

Case Report

Duodenal duplication cyst: A rare cause of geriatric gastrointestinal bleeding

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Abstract

Duodenal duplication cysts are rare congenital anomalies usually found in children, but rare cases, often presenting with pancreatitis, have been reported in adults. We report the case of a duodenal duplication cyst in an 81-year-old man who presented with gastrointestinal bleeding. Despite endoscopy and endoscopic ultrasonography, duodenal duplication was not diagnosed until exploratory laparotomy was performed to remove the lesion. To the best of our knowledge, this was the first case of a duodenal duplication cyst that presented with gastrointestinal bleeding in an elderly adult. The patient's recovery was uneventful. A high index of suspicion is required to diagnose duodenal duplication cysts, particularly in elderly patients with an atypical presentation.

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1. Introduction

With an estimated prevalence of 1:4500 to 1:10,000 in the general population, intestinal duplications are infrequently encountered congenital anomalies.¹ Most duplications are detected when children present with gastrointestinal (GI) bleeding or with symptoms related to obstruction, and less than 30% of all cases of intestinal duplications are diagnosed in adults.² Intestinal duplications in the duodenum are extremely rare and comprise less than 5% of all intestinal duplications. Their presenting symptoms are nonspecific, and are closely related to the type, size, and location of the lesions. Because of the rarity of the condition, physicians seldom suspect this diagnosis. We describe a rare case of duodenal duplication with heterotopic gastric lining in an elderly male, manifesting as GI bleeding. The diagnosis was confirmed by the operative findings and subsequent histopathologic examinations.

2. Case report

An 81-year-old male with mild, well-controlled hypertension sought medical consultation because of a 6-year history of dizziness and three episodes of syncope over the preceding 2 months. An evaluation revealed normocytic anemia, with a hemoglobin level of 7.2 g/dL. All other laboratory examinations yielded normal results. The patient's history showed that he had had intermittent tarry stool passages since he was a child but had ignored them. These symptoms, however, had exacerbated and increased in frequency and severity over the preceding 2 months. The patient denied having any other GI or systemic symptoms or signs, such as weight loss. Although he had mild anemic conjunctiva, physical examination revealed nothing significant.

Pandoscopy revealed that the second portion of the duodenum was normal, and a protruding ovoid mass, 2 cm in diameter, was located within the posterior wall of the first portion of the duodenum (Fig. 1A). Endoscopic ultrasonography (EUS) revealed a well-defined, hypoechoic structure with mixed contents embedded within the muscularis layer (Fig. 1B). Preoperative abdominal sonography revealed no abnormal findings for the liver, gallbladder, spleen, kidney, or regional lymph nodes.

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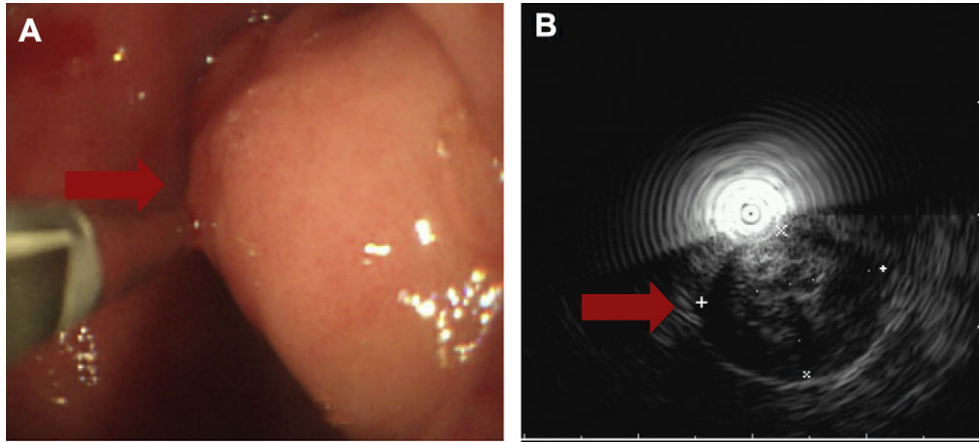


Fig. 1. (A) Pandoscopic examination disclosed a polypoid mass protruding into the lumen of the first portion of the duodenum (arrow). (B) Endoscopic ultrasonography shows a well-defined mixed-echoic mass 1.5 cm in diameter (arrow).

At laparotomy, an old, healed scar with wall thickening was noted in the anterior wall of the pylorus. A longitudinal incision was made in the scar, and a frozen biopsy was performed. Analysis of the frozen biopsy specimen showed that it was a benign ulcer with underlying fibrotic submucosa and muscularis. Further investigation disclosed a cyst structure 2.5 cm in length and 1.5 cm in diameter located at the posterior wall of the first portion of the duodenum (Fig. 2). In manipulation, it was blind-ended and contained all layers of the bowel, and no further communication with the adjacent pancreas or common bile duct was identified. The protruding structure and the cyst on the duodenum were excised completely, leaving the duodenal muscular wall exposed. After this, we primarily closed the lining defect with a 4–0 Vicryl one-layer simple suture. An additional pyloroplasty was performed on the deformed pyloric incision.

The patient's postsurgical recovery was smooth, and he was discharged on the seventh postoperative day. Pathologic examination showed whole-layer alimentary tissue containing the muscular layer and gastric lining (Fig. 3). The cysts had two mucosal layers, with their respective muscularis mucosae, separated by a layer of submucosa. No evidence of further symptoms or GI bleeding was found 6 months after the operation.

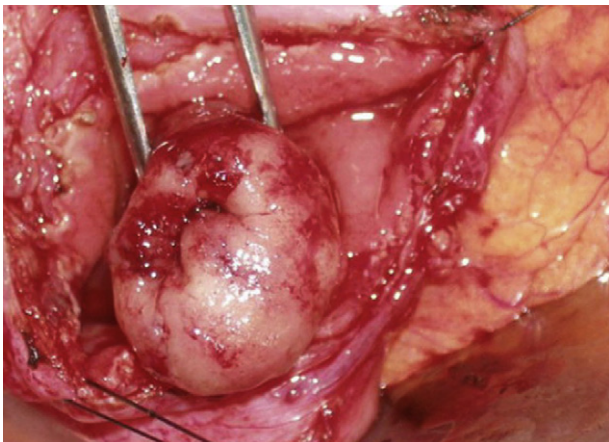


Fig. 2. Intraoperative photograph shows a cyst structure near the pylorus.

3. Discussion

Intestinal duplications are rare congenital anomalies that form during the embryonic development of the human digestive organs. The definition includes a smooth-muscle coat, an alimentary epithelial lining and an intimate attachment to the GI tract.³ They occur at any level of the alimentary tract, and their sizes and shapes vary greatly. Jejunal duplications are most common, followed by gastric and colonic ones, whereas duodenal ones are the rarest, constituting less than 5% of all intestinal duplications.

Like other intestinal duplications, most duodenal duplications are diagnosed in childhood, and such a lesion detected late in an adult is extremely rare. They have diverse clinical presentations, depending on the conditions they give. In cases with distension or inflammation, acute abdomen may manifest. In cases compressing the peripheral organs, such as the adjacent bowels or pancreatobiliary tree, intestinal obstruction and jaundice may occur.⁴ In a few cases, ectopic gastric mucosa can appear and

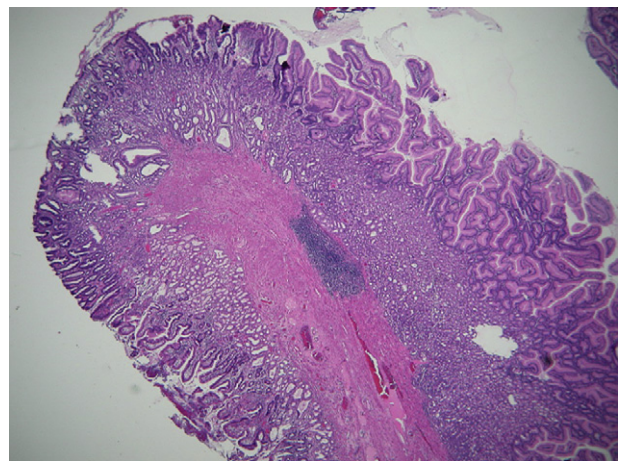


Fig. 3. Histologic section of the surgical specimen shows the characteristic features of a duplication cyst: transmurality of the lesion with gastric and duodenal linings and their respective muscularis mucosae (hematoxylin & eosin, 20 \times).

cause ulceration, bleeding, and even perforation.^{5,6} Complicated bleeding is mostly found in infants, probably because of their low blood volume. In the present case, probably owing to the physical reserve of the patient and the small size of the lesion, occult bleeding may have existed throughout his life with no overt clinical symptoms, until late in life, when exacerbation of the ulcer was discovered by dizziness and syncope.

The preoperative diagnosis of intestinal duplications is rarely accurate. A diagnosis is made when on ultrasonography, a mass is found with an inner hyperechoic rim corresponding to the mucosa and submucosa surrounded by a hypoechoic layer representing the muscularis propria.^{7,8} Barium studies of GI series help to demonstrate the mass effect and displacement of normal alignment,⁸ whereas a technetium scan can aid in the detection of heterotopic gastric mucosa in cases complicated with bleeding.⁹ Recently, a multiplanar approach using computed tomography and magnetic resonance imaging has facilitated the evaluation of the nature and involvement of the lesion in more complicated cases.⁸ In this case, the preoperative endoscopy and EUS revealed a submucosal tumor, which was probably due to its small opening and the filled secretion that masked the cystic structure. Because there were no signs of malignancy, such as tumor size exceeding 30 mm or irregular margins on EUS,¹⁰ the first impression was that of benign or limited disease. Smooth muscle neoplasm was favored regarding its muscularis origin, although heterotopic pancreas, lymphoma, carcinoid, granular cell tumor, cyst, and lipoma can also occur in rare situations.¹¹ The presence of intermediate to hypoechoic echogenicity on EUS further excluded the diagnosis of anechoic cyst, as well as hyperechoic granular cell tumor and lipoma. In addition, heterotopic pancreas, lymphoma, and carcinoids infrequently show regular margins on EUS. Thus, a diagnosis of myogenic tumor was favored in this case.

For any intestinal duplication, definite diagnosis necessitates histologic analyses. EUS-assisted needle biopsy offers a less invasive technique for histologic assessment, but the diagnostic yields for submucosal lesions vary greatly in every series. Although the recent biopsy technique as well as cytomorphologic and immunochemical evaluations have greatly enhanced diagnostic accuracy for smooth-muscle neoplasm, the diagnostic value for other submucosal lesions and sensitivity for malignancy detection remain controversial.¹⁰ A preoperative needle biopsy was not performed in this case. Computed tomography was not arranged because the lesion presented as a small, well-defined submucosal tumor, and the preoperative abdominal sonography did not exhibit any significant lesions within the liver, which favored local disease, enabling curative resection by surgery. Some authors suggest the need for preoperative or intraoperative cholangiopancreatography to exclude biliopancreatic communication with the duodenal duplication cyst.¹² In this case, we did not arrange such a study because we did not think of the diagnosis preoperatively, and the gross findings and hand palpation did not identify any communication with the papilla of Vater or common bile duct during surgery.

Surgical intervention for symptomatic or complicated duplications is always justified. However, the indications for those who are asymptomatic remain controversial. Not treating them leaves

the possibility of the development of neoplastic changes of the lining and other complications.^{12,13} In this regard, surgical resection should be the treatment of intestinal duplications whenever possible. Segmental resection is preferred because of a common blood supply with the normal alimentary tract. Recently, advances in therapeutic endoscopy, such as endoscopic mucosal resection and endoscopic submucosal dissection, provides a viable option for resecting neoplastic lesions of the alimentary tract with a low invasive potential.¹⁴ They have also been widely introduced to submucosal lesions, and occasionally succeed in the removal of duplication cysts in older children and adults.^{12,15} There are, however, certain limitations. Typical exclusion criteria include a size exceeding 3 cm, the presence of ulceration mucosa, extension into the muscular propia, and the presence of signs indicating malignancy.¹⁰ In the present case, we treated the patient with excision of the protruding cyst and its whole duodenal lining by open surgery, with the remaining lining primarily closed with 4–0 Vicryl to cover the exposed posterior duodenal muscular layer. The approach was the same as that used for endoscopic resections in small gastric/duodenal submucosal lesions. Our treatment avoided conventional pancreaticoduodenectomy, which is often used for such lesions.

Duplications in the duodenum are extremely rare, and intestinal duplications of any location are rarely diagnosed in adults. We have reported an unusual occurrence of duodenal duplication in an elderly patient with the uncommon clinical manifestation of GI bleeding. As in previously reported cases, the preoperative diagnosis of this patient was challenging. Surgical resection was justified because any GI mass complicating bleeding in an elderly patient should be considered malignant until proven otherwise. Given the recent success of endoscopic procedures in excising duplication cysts in older children and adults, a high index of suspicion facilitating preoperative diagnosis of the condition may help to avoid surgery in some cases.

References

1. Leffall Jr LS, Jackson M, Press H, Syphax B. Duplication cyst of the duodenum. *Arch Surg* 1967;**94**:30–4.
2. Browning R. Duodenal duplications. *Rev Surg* 1963;**20**:226–9.
3. Gross RE, Holcomb Jr GW, Farber S. Duplications of the alimentary tract. *Pediatrics* 1952;**9**:448–68.
4. Mosca F, Stracqualursi A, Persi A, Latteri S. Duodenal duplication: report of 2 cases in adults and review of the literature. *Chir Ital* 2001;**53**:883–91.
5. Merrot T, Anastasescu R, Pankevych T, Tercier S, Garcia S, Alessandrini P, et al. Duodenal duplications: clinical characteristics, embryological hypotheses, histological findings, treatment. *Eur J Pediatr Surg* 2006;**16**:18–23.
6. Arbell D, Lebenthal A, Blashar A, Shmushkevich A, Gross E. Duplication cyst of the duodenum as an unusual cause of massive gastrointestinal bleeding in an infant. *J Pediatr Surg* 2002;**37**:E8.
7. Barr LL, Hayden Jr CK, Stansberry SD, Swischuk LE. Enteric duplication cysts in children: are their ultrasonographic wall characteristics diagnostic? *Pediatr Radiol* 1990;**20**:326–8.
8. Hur J, Yoon CS, Kim MJ, Kim OH. Imaging features of gastrointestinal tract duplications in infants and children: from oesophagus to rectum. *Pediatr Radiol* 2007;**37**:691–9.
9. Brown RL, Azizkhan RG. Gastrointestinal bleeding in infants and children: Meckel's diverticulum and intestinal duplication. *Semin Pediatr Surg* 1999;**8**:202–9.

10. Polkowski M, Butruk E. Submucosal lesions. *Gastrointest Endosc Clin N Am* 2005;**15**:33–54.
11. Okanobu H, Hata J, Haruma K, Mitsuoka Y, Kunihiro K, Manabe N, et al. A classification system of echogenicity for gastrointestinal neoplasms. *Digestion* 2005;**72**:8–12.
12. Antaki F, Tringali A, Deprez P, Kwan V, Costamagna G, Le Moine O, et al. A case series of symptomatic intraluminal duodenal duplication cysts: presentation, endoscopic therapy, and long-term outcome (with video). *Gastrointest Endosc* 2008;**67**:163–8.
13. Hata H, Hiraoka N, Ojima H, Shimada K, Kosuge T, Shimoda T. Carcinoid tumor arising in a duplication cyst of the duodenum. *Pathol Int* 2006;**56**:272–8.
14. Conlin A, Kaltenbach T, Kusano C, Matsuda T, Oda I, Gotoda T. Endoscopic resection of gastrointestinal lesions: advancement in the application of endoscopic submucosal dissection. *J Gastroenterol Hepatol* 2010;**25**:1348–57.
15. Tekin F, Ozutemiz O, Ersoz G, Tekesin O. A new endoscopic treatment method for a symptomatic duodenal duplication cyst. *Endoscopy* 2009;**41**(Suppl 2):E32–3.