








Imaging Findings of an Intraluminal Duodenal Diverticulum Associated with Adult Duodeno-Duodenal Intussusception and Recurrent Pancreatitis: A Case Report

성인 십이지장-십이지장 장중첩증 및 재발성 췌장염과 관련된 관강내 십이지장 계실의 영상 소견: 증례 보고

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Intraluminal duodenal diverticulum (IDD) is a rare congenital abnormality, consisting of a sac-like mucosal lesion in the duodenum. Cases of IDD can present with gastrointestinal bleeding, duodenal obstruction, or pancreatitis. Here, we report a rare case of a 25-year-old female presenting with IDD complicated by duodeno-duodenal intussusception and recurrent pancreatitis. The diagnosis was based on findings from radiologic examinations (CT and MRI), upper gastrointestinal series (barium swallow), and gastroduodenofiberscopy. Laparoscopic excision of the presumed duodenal duplication was performed. The subsequent histopathologic evaluation of the excised sac revealed normal mucosa on both sides, but the absence of a proper muscle layer confirmed the diagnosis of IDD. Radiologic detection of a saccular structure in the second portion of the duodenum can indicate IDD with duodeno-duodenal intussusception as the lead point.

Index terms Diverticulum; Intussusception; Acute Pancreatitis; Endosonography

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INTRODUCTION

Intraluminal duodenal diverticulum (IDD) is a rare congenital abnormality, consisting of a sac-like mucosal lesion in the duodenum that lies close to the ampulla of Vater (1). While IDD can remain asymptomatic, gastrointestinal symptoms typically occur in the 3rd decade of life. They are usually associated with complications, such as bleeding, duodenal obstruction, or pancreatitis. Acute pancreatitis associated with IDD is present in less than 20% of reported cases (2).

Since the duodenum has a fixed position in the retroperitoneum, duodeno-duodenal intussusception is an extremely rare cause of duodenal obstruction. Indeed, most of the intestinal intussusception in IDD cases represent complications secondary to intestinal masses (3).

Here, we describe a rare case of IDD presenting with duodeno-duodenal intussusception and recurrent pancreatitis.

CASE REPORT

A 25-year-old female was admitted to the emergency department of our hospital with a complaint of epigastric pain. Laboratory test results showed elevated levels of serum amylase (2076 IU/L; normal range: 30–110 IU/L) and lipase (> 3000 IU/L; normal range: 0–160 IU/L).

The patient had experienced acute pancreatitis a month ago, although she had no history of alcohol use. In the past, she had abdominal pain and visited a hospital 10 years ago. However, there was no evidence of pancreatitis, although a duodenal diverticulum was detected on endoscopy.

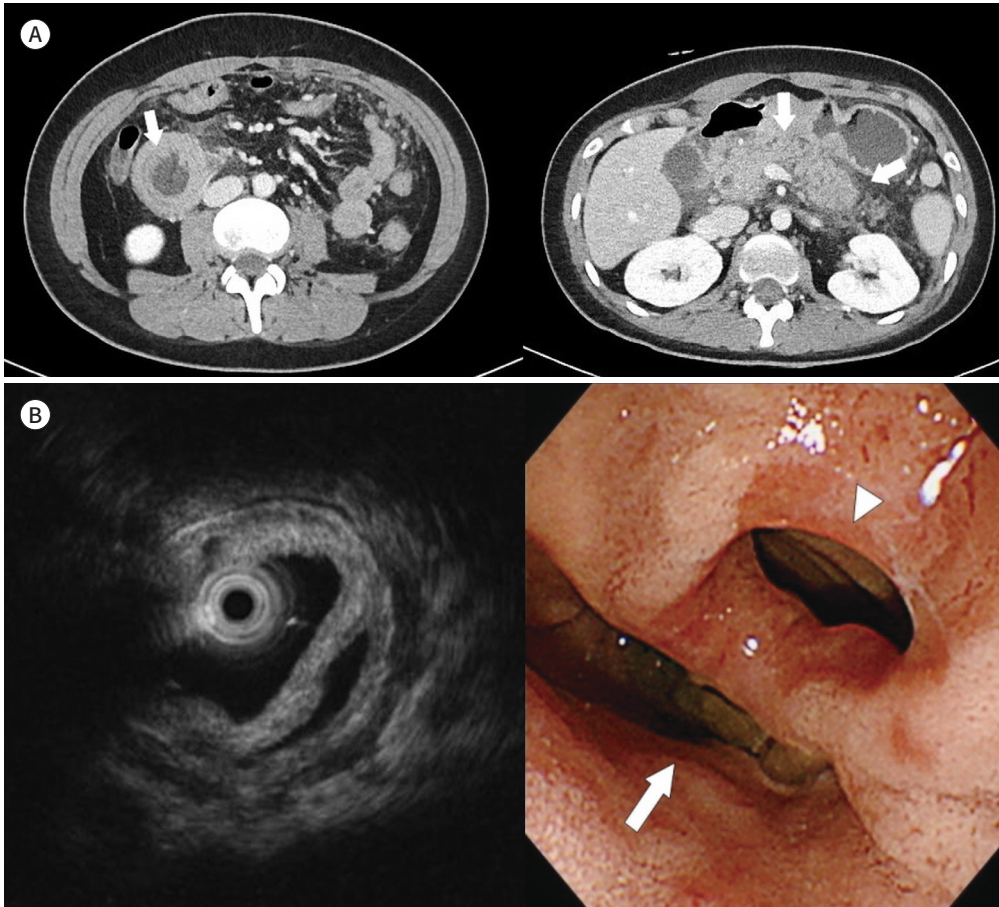
Abdominal-pelvic CT showed characteristics of duodeno-duodenal intussusception (“target sign”) in the second portion of the duodenum, but no lead point was identified. Characteristic findings of acute pancreatitis (i.e., edematous pancreas with peripancreatic fat infiltration and fluid collection), dilatation of the gallbladder, and an extrahepatic duct were also noted (Fig. 1A). An initial gastroduodenofiberscopy (GFS) via endoscopic US revealed that the intussusception was spontaneously released. There was no lead point except for fold thickening of the duodenum around the ampulla of Vater (Fig. 1B, left). The upper gastrointestinal series (UGIS; barium swallow) demonstrated a contrast-filled sac that was surrounded by a narrow lucent line in the contrast-filled duodenum just above the ampulla of Vater, which might represent the wall of the IDD. During the examination, the sac moved in accordance with normal peristalsis, and duodenal obstruction was not observed (Fig. 1C). On the retrospective review of MRI performed on the patient’s first visit, axial and coronal T2-weighted turbo spin-echo sequence showed a fluid-filled sac-like structure in the duodenum, arising just above the ampulla of Vater (Fig. 1D). Repeat GFS revealed two lumens in the duodenum. The larger one ended up as a blind pouch, and the smaller one led to the distal lumen of the duodenum. Minor and major papillae opened in the pouch, where the papillae were blocked by impacted food material (Fig. 1B, right).

Although the initial findings (from CT, MRI, GFS, and UGIS) suggested differential diagnoses of various congenital anomalies, such as a communicating duodenal duplication cyst or duodenal web, the laparoscopic treatment and subsequent pathology (described below) con-

Fig. 1. A 25-year old female with intraluminal duodenal diverticulum associated with duodeno-duodenal intussusception and recurrent pancreatitis, presenting with epigastric pain.

A. Contrast-enhanced-abdominal axial CT scans show duodeno-duodenal intussusception, presenting a “target sign” (left, arrow), at the second portion of the duodenum and edematous pancreas with peripancreatic fat infiltration and fluid collection, indicating acute pancreatitis (right, arrows).

B. Endoscopic US (left) shows two lumens in the second portion of the duodenum, with the endoscope in the blind pouch. After spontaneous reduction of intussusception, repeat gastroduodenofiberscopy (right) shows two lumens in the duodenum. The larger one ended up a blind pouch (arrow) and the smaller led to the distal lumen of the duodenum (arrowhead). Minor and major papillae opened in the pouch, all being blocked by impacted food material.



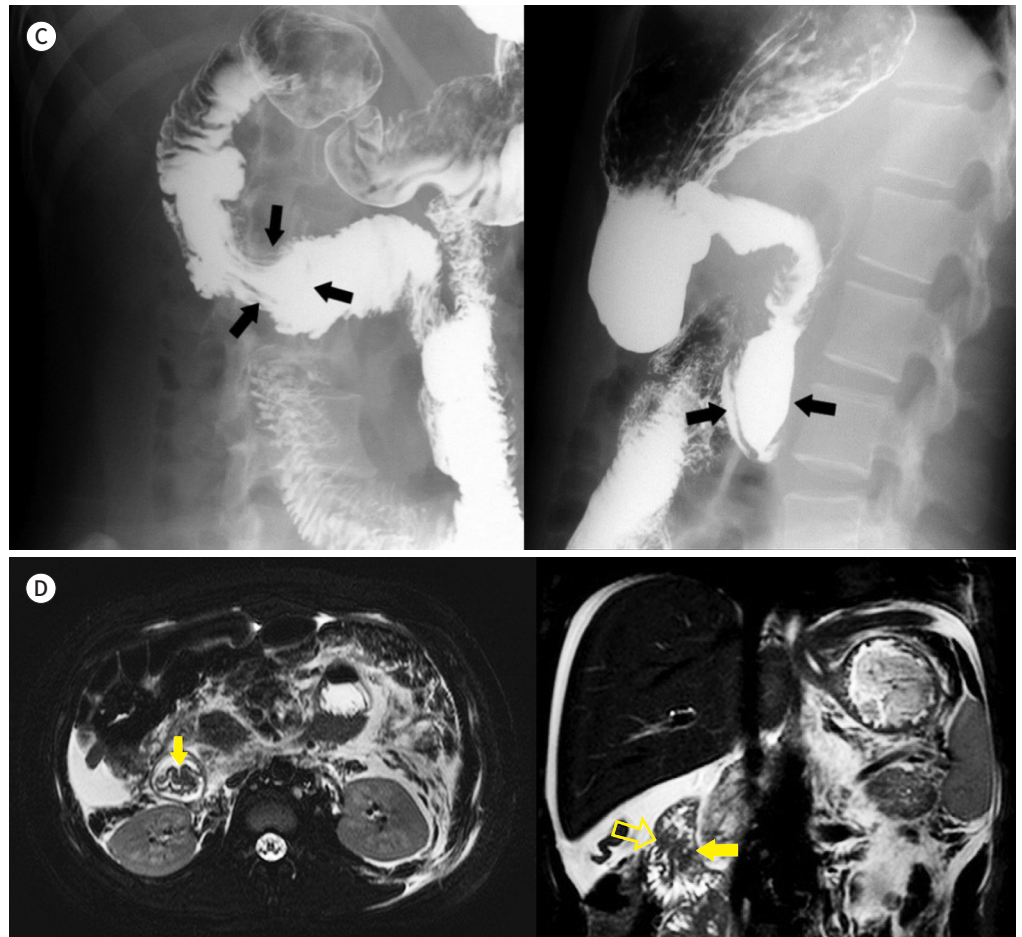
firmed IDD as the final diagnosis. Laparoscopic excision was performed to remove the IDD and to prevent recurrent pancreatitis. During laparotomy, a diverticular sac was identified and pulled outward to evert the duodenum. At the end of the everted duodenum, a small orifice leading to the stomach was found, which corresponded to the smaller lumen on the GFS. A small hole secreting serous fluid, which was the minor ampulla adjacent to the diverticular orifice, was also identified as the end of the everted duodenal mucosa (Fig. 1E). The sac was excised and sent for histopathologic evaluation, which determined the sac covering to consist of normal mucosa (on both sides), but without a proper muscle layer (Fig. 1F). Therefore, IDD was confirmed as the final diagnosis.

This study was approved by the Institutional Review Board of Ewha Womans University Mokdong Hospital (IRB No. 2020-10-026), and the requirement for informed consent was waived.

Fig. 1. A 25-year old female with intraluminal duodenal diverticulum associated with duodeno-duodenal intussusception and recurrent pancreatitis, presenting with epigastric pain.

C. Upper gastrointestinal series with the images obtained by placing the patient in the supine (left) and prone (right) positions show a contrast-filled sac surrounded by a narrow lucent line, which might represent the wall of intraluminal duodenal diverticulum in the second portion of the duodenum, just above the ampulla of Vater, indicating the “wind-sock” sign (arrows).

D. MR images obtained using axial T2-weighted turbo spin-echo sequence shows a fluid-filled sac-like structure in the duodenum arising just above the ampulla of Vater (left, arrow). Coronal T2-weighted turbo spin-echo sequence also shows the same structure in the duodenum (right, open arrow) just above the ampulla of Vater (right, arrow).



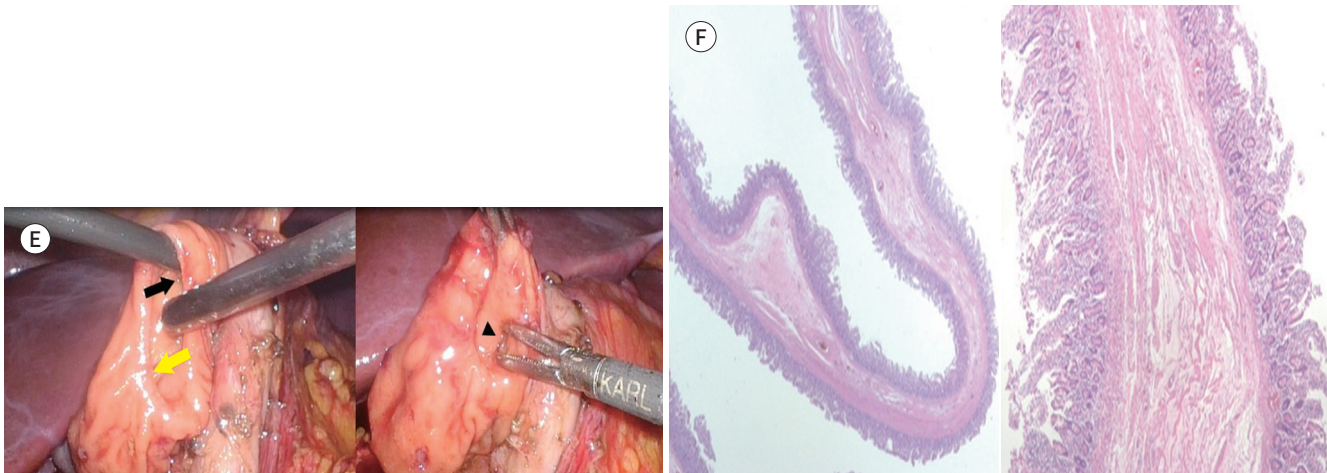
DISCUSSION

IDD is a rare congenital anomaly that represents a subtype of the duodenal web or diaphragm (1). Normally, the lumen of the duodenum becomes obliterated at 4–6 weeks of gestation due to rapid cell division; the subsequent recanalization is usually completed by 12 weeks of gestation (4). Incomplete or impaired recanalization of the lumen results in intraluminal lesions. Duodenal diverticula are classified as extraluminal duodenal diverticula (EDD) and IDD (5). In EDD, a sac of the mucosal or submucosal layer herniates through a muscular defect in the duodenal wall. In contrast, IDD consist of a sac that protrudes into the duodenal lumen, usually within the second portion of the duodenum (6).

Fig. 1. A 25-year old female with intraluminal duodenal diverticulum associated with duodeno-duodenal intussusception and recurrent pancreatitis, presenting with epigastric pain.

E. Intraoperative gross findings shows a sac-like structure pulled from the second portion of the duodenum through the laparotomy. Note the orifice leading to the proximal duodenum and the stomach (black arrow). Another orifice to the diverticular lumen is found (yellow arrow). A small hole secreting serous fluid is identified at the margin of the diverticular orifice, which was the minor ampulla (arrowhead).

F. Pathological analysis of the excised sac with hematoxylin-eosin staining (right, $\times 40$; left, scan view). Two representative views from the microscopic analysis, shows the duodenal mucosa and a submucosal layer in the diverticulum. A proper muscularis mucosa layer is not identified in the diverticulum.



Clinically, IDD is detected by UGIS using the duodenal “windsock” sign, appearing as a thin radiolucent line around the diverticulum (7). CT and MRI scans revealed a fluid-filled sac within the duodenum, similar to the pathognomonic “windsock” appearance mentioned above (1). Histopathologically, the sac appears very similar to a duplication cyst in the duodenum. However, the duodenal duplication cyst has a wall made of muscularis propria and presents peristalsis (which facilitates the outflow of bile) (6). IDD lacks both the muscularis propria layer and peristalsis. MRI findings can help differentiate between the two diseases. In the case of a duodenal duplication cyst, the signal does not change after superparamagnetic oral contrast medium administration, remaining high in T2-weighted sequences. On the contrary, IDD should become hypointense because of its connection with the lumen (6). Bremer differentiated congenital duplications from the diverticula in the presence of all normal intestinal layers (8).

The clinical presentation most frequently includes gastrointestinal bleeding secondary to ulceration of the diverticula and acute pancreatitis in 25% of cases, and acute pancreatitis in 20%. Postprandial epigastric colicky pain, vomiting, diverticulitis, and recurrent intussusception have also been reported (9). IDD symptoms do not appear when the patient is young but occurs mainly in adults, probably because of the enlargement of the diverticulum with age. In terms of the complication of recurrent pancreatitis, as in our patient, the underlying pathogenic mechanism remains unclear. A widely accepted hypothesis involves the reflux of diverticular content through the papilla of Vater (2). In accordance with peristalsis, food material fills the IDD and obstructs the papillae. Duodeno-duodenal intussusception is extremely rare because the duodenum is fixed in the retroperitoneal position and is always associated with a benign lead point, such as Brunner’s gland hamartomas, lipomas, and adenomas (10).

Our case demonstrated an IDD as the lead point of a duodeno-duodenal intussusception, involving the second portion of the duodenum. Since all the symptoms of IDD are due to anatomical issues, the ultimate treatment is surgery, such as anterior duodenotomy and excision of the intraluminal pouch (7).

In conclusion, although IDD is very uncommon, it should be listed as a potential lead point of duodeno-duodenal intussusception, especially in cases harboring a saccular structure in the second portion of the duodenum, as detected by radiologic imaging studies.

Author Contributions

Resources, L.H., Y.S.Y., P.S.H.; writing—original draft, Y.G.Y.; and writing—review & editing, L.J.K.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Kim JM, Lee NK, Kim S, Kim DU, Kim TU. Intraluminal duodenal diverticulum: CT and gadoteric acid-enhanced MRI findings. *J Korean Soc Radiol* 2015;72:176-179
2. De Rai P, Castoldi L, Tiberio G. Intraluminal duodenal diverticulum causing acute pancreatitis: CT scan diagnosis and review of the literature. *Dig Surg* 2000;17:288-292
3. Larsen PO, Ellebæk MB, Pless T, Qvist N. Acute pancreatitis secondary to duodeno-duodenal intussusception caused by a duodenal membrane, in a patient with intestinal malrotation. *Int J Surg Case Rep* 2015;13:58-60
4. Coley BD. *Caffey's pediatric diagnostic imaging*. 13th ed. Philadelphia: Elsevier 2018:953
5. Peng HL, Su CT, Chang CY, Lau BH. Intraluminal duodenal diverticulum in a child concomitant with an entrapped coin and a duodenal polyp. *Formos J Surg* 2014;47:236-239
6. Guarise A, Faccioli N, Ferrari M, Romano L, Parisi A, Falconi M. Duodenal duplication cyst causing severe pancreatitis: imaging findings and pathological correlation. *World J Gastroenterol* 2006;12:1630-1633
7. Karoll MP, Ghahremani GG, Port RB, Rosenberg JL. Diagnosis and management of intraluminal duodenal diverticulum. *Dig Dis Sci* 1983;28:411-416
8. Bremer IL. Diverticula and duplication of intestinal tract. *Arch Path Lab Med* 1944;38:132-140
9. De Castro ML, Hermo JA, Pineda JR, Carreira M, Dominguez F, Clofent J. Acute bleeding and anemia associated with intraluminal duodenal diverticulum: case report and review. *Gastrointest Endosc* 2003;57:976-979
10. Jeon SJ, Yoon SE, Lee YH, Yoon KH, Kim EA, Juhng SK. Acute pancreatitis secondary to duodenojejunal intussusception in Peutz-Jegher syndrome. *Clin Radiol* 2007;62:88-91

성인 십이지장-십이지장 장중첩증 및 재발성 췌장염과 관련된 관강내 십이지장 계실의 영상 소견: 증례 보고

이가영¹ · 이정경^{1*} · 이희성² · 이선영³ · 박상희⁴

관강내 십이지장 계실은 드문 선천성 이상으로, 십이지장 점막으로 구성된 낭성 병변이며, 위장 출혈, 십이지장 폐색 또는 췌장염과 함께 나타날 수 있다. 이 논문에서는 25세 성인 여성에서 십이지장-십이지장 장중첩증과 재발성 췌장염의 합병증을 일으킨 드문 관강내 십이지장 계실의 사례를 보고하는 바이다. CT, MRI, 상부위장관조영술 등의 영상 검사에서 십이지장의 중복낭을 의심하였으나, 수술로 절제된 조직에서 병리학적으로 고유근육층이 없는 관강내 십이지장 계실로 확진되었다.

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