Duodenal duplication cyst: a rare cause of recurrent pancreatitis

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We report the case of a 54-year-old man with a medical history of recurrent pancreatitis who was referred to our department for a novel episode of epigastric pain. A blood test confirmed the diagnosis of acute pancreatitis, and a biliary origin was suspected because of abnormal liver test results and dilatation of the biliary ducts as shown by US. A CT scan confirmed the bile duct dilatation and showed a voluminous cystic lesion of 40 × 30 mm developed from the duodenal wall (Fig. 1). Upper GI endoscopy identified a submucosal pedunculated lesion likely originating from the papilla area and completely obstructing its lumen. The cystic characteristics of the lesion were confirmed by EUS, but neither the typical double-wall sign nor the communication sign between the cyst and the main bile duct were observed. These findings were compatible with 2 hypotheses: a duodenal duplication cyst (DDC) or a type III choledochocele. Abdominal MRI with 3-dimensional construction was not able to discriminate between the 2 diagnoses. Because the patient was symptomatic and these lesions are associated with malignant potential, an endoscopic resection was proposed (Video 1, available online at www.VideoGIE.org).[1,2]

A progressive debulking piecemeal resection was initially proposed. After several resections, the cyst was clearly identified as arising from the papilla. Consequently, a papillectomy with biliopancreatic stent placement also

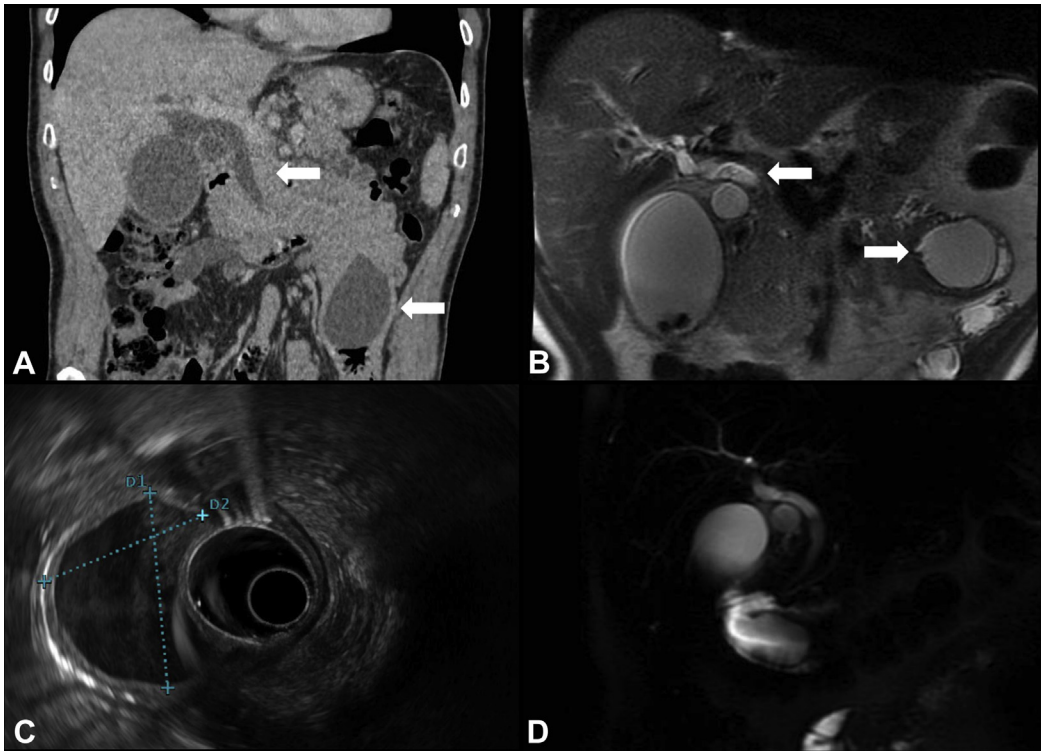

Figure 1. Multimodal morphologic evaluation. **A,** Contrast-enhanced CT coronal image showing dilatation of main duct and cystic lesion obstructing the duodenum (*white arrows* show main duct and cystic lesion). **B,** MRCP coronal image showing cystic lesion obstructing the lumen (*white arrows* show main duct and cystic lesion). **C,** EUS view of lesion measuring 38 (*D1*) × 32 (*D2*) mm. The cyst contains a heterogeneous material compatible with sludge. **D,** MRCP 3-dimensional construction showing a regular cholangiopancreatogram without direct connection to the cyst.

Written transcript of the video audio is available online at www.VideoGIE.org.

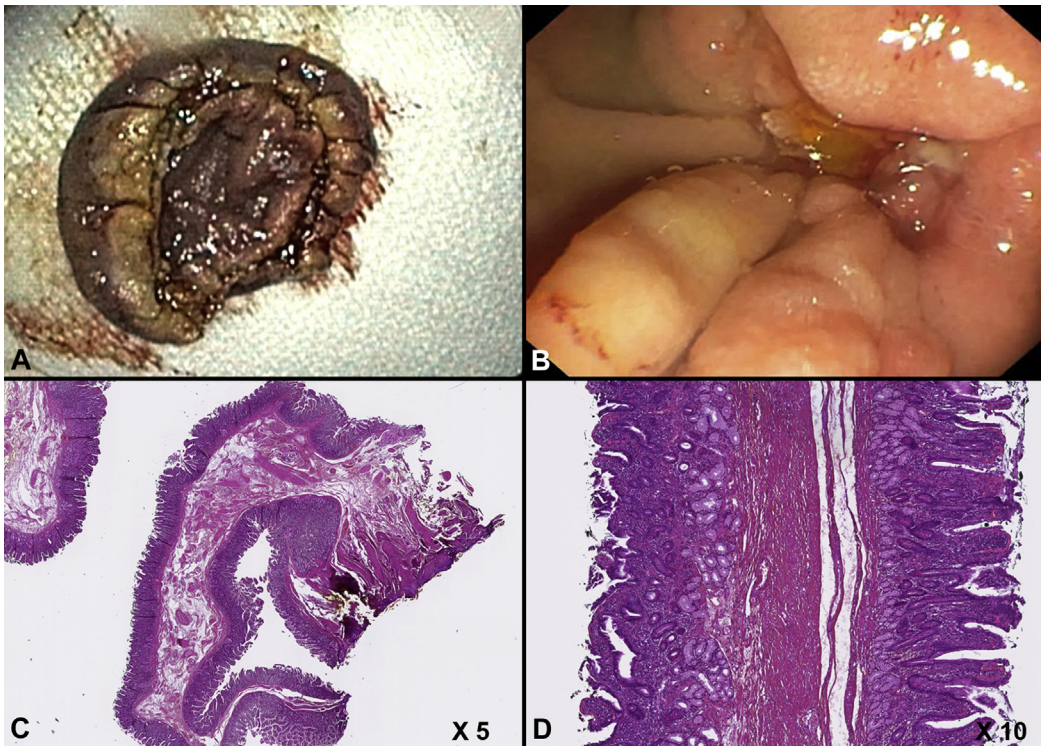

Figure 2. Endoscopic and histologic results after resection. **A,** Macroscopic view of first resection showing digestive mucosa covering both sides of the lesion. **B,** Endoscopic view at 1 month identifying complete re-epithelialization of the resected area. Both pancreatic and biliary orifices are easily identified. **C, D,** Histologic analysis of duodenal duplication cyst after hematoxylin-eosin-saffron staining, at different levels of magnification, showing typical duodenal mucosa lining both lumina, separated by a submucosal layer and containing smooth muscle.

was performed. No adverse events occurred. Endoscopic evaluation 1 month after the procedure showed a completely re-epithelialized papilla. Histologic analysis confirmed the diagnosis of benign DDC with typical features such as intestinal mucosal layer on both sides of the lesion, separated by a submucosal layer containing 2 muscularis mucosae (Fig. 2).

Duodenal duplication cysts are a rare congenital abnormality that can cause digestive symptoms. It has an estimated prevalence of less than 1 per 100,000 persons.[3] Usually diagnosed during childhood, it can remain asymptomatic until adulthood. The clinical manifestations of a DDC are numerous, including abdominal pain, nausea and vomiting, pancreatitis, cholestasis or hepatitis, and GI bleeding. In adults, the most frequent presentation is acute pancreatitis.[4]

The diagnosis of duodenal duplication cysts can be challenging. The main differential diagnosis of a DDC is the type III choledochal cyst (choledochocele), according to the classification by Todani et al.[5] The DDCs represent 5% of all GI duplications and have a cystic structure bulging into the duodenal lumen and developed distally from the ampulla of Vater. A cyst–pancreaticobiliary duct connection is identified in almost 29% of cases.[4] Therefore, the cyst can be filled with duodenal secretions, biliary secretions, or both. By contrast, choledochoceles have a cystic structure originating from the main bile duct and protruding proximally from the papilla. The principal distinguishing feature between these 2 entities is their histologic characteristic: DDCs are covered both inside and out with duodenal mucosa containing a distinct layer of smooth muscle.[6]

Both duodenal duplication cysts and choledochoceles are associated with an evolution to malignancy,[1,2] justifying their resection. Endoscopic resection is minimally invasive and has been proposed as an alternative in selected patients. Antaki et al[7] reported a series of 7 successful endoscopic resections for benign symptomatic DDCs. The basic principle in all cases was a marsupialization of the cyst to allow free drainage into the duodenal lumen. There were no major events and no local recurrence after 7 months of follow-up. Several case reports have also demonstrated the success of this innovative approach, indicating that endoscopic resection is a safe and effective technique in selected cases.[8,9]

DISCLOSURE

All authors disclosed no financial relationships relevant to this publication.

Abbreviation: DDC, duodenal duplication cyst.

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