Case report

Pancreatic intraductal papillary mucinous neoplasm associated colloid carcinoma

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ABSTRACT

Colloid carcinomas are rare pancreatic tumors characterized by the presence of mucin pools with scarce malignant cells. Most of these neoplasms arise from intestinal-type intraductal papillary mucinous neoplasms (IPMNs). We report a case of a 77-year-old male patient who presented with weight loss, asthenia, lumbar pain and diabetes. Imaging studies revealed a mixed-type IPMN with high-risk features and a possible invasive component. The patient underwent surgical resection and the histology confirmed an invasive colloid carcinoma of the pancreas associated with an intestinal-type IPMN. Although invasive ductal and colloid carcinomas may look similar on imaging studies, its distinction is important because the latter have a better prognosis.

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Introduction

Invasive carcinoma arising from intraductal papillary mucinous neoplasms (IPMNs) can be classified as tubular (resembling the classical pancreatic ductal carcinomas) or as colloid types [1]. The colloid carcinomas of the pancreas are histologically similar to colloid tumors located in other exocrine epithelia, such as in the breast and colon. These tumors have an indolent behavior and carry a better prognosis than invasive ductal carcinomas of the pancreas [2].

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Case report

A 77-year-old male patient presented with a 2-month history of weight loss, asthenia, lumbar pain, and new onset of diabetes mellitus. Past medical history included abdominal aneurysm, dyslipidemia, and high blood pressure. Moreover, he reported a history of tobacco and alcohol abuse in the past.

Prior workup on another institution included a colonoscopy to exclude colorectal cancer and an abdominal computed tomography (CT). The abdominal CT raised suspicion of an IPMN with a solid component in the head of the pancreas.

The biochemical evaluation revealed normal liver function tests and a normal level of Carbohydrate Antigen 19-9 (CA 19-9) (17 U/mL for a normal <37 U/mL). The levels of carcinoembryonic antigen (CEA) were increased (6.5 ng/mL, normal <2.5 ng/mL).

To better characterize those findings, the patient underwent magnetic resonance cholangiopancreatography (MRCP). The MRCP revealed several pancreatic cystic lesions in association with a diffuse dilatation of the main pancreatic duct (up to 18 mm in diameter), which was filled with hyperintense content on T2-weighted images (Fig. 1). Some enhancing solid mural nodules were seen along the main pancreatic duct in the pancreas head and body, with restriction to diffusion (Fig. 2). These findings are in accordance with a mixed-type IPMN with high-risk features and a likely invasive component. The cystic mass contacted the first part of the duodenum, the intra-pancreatic portion of the common bile duct, and the gastric antrum. There were no liver metastasis, peritoneal nodules, or ascitis.

Total pancreatectomy with splenectomy and distal gastrectomy was performed, to achieve negative surgical margins. The postoperative period was uneventful.
Fig. 3 – Histopathological findings. (A) Gross images of sequential sections of the resected specimen at the pancreatic head level showing pancreatic cysts (arrows) and dilation of the main pancreatic duct. (B and C) Histological analysis reveal large mucin pools with scarce malignant cells, some arranged into small groups (arrowhead), findings that are typical of colloid carcinoma (Hematoxylin and Eosin [H&E], scale bar corresponds to 100 μm [B] and 200 μm [C]). (D) Other tumor areas showed isolated malignant cells floating inside the mucinous lakes (H&E, scale bar corresponds to 50 μm). (E) Associated intestinal-type IPMN with low-grade (white arrow) to high-grade dysplasia (black arrow) (H&E, scale bar corresponds to 100 μm). IPMN, intraductal papillary mucinous neoplasm.

Fig. 4 – Intestinal-type IPMN: immunohistochemical staining show that the neoplastic cells display expression for the markers CK7, CK20, CDX-2, MUC2, and MUC5AC (scale bar corresponds to 1 mm). H&E, Hematoxylin and Eosin; CK, cytokeratin; CDX-2, caudal-related homeobox gene 2; MUC, mucin; IPMN, intraductal papillary mucinous neoplasm.

The histologic examination revealed an invasive colloid carcinoma in the pancreas head and body, measuring more than 40 mm, associated with an intestinal mixed-type IPMN (Figs. 3 and 4). There was high-grade dysplasia in the main pancreatic duct. The circumferential margins of resection were negative, and all the lymph nodes were negative for malignant cells. Furthermore, there was no perineural or lymphovascular invasion.

Discussion

IPMNs are mucin-producing exocrine pancreatic neoplasms. According to the anatomic ductal involvement, they can be classified into main duct, side branch, and mixed type [1,3].

On the other hand, according to their histological features, IPMNs are divided into 4 subtypes: gastric, intestinal, pan-
creatobiliary, and oncocytic [1,4], each with different malignant potential, the pancreatobiliary type being the one with a higher risk of malignancy [4].

Features that suggest malignancy of an IPMN are jaundice, a solid component or enhancing mural node(s) measuring 5 mm or more, a main pancreatic duct dilation of 10 mm or more, and cystic lesions measuring more than 3 cm in size [4,5].

Most of the invasive carcinomas arising from IPMN are ductal (tubular) carcinomas, resembling classic ductal adenocarcinoma. Nevertheless, a smaller number are colloid carcinomas, mixed or anaplastic tumors. Colloid carcinoma or mucinous noncystic carcinoma represents 1%-3% of all invasive pancreatic adenocarcinomas [6]. Typically, colloid carcinomas arise from intestinal-type IPMNs [1,7], whereas tubular carcinomas arise from pancreatobiliary and gastric types IPMNs [1].

Histologically, colloid carcinomas are characterized by extracellular mucin lakes with sparse malignant cells [6,8]. Due to their origin, they express the intestinal markers MUC2 and CDX2 [7].

Colloid carcinomas have a better prognosis than tubular carcinomas [1]. Its 5-year survival rate is estimated to be 57%, whereas that of tubular carcinomas is 12% [8].

IPMN-associated tubular and colloid carcinomas may look identical in imaging studies. Fouladi et al. [1] recently reviewed the role of some CT features to distinguish colloid from tubular carcinomas. A sudden caliper change of the main pancreatic duct associated with a solid mass that does not communicate with the duct is the most accurate sign to differentiate those tumors, suggesting a tubular carcinoma. Conversely, cystic masses communicating with the main pancreatic duct are more likely to represent colloid carcinomas.

Surgical treatment of main-duct and mixed IPMNs with worrisome features must be considered in all patients fit for surgery [5]. There are no established guidelines to specifically address the adjuvant treatment of resected invasive colloid carcinomas [1,3]. Adjuvant chemotherapy may improve survival in patients with tubular carcinomas but not in those with colloid carcinomas [7].

Long-term surveillance is required after resection of invasive IPMNs to detect a possible recurrence [3]. The disease-free of survival of recurrent resected IPMN-associated invasive neoplasms is of 29 months [5].

**Patient consent**

The authors obtained written informed from the patient for publication of this case report.

**REFERENCES**

[1] Fouladi DF, Raman SP, Hruban RH, Fishman EK, Kawamoto S. Invasive intraductal papillary mucinous neoplasms: CT features of colloid carcinoma versus tubular adenocarcinoma of the pancreas. Am J Roentgenol 2020;214(5):1092–100.

[2] Poultides GA, Reddy S, Cameron JL, Hruban RH, Pawlik TM, Ahuja N, et al. Histopathologic basis for the favorable survival after resection of intraductal papillary mucinous neoplasm-associated invasive adenocarcinoma of the pancreas. Ann Surg 2010;251(3):470–6.

[3] Hirono S, Yamaue H. Surgical strategy for intraductal papillary mucinous neoplasms of the pancreas. Surg Today 2020;50(1):50–5.

[4] Hecht EM, Khatri G, Morgan D, Kang S, Rho-sale 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 2020;46(4):1586–606.

[5] Del Chiario M, Besselink MG, Scholten L, Bruno MJ, Cahen DL, Gress TM, et al. European evidence-based guidelines on pancreatic cystic neoplasms. Gut 2018;67(5):789–804.

[6] Yoon MA, Lee JM, Se HK, Jae YL, Joon KH, Byung IC, et al. MRI features of pancreatic colloid carcinoma. Am J Roentgenol 2009;193(4):W308–13.

[7] Marchegiani G, Andrianello S, Dal Borgo C, Secchettin E, Melisi D, Maleo G, et al. Adjuvant chemotherapy is associated with improved postoperative survival in specific subtypes of invasive intraductal papillary mucinous neoplasms (IPMN) of the pancreas: it is time for randomized controlled data. Hpb 2019;21(5):596–603.

[8] Asey NV, Pierson C, Sarkar F, Abrams J, Weaver D, Conlon KC, et al. Colloid (mucinous noncystic) carcinoma of the pancreas. Am J Surg Pathol 2001;25(1):26–42.