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A case series of symptomatic intraluminal duodenal duplication cysts: presentation, endoscopic therapy, and long-term outcome (with video) CME

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Background: Duodenal duplication cysts are rare congenital anomalies. Symptomatic cases have classically been treated by surgical resection, which can be complex because of the close proximity of the cysts to the papilla.

Objective: To describe a series of 8 patients with symptomatic duodenal duplication cysts who were treated endoscopically, with a special focus on the long-term outcome.

Design: Retrospective case series.

Setting: Three tertiary-care European academic hospitals.

Patients: Eight patients, age 8 to 72 years, were treated endoscopically for symptomatic intraluminal duodenal duplication cysts between 1981 and 2006. Seven patients presented with acute pancreatitis, and one patient presented with jaundice.

Intervention: Endoscopic incision and marsupialization of the cysts was performed by using a variety of endoscopic tools (needle-knife and regular sphincterotomes, cystotomes, and polypectomy snares).

Main Outcome Measurements: Technical success of endoscopic intervention and long-term clinical recurrence of symptoms.

Results: No major complications occurred. All patients remained asymptomatic at a median follow-up of 7.3 years.

Limitations: Retrospective study; the small number of patients.

Conclusions: The endoscopic treatment of symptomatic intraluminal duodenal duplication cysts is a safe and effective technique, with excellent long-term results. It represents a minimally invasive alternative to surgical resection and might be considered the preferred therapeutic modality for these cases.

GI duplications are uncommon congenital anomalies that can occur at any level of the GI tract. Their estimated prevalence is 1:4500 to 1:10,000 in the general population.[1] Most symptomatic cases are diagnosed in children and usually present with obstructive or bleeding symptoms. Duodenal duplication cysts are extremely rare, representing less than 5% of all GI duplications. Acute pancreatitis or biliary obstruction has rarely been attributed to these cysts.[2-12] Treatment has classically involved surgical resection,[13-16] which can be complex because of the close proximity of the cysts to the papilla and the biliopancreatic confluence. Endoscopic therapy has been used as an alternative to surgery in a few selected cases.[17-22] We describe the endoscopic technique used in the treatment of intraluminal duodenal duplication cysts and the long-term results of this minimally invasive therapy.

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PATIENTS AND METHODS

Eight patients were treated endoscopically for symptomatic duodenal duplication cysts in 3 tertiary-care European university hospitals, Gemelli Hospital (case nos. 1 and 2), St-Luc Hospital (case nos. 3 and 4), and Erasme hospital (case nos. 5-8), between 1981 and 2006 (Table 1).

TABLE 1. Patient demographics

Case no.	Sex	Age at symptom onset, y	Clinical presentation	Initial workup	Year of presentation
1	M	25	AP	US, CT	1997
2	M	72	Jaundice	MRCP	2002
3	F	21	RAP	US, CT, EUS, ERCP, MRCP	2004
4	M	44	AP, chronic GI blood loss	US, EUS, ERCP, MRCP	2005
5	M	7	RAP	UGIS, US	1981
6	M	43	AP	CT, ERCP	1993
7	F	17	RAP	MRCP	1996
8	F	26	RAP	EUS, MRCP	2006

AP, Acute pancreatitis (1-2 episodes); *RAP,* relapsing acute pancreatitis (≥3 episodes); UGIS: Upper-GI x-ray study.

The onset of symptoms was at a median age of 25.5 years (range 7-72 years). Five patients (62.5%) were men. Four patients (50%) presented with acute relapsing pancreatitis (≥3 episodes). The remaining patients presented with one or two episodes of acute pancreatitis (n = 3) and jaundice (n = 1). The presentation was associated with chronic GI blood loss in one case. A combination of several different tests was used in the diagnostic workup. A fluid-filled cystic structure in the paraduodenal area was noted in all cases, and, in several patients, the diagnosis of duodenal duplication was suspected before endoscopy. The initial differential diagnosis included choledochocele (congenital choledochal cyst type III[23]), pancreatic pseudocyst, and ampulloma. During endoscopy, duodenal duplication was diagnosed when a fluid-filled structure that protruded inside the duodenal lumen was seen (Fig. 1A), along with normal cholangiograms and pancreatograms (Fig. 1B). In the most recent cases, a diagnosis was achieved by EUS and/or MRCP, without the need for a diagnostic ERCP (Fig. 2A). In the 3 cases in which EUS was performed, an intramural anechoic lesion, separate from the pancreatic and biliary ductal systems, was noted. The normal-looking papilla was always found on the proximal side of the protrusion, whereas, it is usually found on the distal side of the protrusion in choledochoceles (Fig. 3). In the 7 cases in which cholangiograms were obtained, emptying of contrast from the bile ducts into the cyst lumen was noted in many but not all cases (4 cases), which caused enlargement of the cyst and explained the obstructive phenomenon leading to acute pancreatitis. Treatment was performed at a median of 24 months (range 2-180 months) after the onset of symptoms.

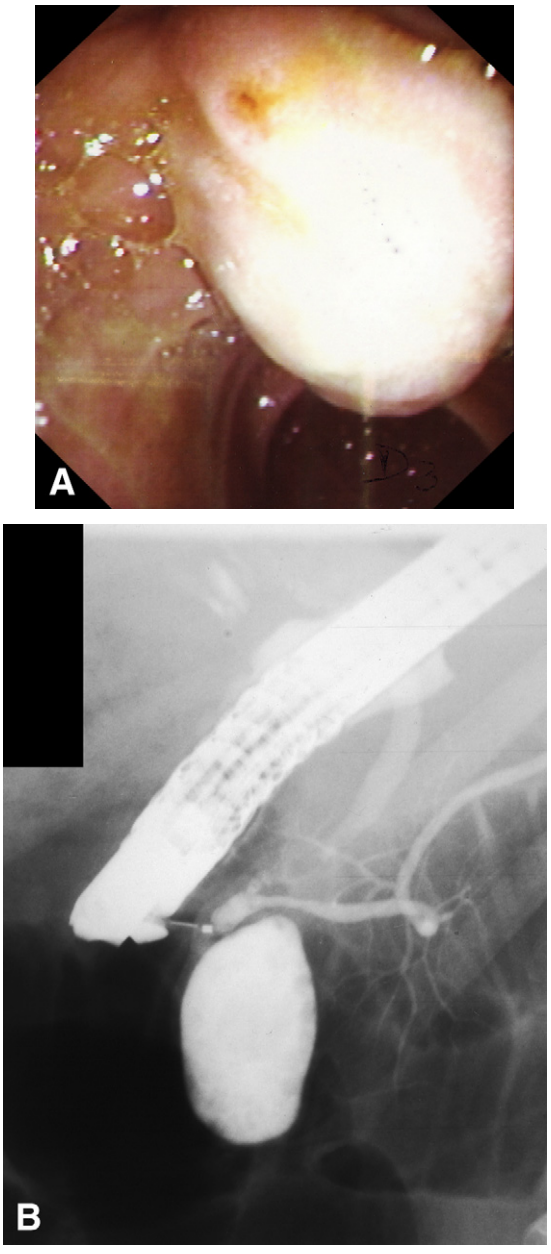

Figure 1. Duodenal duplication cyst (case no. 6). **A,** Typical endoscopic appearance, with the normal papilla proximal to the cyst. **B,** Radiographic appearance of the same duodenal duplication cyst during ERCP, with a normal cholangiogram and pancreatogram. Note the presence of stones inside the duplication cyst.

Techniques

All therapeutic procedures were performed with standard, adult, side-viewing duodenoscopes. Procedures were performed with the patient under general anesthesia in 6 cases and sedation in 2 cases. The basic principle in all cases was marsupialization of the cyst to allow for free drainage into the duodenal lumen (Table 2). The initial puncture through the cyst roof was performed by using a needle-knife sphincterotome (n = 5) or a cystotome (n = 1). In 4 cases, the endoscopist used a different instrument (regular sphincterotome [n = 2], an insulated-tip knife [n = 1],

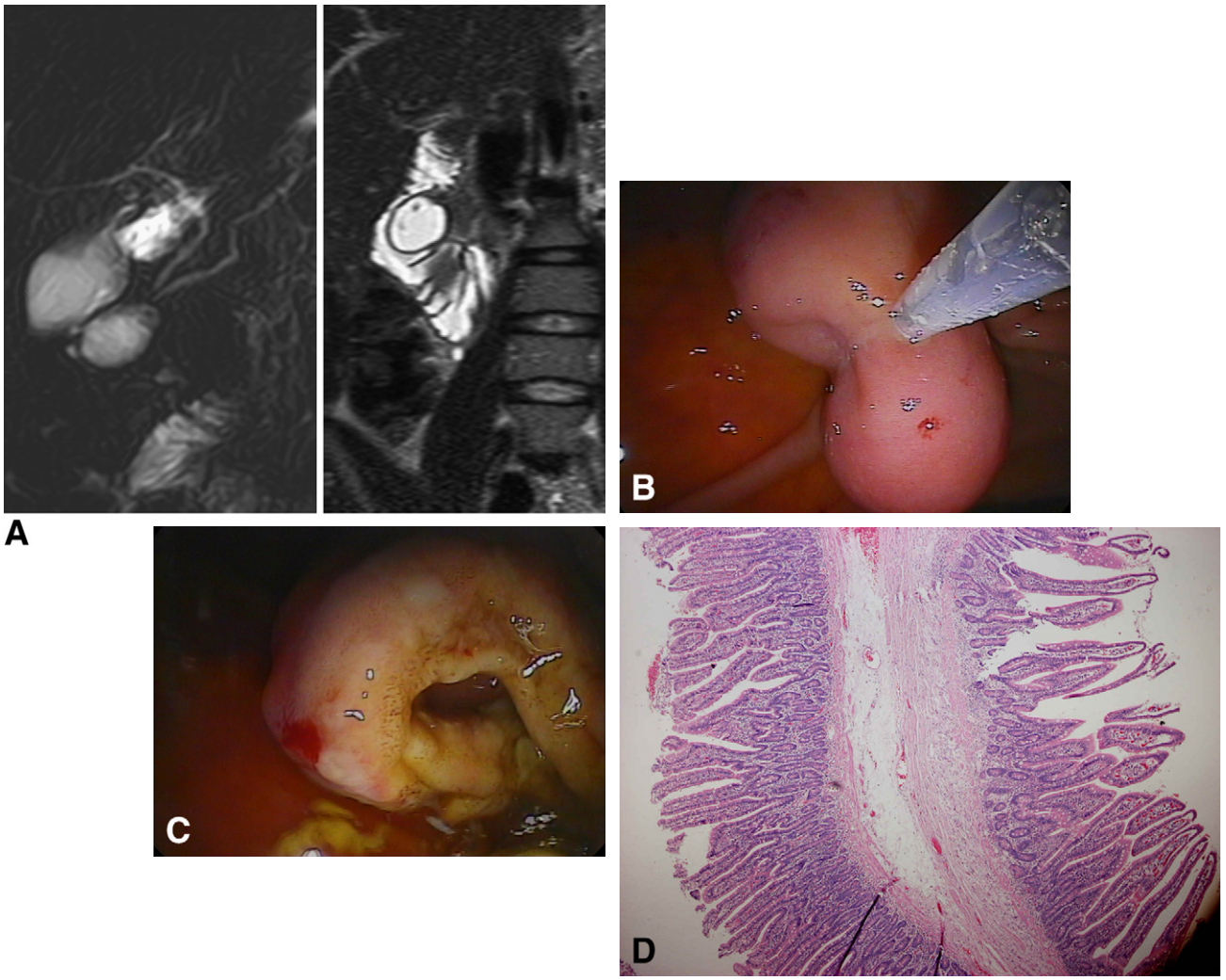

Figure 2. The diagnosis and endoscopic therapy of a symptomatic duodenal duplication cyst in a 26-year-old patient (case no. 8). **A,** MRCP, showing the duodenal duplication cyst, with normal biliary and pancreatic ductal systems. **B,** Positioning of the polypectomy snare around the duplication cyst. **C,** Endoscopic appearance of the duplication cyst immediately after treatment; note the wall layers of the duplication. **D,** Histologic section of the snare resection specimen, showing the characteristic features of a duplication cyst: two layers of duodenal mucosa (with their respective muscularis mucosae) separated by a layer of submucosa (H&E, orig. mag. ×40)

or an inverted sphincterotome [n = 1]) to enlarge the incision through the cyst roof. Subsequent partial resection of the cyst wall was performed by using a polypectomy snare (n = 2). In the remaining two cases, a partial resection of the cyst wall was performed with a snare, without an initial incision (Fig. 2B and C; Video 1, available online at www.giejournal.org). When present, gallstones were evacuated from inside the cyst lumen by using a balloon-tipped catheter (n = 1). Biliary and pancreatic stents were temporarily placed in one case. Sphincterotomy was performed in 3 cases.

RESULTS

Technical success was achieved in all patients during a single endoscopic procedure, with no major complications. Minor arterial bleeding occurred during incision in one case and was immediately controlled endoscopically with bipolar coagulation and application of two clips. Blood transfusions were not required. There were no cases of delayed bleeding, perforation, or pancreatitis.

Histopathology

Biopsy samples were obtained from inside the cysts in all cases. Results of histopathologic analysis showed benign duodenal mucosa, which confirmed the diagnosis. The characteristic features of a duplication cyst: two duodenal mucosal layers (with their respective muscularis mucosae), separated by a layer of submucosa (Fig. 2D), were observed in the cases in which the snare resection specimen was retrieved.

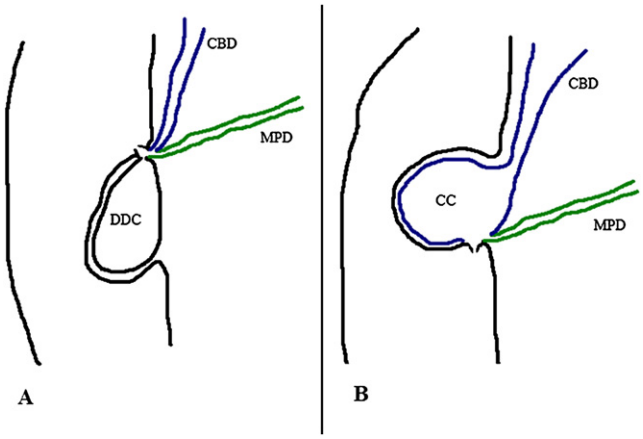

Figure 3. A and **B,** Diagram showing the difference between a duodenal duplication cyst (**A**) and a choledochocele (**B**) with regard to their relationship with the papilla. *CBD,* common bile duct; *MPD,* main pancreatic duct; *DDC,* duodenal duplication cyst, *CC,* choledochocele.

TABLE 2. Endoscopic intervention

Case no.	Age at treatment, y	Anesthesia	Initial puncture	Other instruments	Resection	Sphincterotomy	Complications
1	40	General	NK	Inverted sphincterotome		EBS	None
2	72	Sedation	NK	IT knife		EBS	None
3	24	General	Cystotome	Sphincterotome			None
4	47	General	NK			EBS and EPS*	Minor bleeding
5	8	General	NK	Sphincterotome	Snare		None
6	43	Sedation	NK		Snare		None
7	18	General			Snare		None
8	31	General			Snare		None

NK, Needle-knife sphincterotome; IT, insulated-tip; *EBS,* endoscopic biliary sphincterotomy; *EPS,* endoscopic pancreatic sphincterotomy.
*With placement of biliary and pancreatic stents.

Follow-up and long-term outcome

Follow-up was defined as the interval between the therapeutic endoscopic intervention and the last contact with the patients by clinic visits or telephone conversations. The median follow-up was 7.3 years (range 5 months to 13 years). All patients remained asymptomatic, with no further episodes of pancreatitis, cholestasis, or bleeding (Table 3). Subsequent investigation with MRCP (n = 4) and/or endoscopy (n = 6) was performed at a median of 7 months after treatment (range 2 months to 12 years) and revealed the atrophic cyst remnant, with no additional abnormalities (Fig. 4). All biopsy specimens obtained at a follow-up endoscopy showed normal duodenal mucosa.

DISCUSSION

Intraluminal duodenal duplication cysts are rare congenital malformations[1] that usually present in childhood with obstructive or bleeding symptoms.[2-12] The majority of our patients presented with episodes of acute pancreatitis. We suspect that the pathophysiology is related to the occlusion of the pancreatic ductal system by the distended duplication cyst, filled with secretions, sludge, or stones.

With the increased availability of MRCP and EUS, a diagnosis of duodenal duplication can be reached nowadays, without the need for diagnostic ERCP. Either one of these tests can be used, depending on local expertise, to examine the suspected lesion and to exclude anomalies of the biliary and pancreatic ductal systems, which, in combination with the typical endoscopic appearance and location of the lesion distal to a normal-looking papilla, will confirm the diagnosis of a duodenal duplication cyst.

Therapy has classically involved surgical resection.[13-16] However, the close proximity of the major papilla and the associated risk of surgical complications stimulated an interest in treating these cases endoscopically. A few individual case reports of duodenal duplication cysts,

TABLE 3. Treatment outcome and follow-up

Case no.	Subsequent testing	Recurrent symptoms	Length of follow-up, mo
1	MRCP and ERCP	None	120
2	ERCP	None	55
3	EGD and MRCP	None	26
4	MRCP	None	12
5	ERCP	None	152
6	US and ERCP	None	160
7	MRCP	None	125
8	EGD	None	5

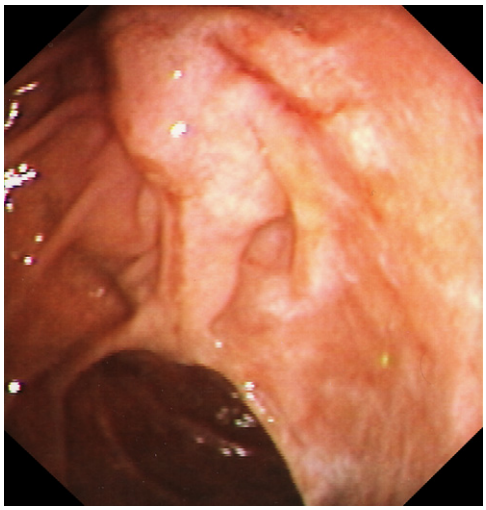

Figure 4. Endoscopic appearance of the atrophic cyst remnant 13 years after treatment of a duodenal duplication cyst (case no. 5).

treated by using a variety of endoscopic techniques, have been previously published.[17-22] Our article represents the largest case series ever described for such a rare entity. The same standard basic principles of incision and marsupialization, using available endoscopic tools, were applied in all cases. The only complication was a case of minor bleeding at the time of endoscopy.

Our experience leads us to believe that resection of the cyst roof by using a standard polypectomy snare or a large incision of the roof of the duplication is probably sufficient to cure the patient. Snare resection provides a better specimen than biopsy specimens for histologic confirmation of the diagnosis. Additional sphincterotomy is not necessary, because the sphincter area is intact in cases of duodenal duplication. A sphincterotomy was performed in 3 of our cases, with no additional benefit to unroofing the duplication and with a potential risk for complications. This contrasts with the endoscopic therapy of choledochoceles for which a wide biliary sphincterotomy is the treatment of choice.[24]

All our patients remained asymptomatic during the entire follow-up period. This confirms that providing drainage and avoiding compression of the biliary and pancreatic ductal systems is sufficient to prevent further symptoms. Therefore, endoscopic therapy can be considered a definitive treatment for these patients.

There are 3 reported cases of malignancy arising inside a duodenal duplication cyst.[25-27] Compared with surgical resection, endoscopic therapy does not always result in complete ablation of the cyst mucosa; however, avoiding stasis of secretions inside the cyst over a lifetime could hypothetically have a protective effect. Some investigators have also attributed malignant degeneration to the presence of ectopic gastric tissue inside the cyst; this was not found in any of our patients. We usually recommend a repeat endoscopy with follow-up biopsies 6 to 12 months after treatment.

In conclusion, endoscopic treatment of symptomatic intraluminal duodenal duplication cysts is a safe and effective technique. The long-term outcome is excellent, with complete resolution of symptoms. Therefore, endoscopic therapy represents a minimally invasive alternative to surgical resection and might be considered the preferred therapeutic modality for these cases.

DISCLOSURE

The authors report that there are no disclosures relevant to this publication.

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