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Myofibroblastic Inflammatory Tumor Presenting as Recurrent Acute Pancreatitis and Obstructive Jaundice

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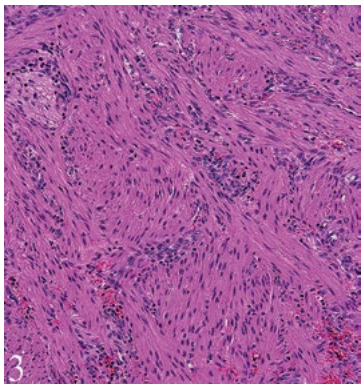

A 38 year-old male, smoker, with occasional alcohol consumption, episodes of high blood pressure, recurrent acute pancreatitis (2010 and 2014) and cholecystectomy for acalculous gallbladder was admitted in November 2014 with severe epigastric pain. Clinical examination revealed upper abdominal pain, altered general status, and mild jaundice. Blood tests evidenced increased amylase and lipase levels (7xULN), moderate hepatocytolysis, cholestatic jaundice with total bilirubin 9.1 mg/dL.

Esophagogastroduodenoscopy revealed edema in the first portion of the duodenum and a villous aspect of the mucosa in the second part of the duodenum with stenosis (Fig. 1). Multiple endobiopsies showed non-specific chronic mucosal inflammation. Computed tomography showed circumferential wall thickening (up to 15 mm), with low contrast caption in the second and third portion of the duodenum (Fig. 2), dilation of the intrahepatic and extrahepatic bile ducts and dilation of the Wirsung duct.

Treatment of acute pancreatitis was administered with temporary benefit.

In December 2014 the patient underwent cephalic duodenopancreatectomy, which revealed a tumor (55/40 mm) located in the muscular layer of the duodenum. Histopathological examination of the surgical specimen revealed a duodenal wall infiltrated by a diffuse tumor composed of uniform bland-looking spindle-cells, with focal atypia, arranged in fascicles, and inflammatory cell infiltrates with frequent eosinophils (Fig. 3). Immunohistochemistry showed positive smooth muscle actin in tumoral cells, CD 117 and CD34 negative, S100 positive in rare tumoral cells, Ki67 positive about 5-10%. Both histopathological and imunohistochemistry aspects confirmed the diagnosis of an inflammatory myofibroblastic tumor (IMT).

Duodenal IMTs are rare tumors; only seven cases have been up to now reported. They are of probable immunological aetiology and preoperatively indistinguishable from adenocarcinomas. Clinically, duodenal IMTs present with gastric outlet obstruction. Surgery is usually curative. Chemotherapy and radiotherapy are controversial, recommended only for incomplete resections and for aggressive IMTs. Recurrence is uncommon.

Our patient is presently asymptomatic, under follow-up, endoscopy and ultrasonography at 6 months, and annual magnetic resonance imaging.

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Conflicts of interest: None to declare.

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