Ampullary cyst with papillary orifice distal to bulge: Not always a choledochocele!

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A 9-year-old girl presented with recurrent episodes of epigastric pain for the past 3 months. Investigations suggested recurrent acute pancreatitis with significantly elevated amylase and lipase levels on several occasions. Contrast-enhanced CT of the abdomen revealed a 3.1- × 1.8- × 3.9-cm thick-walled nonenhancing cystic lesion along the third part of the duodenum (Fig. 1). MRCP revealed a cyst communicating with the common biliopancreatic channel (Fig. 2). A side-viewing examination using a duodenoscope (Olympus TJF Q180V, Olympus Corp, Tokyo, Japan) revealed a smooth extrinsic bulge extending from the second to the third part of the duodenum on the medial wall. The papillary orifice could not be located. An EUS examination using a linear echoendoscope (GF-UCT180, Olympus Corp) revealed a duodenal cyst with layered architecture on the medial duodenal wall that was communicating with the common pancreatobiliary channel. The presumed etiology of pancreatitis in this case is papillary orifice obstruction.

She underwent endoscopic deroofing using a duodenoscope. A linear incision was made using a needle knife (Micro knife, Boston Scientific, Mass, Natick, USA) on the prominent area of the duodenal bulge, from which a free flow of bile was observed. The incision promptly decompressed the projecting duodenal cyst (Video 1, available online at www.giejournal.org), and the papillary orifice emerged on the distal part of the bulge. Minor bleeding was noted during the procedure, which was controlled using electrocoagulation. Histology results from the intracystic biopsy confirmed duodenal mucosal epithelium with a common muscular layer (Fig. 3), suggesting a duodenal duplication cyst (DDC) rather than a choledochocele. The latter can be lined by either bile duct or gallbladder mucosa and, unlike DDC, lacks

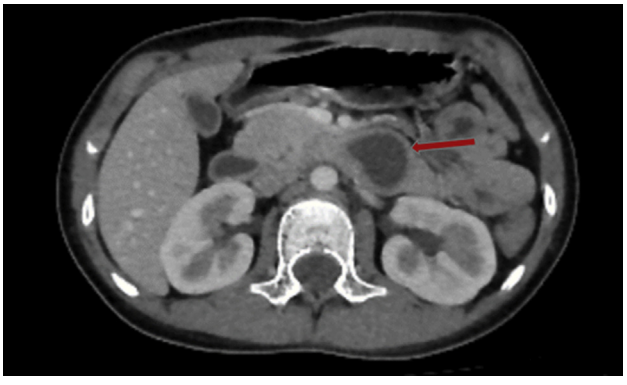

Figure 1. Image showing thick-walled nonenhancing cystic lesion (*arrow*) along the third part of the duodenum.

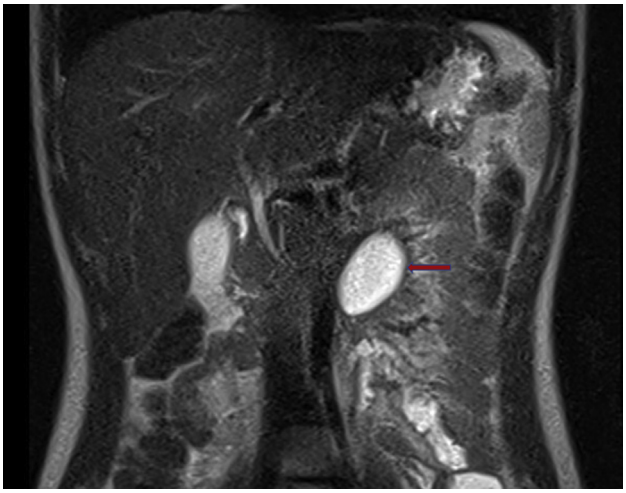

Figure 2. MRCP coronal image showing cystic lesion (*arrow*) obstructing the lumen.

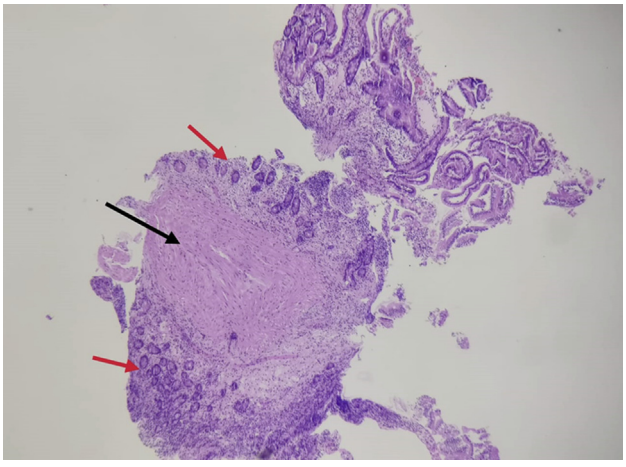

Figure 3. Biopsy specimen showing duodenal mucosal epithelium (*red arrow*) with common muscular layer (*black arrow*).

a smooth muscle layer. The patient remained pain-free in the next 6 months of follow-up.

Although DDCs are generally benign, malignant transformations have been reported. A meta-analysis of 41 cases of duodenal duplication cysts by Chen et al[1] showed that surgical intervention is the most common measure used for treatment. Surgical excision provides complete cure and eliminates the risk of malignancy. However, because of the proximity to the biliopancreatic duct, total resection requires pancreaticoduodenectomy, with its associated morbidity, mortality, and poor quality of life.

Recently, duodenal duplication cysts have been safely treated endoscopically, with significantly lower associated morbidity compared with surgery.[2] It has been postulated that avoiding stasis of biliopancreatic secretions within the cyst, as achieved with endoscopy, provides protection against malignant change.[3]

Patients undergoing endoscopic deroofing of a duplication cyst should be regularly followed. In our case, these aspects were discussed with the patient's parents, and they opted for endoscopic management.

Conventionally, it is accepted that the orifice is located distally in the duodenal papillary cystic mound in choledochocele and proximally in a duplication cyst.[4,5] However, this dictum is not always correct, as observed in our index case. Either way, deroofing the cyst relieved the symptoms of recurrent abdominal pain due to pancreatitis.

DISCLOSURE

All authors disclosed no financial relationships.

Abbreviation: DDC, duodenal duplication cyst.

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