

[ CASE REPORT ]

## Pancreatic Colloid Carcinoma Presenting with Acute Pancreatitis

Masakuni Fujii, Yuki Okamoto, Shin-ichi Fujioka and Junji Shiode

### Abstract:

Pancreatic colloid carcinoma, also known as mucinous non-cystic carcinoma, is a rare subtype of pancreatic cancer accounting for 1-3% of the pancreatic malignant neoplasms. We herein report a woman who initially presented for acute pancreatitis. Computed tomography showed pancreatic swelling due to acute pancreatitis and a 16-mm mass with an enhanced margin in the pancreatic tail. We performed endoscopic ultrasound fine-needle aspiration. The patient was diagnosed with pancreatic colloid carcinoma, and distal pancreatectomy was performed. This case indicates that pancreatic colloid carcinoma should be considered as a differential diagnosis of pancreatic tumor presenting with acute pancreatitis.

**Key words:** pancreatic colloid carcinoma, rare tumor, acute pancreatitis

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### Introduction

Pancreatic colloid carcinoma, also known as mucinous non-cystic adenocarcinoma, is a unique well-differentiated type of invasive adenocarcinoma of the pancreas (1). It is a rare subtype of pancreatic cancer that only accounts for only 1-3% of the malignant neoplasms of the exocrine pancreas. The incidence is suspected to be only a few cases per 1 million individuals per year (2-4). The clinical manifestations of pancreatic colloid carcinoma are similar to those of invasive pancreatic adenocarcinoma; however, the histopathology has unique features (2).

We herein report our experience with a case of pancreatic colloid carcinoma complicated by acute pancreatitis.

### Case Report

A 71-year-old Japanese woman with hypertension, hyperlipidemia, chronic nephritis, and hypothyroidism presented to our institution with nausea, vomiting, and epigastric pain with radiation to the back of 2 days' duration. She was a never smoker and denied alcohol abuse.

Her vital signs were notable for tachycardia at 112 beats/min, and she was hypertensive at 160/108 mmHg. A physi-

cal examination was notable for epigastric pain. Pertinent laboratory findings included a leukocyte count of 17,190/ $\mu$ L, C-reactive protein 1.37 mg/dL, amylase 3,982 U/L (40-126 U/L), and lipase 3,482 U/L (11-59 U/L).

Abdominal contrast-enhanced computed tomography (CT) showed pancreatic swelling due to acute pancreatitis and a 16-mm mass with an enhanced margin in the pancreatic tail (Fig. 1). The acute pancreatitis was mild. The patient was treated with fasting, transfusion, and antibiotics. The treatment effect was good. The abdominal symptoms and vital signs improved within six days. The amylase and lipase levels, leukocyte count, and C-reactive protein levels normalized within nine days. The patient recovered and was able to consume a sufficient amount of food within nine days. Acute pancreatitis improved with conservative management as a further evaluation of her mass was pursued.

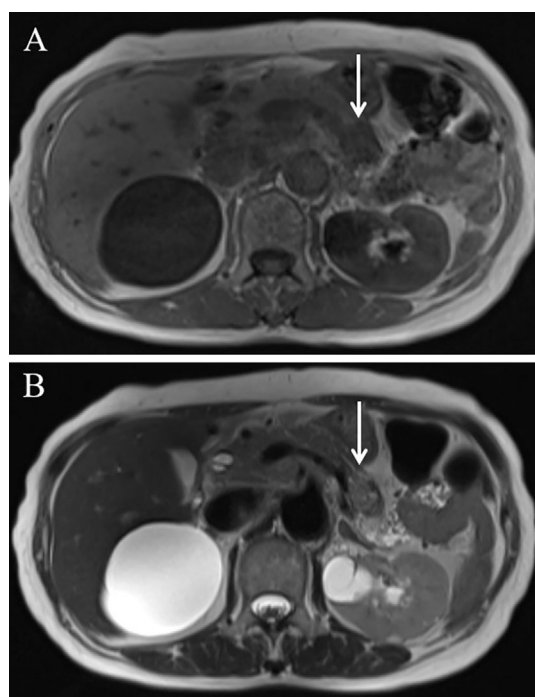
Abdominal ultrasound showed a hypoechoic uniform mass of 18×17 mm in the pancreatic tail (Fig. 2). On magnetic resonance imaging (MRI), the tumor had a low T1 and high T2 signal (Fig. 3). Magnetic resonance cholangiopancreatography (MRCP) showed the dilation of the pancreatic duct within the tail of the pancreas due to the tumor (Fig. 4). We then sent the patient for endoscopic retrograde cholangiopancreatography (ERCP), which revealed a normal duodenal papilla but disconnection of the pancreatic tail duct



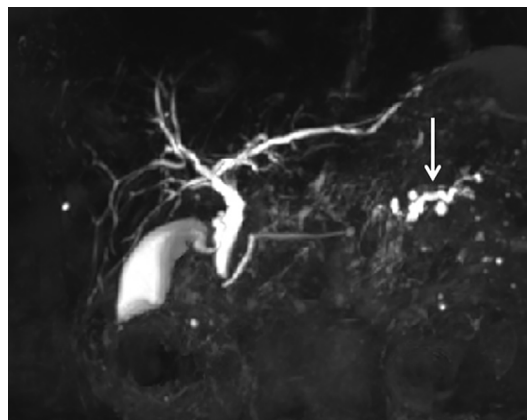
**Figure 1.** Abdominal contrast-enhanced computed tomography (CT) showed pancreatic swelling due to acute pancreatitis (arrowhead) and a 16-mm mass with an enhanced margin in the pancreatic tail (arrow).



**Figure 2.** Abdominal ultrasound showed a hypoechoic boundary-clear internal uniform mass 18×17 mm in size in the pancreatic tail (arrow).



**Figure 3.** On magnetic resonance imaging (MRI), the tumor had a low T1 (arrow) (A) and high T2 (arrow) (B) signal.



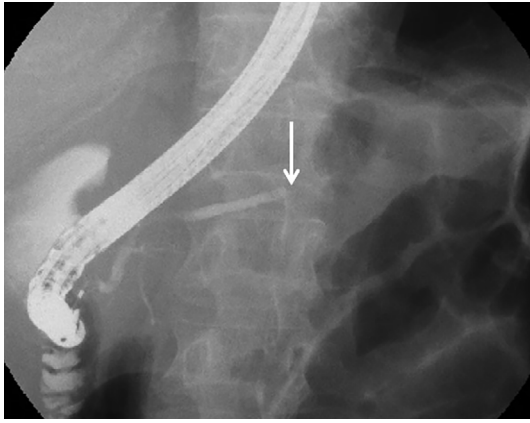
**Figure 4.** Magnetic resonance cholangiopancreatography (MRCP) showed the dilation of the pancreatic duct within the tail of the pancreas due to the tumor (arrow).

due to the tumor (Fig. 5). Pancreatic juice cytology was negative. [18F]-fluorodeoxyglucose positron emission tomography/CT (FDG PET/CT) showed an abnormal accumulation (maximum standardized uptake value: 4.35) (Fig. 6).

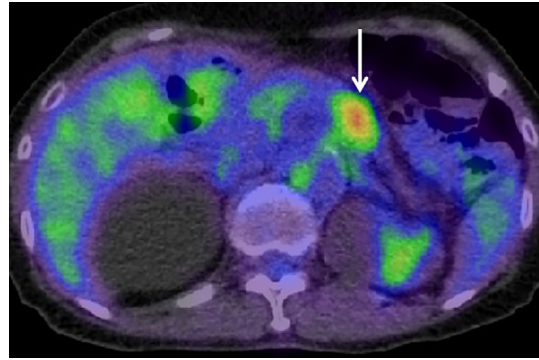
We suspected pancreatic ductal adenocarcinoma and performed endoscopic ultrasound fine-needle aspiration (EUS-FNA) through the transgastric route using a 25-gauge FNA needle. The pathological findings revealed rich mucin and glands composed of malignant cells with marked variation in nuclear size and disorderly arrangement of nuclei (Fig. 7). We diagnosed the patient with pancreatic colloid carcinoma, and she underwent distal pancreatectomy with

splenectomy 35 days after the onset of acute pancreatitis. The resected specimen was a pale beige tumor, 35×25 mm in size and rich in mucin. The tumor surface showed gelatinous nodules containing highly viscous mucus, and whitish fibrous tissue was observed at the margins and between the nodules.

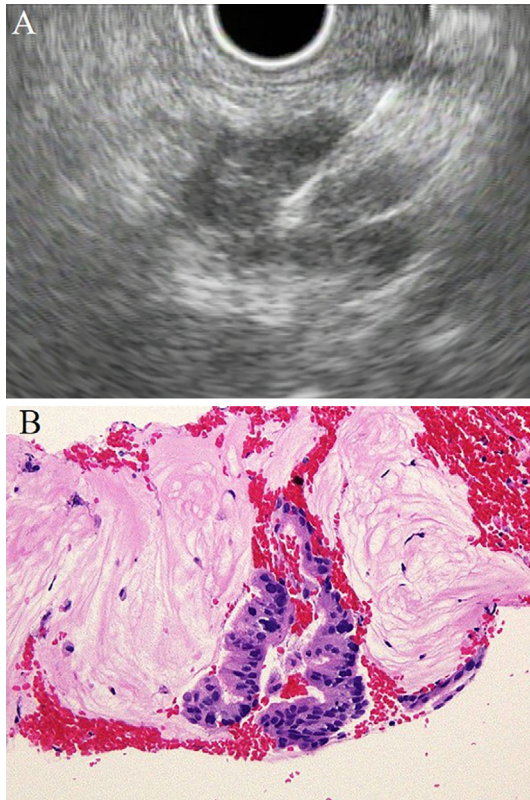
A histopathological examination revealed mucin pools with floating clumps and strands of malignant tumor cells. The mucin pools had fibular membrane and they were lined by a cuboidal, cylindrical epithelium with tumor cells (Fig. 8), confirming our diagnosis of pancreatic colloid carcinoma, stage 1B, T2N0M0. There were no pseudocysts or intraductal papillary mucinous neoplasms (IPMNs) in the pathological findings of the resection specimen. At six months after surgery, S1 (tegafur, gimeracil and oteracil potassium) was administered as adjuvant chemotherapy. This patient has had no recurrence for 2,568 days since surgery



**Figure 5.** Endoscopic retrograde cholangiopancreatography (ERCP) revealed the disconnection of the pancreatic tail duct due to the tumor (arrow).



**Figure 6.** [18F]-fluorodeoxyglucose positron emission tomography/computed tomography (FDG PET/CT) showed an abnormal accumulation (maximum standardized uptake value, 4.35) (arrow).



**Figure 7.** Endoscopic ultrasound fine-needle aspiration (EUS-FNA) through the transgastric route using a 25-gauge FNA needle (A) revealed rich mucin and glands composed of malignant cells with marked variation in nuclear size and disordered arrangement of nuclei (Hematoxylin and Eosin staining,  $\times 40$ ) (B).

and is currently alive.

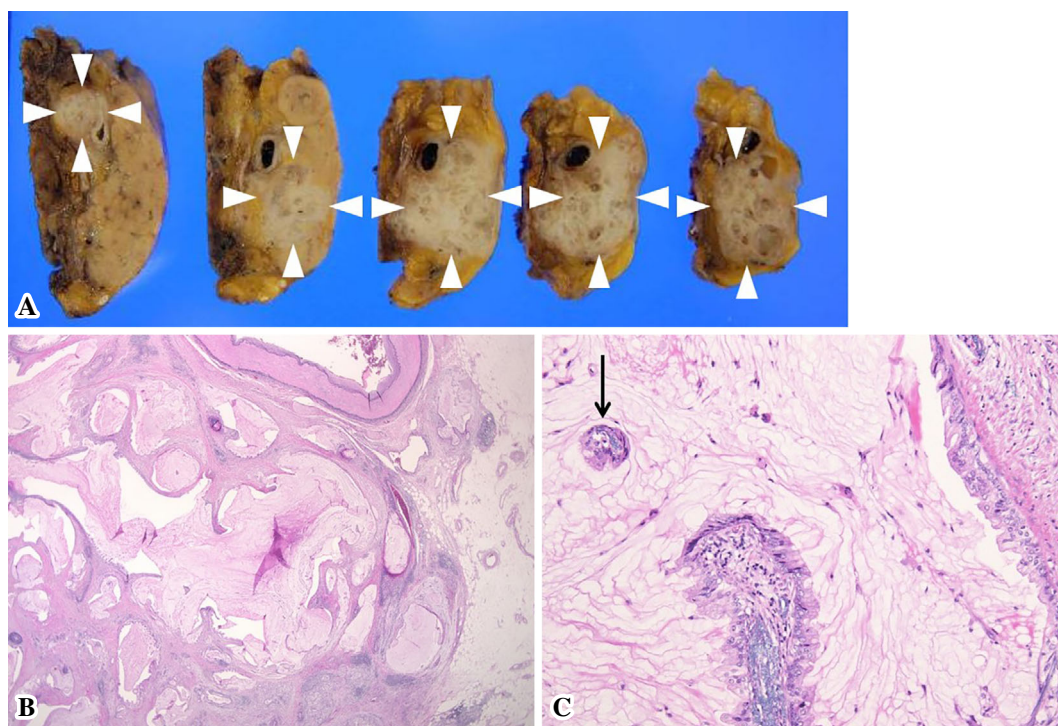
## Discussion

Pancreatic colloid carcinoma was first described in a case report by Muir in 1952 (1, 5). In a study of 17 patients di-

agnosed with pancreatic colloid carcinoma in 2001, Adsay et al. considered it a rare mucous-producing tumor that should be distinguished from other pancreatic neoplasms (1, 6). Colloid carcinoma of the pancreas, also known as mucinous noncystic carcinoma, is a rare pancreatic neoplasm and represents about 1% of all pancreatic tumors (3, 6, 7).

A microscopic investigation of pancreatic colloid carcinoma reveals that the tumors consist of separating pools containing mucin and floating clumps or strands of malignant cells; the mucin pools are lined in part by the cuboidal or well-differentiated epithelium (2). According to the Armed Forces Institute of Pathology (AFIP) definition established in 2007, the colloid component of colloid carcinoma should comprise at least 80% of the neoplasm (1). Colloid carcinomas most often develop in the head of the pancreas (2). The diameter of colloid carcinoma ranges between 1.2 and 16.0 cm, which is greater than that of tubular ductal adenocarcinoma at presentation (2, 3, 6). Pancreatic colloid carcinoma is derived from a subtype of invasive pancreatic ductal adenocarcinoma, IPMN and mucinous cystic neoplasm (MCN). Many pancreatic colloid carcinomas are reportedly derived from IPMN (7). The present case did not show any pathological findings of IPMN or MCN, suggesting a subtype of invasive pancreatic ductal adenocarcinoma. MUC staining is useful in the diagnosis of pancreatic colloid carcinoma. MUC1 appears to be a marker of an aggressive phenotype, such as pancreatic ductal carcinoma. MUC2 is commonly expressed in tumors with an indolent course, such as some IPMNs, and specifically in pancreatic colloid carcinomas involving different organs (1, 8, 9).

Various reports have described the prognosis of pancreatic colloid carcinoma. Pancreatic colloid carcinomas have a better median overall survival (40-60%) than tubular carcinomas, which behave similarly to pancreatic ductal adenocarcinoma (5-year overall survival: 10-20%) (7, 10-12). Pancreatic colloid carcinomas, as well as those of other organs (e. g. breast and skin) tend to exhibit an indolent course with a favorable 5-year survival of 57-72% after resection (1, 5, 13, 14). However, Seidel et al. reported that the



**Figure 8.** The tumor surface showed gelatinous nodules containing highly viscous mucus, and whitish fibrous tissue was observed at the margins and between the nodules (arrowhead) (A). A histopathological examination revealed mucin pools with floating clumps and strands of malignant tumor cells (arrow). The mucin pools had a fibular membrane and were lined by a cuboidal, cylindrical epithelium with tumor cells [Hematoxylin and Eosin (H&E) staining,  $\times 4$ ] (B), (H&E staining,  $\times 40$ ) (C).

prognosis (5-year overall survival 29%) was similar to that of pancreatic ductal adenocarcinoma (15). Furthermore, the prognosis in 13 resected cases (median survival: 24 months, 5-year overall survival: 30%) was reported to be similar to that of ductal adenocarcinoma (16). No specific guidelines exist for the treatment of colloid carcinoma at present; however, the mainstay treatment should be considered to be surgery if there is no distant metastasis, surrounding organ invasion, or vessel encasement (2). The resection rate of pancreatic colloid carcinoma was 75% in a report of 28 cases (17). There is no consensus concerning the appropriate chemotherapy for pancreatic colloid carcinomas (18). The accumulation of more cases is needed for the establishment of further supporting evidence.

Our case had good prognosis after surgery. The tumor caused acute pancreatitis, so we were able to detect the lesion relatively early. The favorable outcomes of pancreatic colloid carcinoma highlight the importance of its detection, diagnosis, and prompt treatment. To improve the preoperative diagnostics, it is important to be aware of the imaging characteristics, with characteristic CT and MRI findings reported to be an enhanced tumor border in the early phase and slightly dendritically enhanced inside of the tumor in the late phase on cystography CT. The mass tends to appear round or with lobular margins with distinct boundaries (2, 19). MRI shows a low signal on T1-weighted imaging and a high signal on T2-weighted imaging. MRCP shows a lighter high signal than normal water or mucus, re-

flecting the highly viscous nature of the gelatinous mucous nodule (20). There are no unique findings on PET/CT for pancreatic colloid carcinomas, but PET/CT may still be performed to determine the potential presence of locoregional and distant metastases as well as the metabolic tumor activity, which may further improve patient management (1). Our case presented with relatively typical imaging findings.

Due to the rarity of the diagnosis, pancreatic colloid carcinomas are typically diagnosed after surgical resection rather than based on a preoperative workup (7). The diagnosis of pancreatic colloid carcinoma may be challenging, as the yield from pancreatic juice cytology via ERCP is low, and EUS-FNA requires expertise training and carries its own risk of potential primary tumor seeding (2, 21). However, while EUS-FNA comes with risks, it allows large amounts of mucin and benign-appearing glandular epithelium or single cells to be observed. FNA is reportedly a useful diagnostic modality and can be used to provide a rapid and accurate diagnosis in cases of pancreatic colloid carcinoma (2). We suspected pancreatic ductal adenocarcinoma in the present study and performed EUS-FNA, which provided findings prompting us to suspect pancreatic colloid carcinoma preoperatively. Recently, preoperative chemotherapy for resectable pancreatic cancer has been performed increasingly frequently (22). Therefore, the preoperative diagnosis of pancreatic colloid carcinoma and conventional pancreatic cancer is very important in determining the treatment strategy. Satoh et al. reported a case of pancreatic colloid carci-

noma diagnosed by contrast-enhanced endoscopic ultrasound-guided fine-needle aspiration (23). As above, the accumulation of more cases is needed for the establishment of further supporting evidence.

Clinically, pancreatic colloid carcinoma presents with signs and symptoms similar to those of pancreatic ductal adenocarcinoma, such as abdominal pain, jaundice, weight loss, an abdominal mass, and acute pancreatitis. However, acute pancreatitis often leads to a situation enabling the discovery of pancreatic cancer. Acute pancreatitis may constitute an early symptom of pancreatic cancer, as around 1% of acute pancreatitis admissions are due to pancreatic cancer (24, 25). For our case, we suspected that the obstruction of the pancreatic duct by mucus or the mass had caused acute pancreatitis. When treating acute pancreatitis, we should be alert for the presence of pancreatic cancer, including pancreatic colloid carcinoma.

The incidence of pancreatic colloid carcinoma appears to exhibit no gender predominance, and the median age at the first presentation has been reported to be 59-69 years old. Tumor biomarkers, including CEA and CA19-9, are also elevated (2-4), although in our case they were normal. In addition, a unique feature of our case is that it was diagnosed in the setting of acute pancreatitis. Furthermore, our case has been followed for 2,568 days, proving a unique addition to the literature, where cases with long-term follow-up of pancreatic colloid carcinoma outcomes are scarce.

In conclusion, pancreatic colloid carcinoma, also known as mucinous non-cystic carcinoma, is a rare subtype of pancreatic cancer. Although pancreatic colloid carcinoma is a disease that is difficult to diagnose, the utility of understanding distinct imaging characteristics allows for both a prompt diagnosis and prompt management. In addition, it is always important to consider a mass effect when assessing a patient with an unknown pancreatitis etiology, without forgetting that pancreatic colloid carcinoma may be the culprit lesion.

**The authors state that they have no Conflict of Interest (COI).**

## References

- Jiang L, Tang Q, Panje CM, Nie H, Zhao G, Shi H. Assessment of pancreatic colloid carcinoma using 18F-FDG PET/CT compared with MRI and enhanced CT. *Oncol Lett* **16**: 1557-1564, 2018.
- Gao Y, Zhu YY, Yuan Z. Colloid (mucinous non-cystic) carcinoma of the pancreas: a case report. *Oncol Lett* **10**: 3195-3198, 2015.
- Liszka L, Zielinska-Pajak E, Pajak J, Gołka D. Colloid carcinoma of the pancreas: review of selected pathological and clinical aspects. *Pathology* **40**: 655-663, 2008.
- Whang EE, Danial T, Dunn JC, et al. The spectrum of mucin-producing adenocarcinoma of the pancreas. *Pancreas* **21**: 147-151, 2000.
- Muir EG. Colloid carcinoma of the pancreas. *Br J Surg* **40**: 177, 1952.
- Adsay NV, Pierson C, Sarkar F, et al. Colloid (mucinous noncystic) carcinoma of the pancreas. *Am J Surg Pathol* **25**: 26-42, 2001.
- Orcutt ST, Coppola D, Hodul PJ. Colloid carcinoma of the pancreas: case report and review of the literature. *Case Rep Pancreat Cancer* **2**: 40-45, 2016.
- Nakata K, Ohuchida K, Aishima S, et al. Invasive carcinoma derived from intestinal-type intraductal papillary mucinous neoplasm is associated with minimal invasion, colloid carcinoma, and less invasive behavior, leading to a better prognosis. *Pancreas* **40**: 581-587, 2011.
- Adsay NV, Merati K, Nassar H, et al. Pathogenesis of colloid (pure mucinous) carcinoma of exocrine organs: coupling of gel-forming mucin (MUC2) production with altered cell polarity and abnormal cell-stroma interaction may be the key factor in the morphogenesis and indolent behavior of colloid carcinoma in the breast and pancreas. *Am J Surg Pathol* **27**: 571-578, 2003.
- Waters JA, Schnelldorfer T, Aguilar-Saavedra JR, et al. Survival after resection for invasive intraductal papillary mucinous neoplasm and for pancreatic adenocarcinoma: a multi-institutional comparison according to American Joint Committee on Cancer Stage. *J Am Coll Surg* **213**: 275-283, 2011.
- Poultides GA, Reddy S, Cameron JL, et al. Histopathologic basis for the favorable survival after resection of intraductal papillary mucinous neoplasm-associated invasive adenocarcinoma of the pancreas. *Ann Surg* **251**: 470-476, 2010.
- Koh YX, Chok AY, Zheng HL, Tan CS, Goh BK. Systematic review and meta-analysis comparing the surgical outcomes of invasive intraductal papillary mucinous neoplasms and conventional pancreatic ductal adenocarcinoma. *Ann Surg Oncol* **21**: 2782-2800, 2014.
- Mostafa ME, Erbarut-Seven I, Pehlivanoglu B, Adsay V. Pathologic classification of "pancreatic cancers": current concepts and challenges. *Clin Oncol* **6**: 59, 2017.
- D'Angelica M, Brennan MF, Suriawinata AA, Klimstra D, Conlon KC. Intraductal papillary mucinous neoplasms of the pancreas: an analysis of clinicopathologic features and outcome. *Ann Surg* **239**: 400-408, 2004.
- Seidel G, Zahurak M, Iacobuzio-Donahue C, et al. Almost all infiltrating colloid carcinomas of the pancreas and periampullary region arise from in situ papillary neoplasms: a study of 39 cases. *Am J Surg Pathol* **26**: 56-63, 2002.
- Nishi M, Kawasaki H, Fujii M, et al. Two cases of mucinous carcinoma of the pancreas. *Suizo* **29**: 98-104, 2014 (in Japanese, Abstract in English).
- Saegusa H, Nakata S, Watanabe M. Mucinous carcinoma of the pancreas: a case report and literature review. *Suizo* **29**: 749-755, 2014 (in Japanese, Abstract in English).
- Rubio-Perez I, Martin-Perez E, Sanchez-Urdazpal L, Corbaton P, Larrañaga E. Colloid carcinoma of the pancreas: a distinct pancreatic neoplasm with good prognosis. Report of a case. *JOP* **13**: 219-221, 2012.
- Ren FY, Shao CW, Zuo CJ, Lu JP. CT features of colloid carcinomas of the pancreas. *Chin Med J* **123**: 1329-1332, 2010.
- Yoon MA, Lee JM, Kim SH, et al. MRI features of pancreatic colloid carcinoma. *AJR Am J Roentgenol* **193**: 308-313, 2009.
- Parwani AV, Ali SZ. Pathologic quiz case: a 52-year-old woman with jaundice and history of necrotizing pancreatitis. Primary colloid carcinoma of the pancreas. *Arch Pathol Lab Med* **129**: 255-256, 2005.
- Motoi F, Kosuge T, Ueno H, et al. Randomized phase II/III trial of neoadjuvant chemotherapy with gemcitabine and S-1 versus upfront surgery for resectable pancreatic cancer (Prep-02/JSAP05). *Jpn J Clin Oncol* **49**: 190-194, 2019.
- Satoh T, Ishiwatari H, Sasaki K. Pancreatic colloid carcinoma diagnosed by contrast-enhanced endoscopic ultrasound-guided fine-needle aspiration. *Dig Endosc* **32**: 150, 2020.
- Kirkegård J, Gaber C, Lund JL, et al. Acute pancreatitis as an early marker of pancreatic cancer and cancer stage, treatment, and

prognosis. *Cancer Epidemiol* **64**: 101647, 2020.  
25. Lankisch PG, Apte M, Banks PA. Acute pancreatitis. *Lancet* **386**: 85-96, 2015.

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