AG-2020-58
doi.org/10.1590/S0004-2803.202000000-60

Malignant branch duct intraductal papillary mucinous neoplasia mimicking pancreatic pseudocyst

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Received: 14 April 2020
Accepted: 16 June 2020

After episodes of acute pancreatitis (AP), it is common to find local complications as pancreatic pseudocyst (PP) or walled-off necrosis (WON). The PP can be asymptomatic[1]. Even though the diagnosis of PP can be straightforward with clinical settings and imaging exams, some patients may require further evaluation if the clinical presentation or the imaging findings don't fully support the diagnosis[1]. The PP is often self-limited and can regress completely or decrease the size in one year[2].

68-year-old man presenting nausea, vomiting and strong abdominal pain located in the upper abdomen. A year ago, he presented the same symptoms and was diagnosed with AP. His medical history included replacement of the ascending aorta and aortic valve, dyslipidemia and systemic arterial hypertension.

At the physical examination he was in a good condition. Laboratory tests presented amylase 753 U/L, lipase 10,000 U/L and CA19.9 2.8 U/L. Computerized tomography showed a thin-walled septa cyst in the head of pancreas (49 x 45 mm), not identified in the routine CT performed during the first episode of AP. Magnetic resonance cholangiopancreatography imaging showed compression of the main pancreatic duct (MD) and common bile duct by the cyst (FIGURE 1).

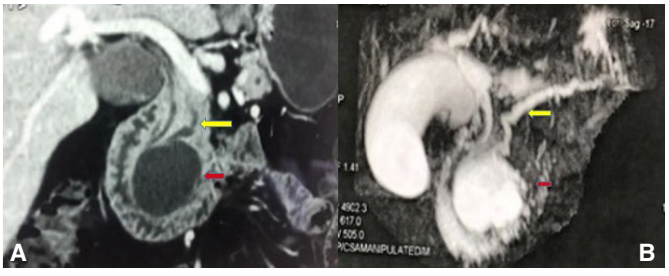

FIGURE 1. (A) CT showed a thin-walled cyst (red arrow) in the head of pancreas 49 x 45 mm with compression of the CBD and MD (yellow arrow). (B) MRI/MRCP pancreatic head cystic lesion with thick wall (red arrow) and compression of the CBD and the MD (yellow arrow).

EUS-FNA revealed a cystic lesion (6.5 x 5.3 cm) with thickened wall and intramural nodule (FIGURE 2). The aspirated liquid was yellow and the biochemistry presented amylase 8 U/L, CEA 651.8 U/L and CA19.9 140,000 U/L. Microhistology found foveolar and atypical cylindrical cells, basal nuclei and mucinous cytoplasm, confirming the diagnosis of an intraductal papilary mucinous neoplasia (IPMN).

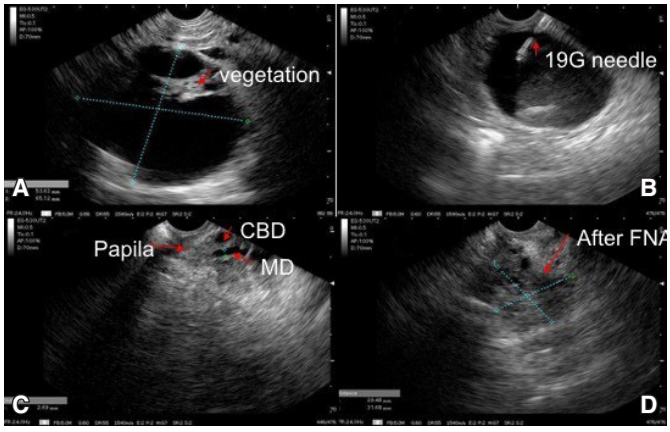

FIGURE 2. EUS imaging. (A) pancreatic cystic lesion with a thick wall, like a vegetation inside; (B) moment of the FNA with a 19G needle; (C) papillary region near to the pancreatic cystic lesion; and (D) imaging after the complete aspiration of the cyst.

The duodenopancreatectomy (DP) was the treatment. Final diagnosis was moderately differentiated invasive ductal adenocarcinoma, associated with IPMN. The epithelium was pancreatobiliary (MUC1+ and MUC5AC+) in branch duct (BD), with high grade dysplasia and invasive carcinoma of 1.2 cm – free margins. One metastatic lymphnode near to the hepatic artery was found. The final pathological staging was pT1cN1 (FIGURE 3 and E-VIDEO*).

Declared conflict of interest of all authors: none
Disclosure of funding: no funding received
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*** Video: https://www.youtube.com/watch?v=F4QTRgtOiTM**

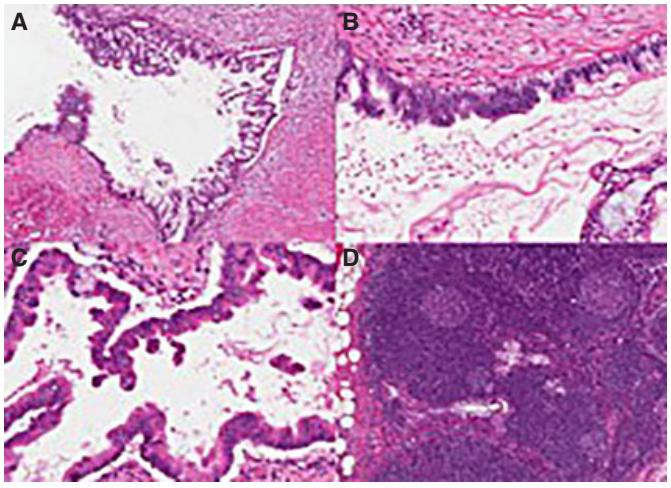

FIGURE 3. The diagnosis was moderately differentiated invasive ductal adenocarcinoma (D), associated with IPMN (A, B, and C), pancreatobiliary epithelium (B and C) in branch duct.

IPMN is a disease difficult to diagnose[3]. More than 60% of patients are men`s aged between 50–70, the same as our subject. Most patients with IPMN are asymptomatic, but the occurrence of two episodes of AP, as we observed in our case, seems to be awarning regarding the evolution of this disease.

When presenting symptoms, about 40% of IPMNs may have invasive carcinoma or high-grade dysplasia[4]. Invasive carcinoma can be detected in MD-IPMN and mixed-IPMN in up to 45% of cases. On the other hand, even if it is a small cyst (<3.0 cm) without worrisome features, BD-IPMNis 15%, as we could observe in our patient[5].

The immunophenotypes of the epithelium found in all types of IPMN are gastric (49 to 66%), intestinal (18 to 36%), pancreatobiliary (7 to 18%) and oncocytic (1 to 8%). In our case, the lining epithelium found was the pancreatobiliary, which has the greatest chance of malignancy[6]. This epithelium can progress to aggressive ductal adenocarcinoma. However, our patient has a better prognosis, since we detected an early ductal adenocarcinoma with an invasion lower than 11 mm, characterized as a T1c[7]. After five years of follow-up, our patient is healthy.

Authors' contribution

Ardengh JC: performed the procedure and the diagnosis. Taglieri E: references search. Ardengh AO: English text and video edition. Micelli-Neto O: figure edition.

Orcid

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