl. Intraductal pancreatic neuroendocrine tumor. Endocr Pathol. 2009;20:262-6. doi: 10.1007/s12022-009-9093-z.
19. Carr RA, Bletsis P, Roch AM, House MG, Zyromski NJ, Nakeeb A, *et al*. Cystic pancreatic neuroendocrine tumors: A more favorable lesion? Pancreatology. 2019;19(2):372-376. doi: 10.1016/j.pan.2019.01.017.
20. Kawamoto S, Johnson PT, Shi C, Singhi AD, Hruban RH, Wolfgang CL, *et al*. Pancreatic neuroendocrine tumor with cystlike changes: evaluation with MDCT. AJR Am J Roentgenol. 2013;200(3):W283-90. doi: 10.2214/AJR.12.8941.
21. Koh YX, Chok AY, Zheng HL, Tan CS, Goh BK. A systematic review and meta-analysis of the clinicopathologic characteristics of cystic versus solid pancreatic neuroendocrine neoplasms. Surgery. 2014;156(1):83-96.e2. doi: 10.1016/j.surg.2014.03.026.
22. Singhi AD, Chu LC, Tatsas AD, Shi C, Ellison TA, Fishman EK, *et al*. Cystic pancreatic neuroendocrine tumors: a clinicopathologic study. Am J Surg Pathol. 2012;36:1666-73. doi: 10.1097/PAS.0b013e31826a0048.
23. Boninsegna L, Partelli S, D'Innocenzio MM, Capelli P, Scarpa A, Bassi C, *et al*. Pancreatic cystic endocrine tumors: a different morphological entity associated with a less aggressive behavior. Neuroendocrinology. 2010;92:246-51. doi: 10.1159/000318771.
24. Manuel-Vazquez A, Ramia JM, Latorre Fragua R, Valle-Rubio A, Arteaga-Peralta V, Ramiro-Pérez C, *et al*. Pancreatic Neuroendocrine Tumors and Intraductal Papillary Mucinous Neoplasm of the Pancreas: A Systematic Review. Pancreas. 2018;47(5):551-555. doi: 10.1097/MPA.0000000000001048.
25. Scott AT, Howe JR. Evaluation and Management of Neuroendocrine Tumors of the Pancreas. Surg Clin North Am. 2019;99(4):793-814. doi: 10.1016/j.suc.2019.04.014.