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Case Report

Endoscopic treatment of periampullary duodenal duplication cyts in an 18-month-old girl

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ABSTRACT

Background. Duodenal duplication cysts (DDC) are rare congenital anomalies of the gastrointestinal tract and periampullary localization with anatomical variants including biliary and pancreatic duct anomalies remains a surgical challenge. Endoscopic treatment of the periampullary DDC (PDDC) communicating with the pancreaticobiliary duct in an 18-month-old girl is presented to discuss the endoscopic treatment options in children.

Case. An 18-month-old girl with a normal prenatal ultrasound (US) was asymptomatic until complaining of abdominal pain and vomiting at 10-months of age. Abdominal US revealed a 1.8 × 2 cm cystic mass adjacent to the second part of the duodenum. The amylase and lipase levels were slightly increased while she was symptomatic. Magnetic resonance cholangiopancreaticography (MRCP) showed a thick cyst wall measuring 1.5 × 2 cm at the second part of the duodenum, consistent with DDC that was suspected to be communicating with the common bile duct. Upper gastrointestinal endoscopy confirmed a bulging cyst in the duodenum lumen. The puncture and injection of the cyst with contrast material confirmed the communication of the duplication cyst with the common bile duct. The unroofing of the cyst was performed with endoscopic cautery. The biopsy obtained from the cystic mucosa revealed normal intestinal histology. Oral feeding was initiated six hours after the endoscopy. The patient has been followed for the last 8 months uneventfully.

Conclusions. Endoscopic treatment of PDDC with various anatomical variants can be considered an alternative to surgical excision in children.

Key words: periampullary, duodenal duplication cyst, endoscopy, children.

Duodenal duplication cysts (DDC) are rare congenital anomalies of the gastrointestinal (GI) tract and constitute 5-7% of all GI duplication cysts.[1] Two embryologic theories have been proposed to explain the development of DDC. They may occur due to duodenal epithelial pinching during the outgrowth of the dorsal pancreatic bud or secondary to epithelial sequestration.[1] DDC are typically adherent to the mesenteric site of the third or fourth part of the duodenum and are made up of an epithelial mucosal lining and a smooth muscle layer.[2]

DDCs localized adjacent to the major papilla and biliary-pancreatic papilla with or without an aberrant pancreatic duct draining into a cyst are defined as periampullary DDC (PDDC).[1] Periampullary type is much rarer and shows a more variable clinical presentation including pancreatitis, bleeding, duodenal obstruction and perforation.[3]

Although endoscopic treatment of PDDC is well defined in the adult population, there is little information regarding the use of endoscopic treatment in children. Bulotta et al.[3] reported

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4 pediatric cases managed with endoscopic intervention and reported favourable outcomes in children aged 11 to 14 years. However, the availability of endoscopic treatment in PDDC for small infants has not been reported. Therefore, an 18-months-old girl with PDDC is reported to discuss the technical tricks and results of endoscopic management of PDDC in young children.

Case Report

A girl weighing 3100 grams was born to a 29-year-old mother at the 38th week of gestation. The prenatal ultrasound (US) was normal and the patient was asymptomatic until 10 months of age. After complaining about abdominal pain and vomiting an abdominal US was performed, revealing a 1.5 x 2 cm cystic mass adjacent to the duodenum (Fig. 1). At the time of the symptoms, the complete blood count and liver function tests were normal. There was a slight increase in lipase (115 U/L normal: < 67 U/L) and pancreatic amylase (125 U/L, normal: 8-53 U/L) levels. The total (0.38 mg/dL, normal: 0.3-1.2 mg/dL) and direct bilirubin levels (0.083 mg/dL, normal: 0-0.2 mg/dL) were also within normal limits. Magnetic resonance cholangiopancreaticography (MRCP) revealed a thick-walled cyst adjacent to the second portion of the duodenum and pancreas, which was consistent with DDC and suspected to communicate with the common bile duct (Fig. 2). After obtaining consent from the parents, the patient underwent an endoscopic evaluation under general anaesthesia. The body weight of the patient was 12 kg at the time of the endoscopic evaluation. The procedure was performed using an adult therapeutic duodenoscope (PENTAX, ED-3490TK, Japan). The cyst was bulging at the second portion of the duodenum (Fig. 3a). The papilla was on the proximal side of the cystic protrusion. The cyst was punctured with a sclerotherapy needle (Micro-Tech Endoscopy, China) and contrast material was injected into the cyst (Fig. 3b). After confirming the communication of cysts with the common bile duct, incision of the cyst wall with a needle knife sphincterotome (Boston Scientific, USA) was performed with an electrosurgical generator (Olympus, ESG-100, pulse cut slow, (Fig. 3c). A biopsy was obtained from both the cyst wall and the inner layer of the cystic cavity. The histopathological evaluation of the biopsies revealed duodenal epithelium in the cyst wall confirming the diagnosis of DDC (Fig. 4). There were no perioperative and postoperative complications, and the patient was fed orally 6 hours after the operation. The patient was released from the hospital the next day and has been monitored for the past eight months without incident. Long-term follow-up is planned for possible malignant transformation.

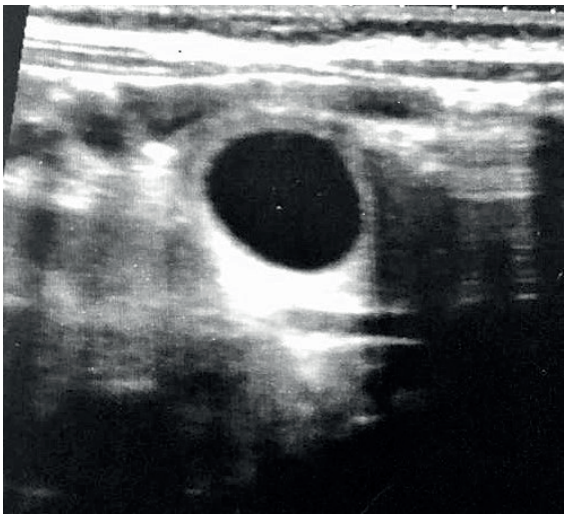

Fig. 1. Ultrasound image shows thick-walled duodenal duplication cyst.

Discussion

PDDC are extremely rare type of duplication cysts in the GI tract and constitutes a major surgical challenge because of the close localization to the biliary and pancreatic tree. Endoscopic treatment of PDDC enables better definition of the intra-luminal cyst, accurate localization of the papilla and distinguishes PDDC from other cystic lesions.[3] Although endoscopic treatment of PDDC is well defined in adults, there is scant information for the pediatric population. In a literature search, Bulotta et al.[3] reviewed 20 pediatric cases with

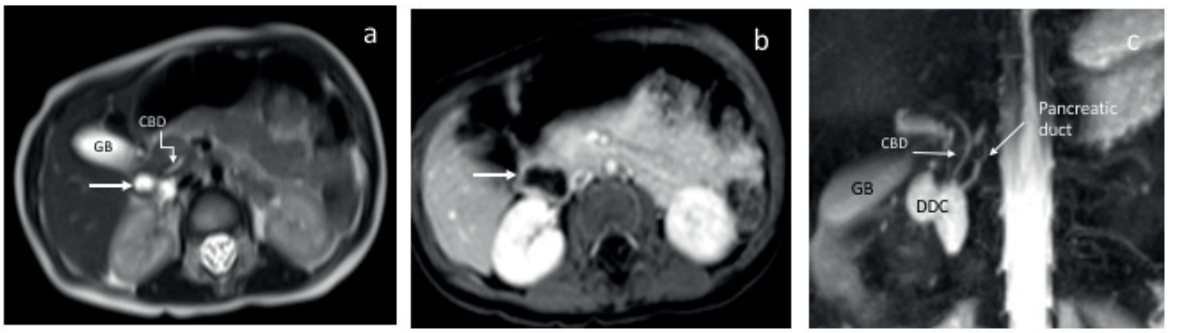

Fig. 2. (a) Axial T2-weighted, and (b) postcontrast T1-weighted, (c) coronal T2-weighted MRCP images show the duodenal duplication cyst (arrows in a and b) adjacent to the duodenum and the pancreas. CBD: common bile duct, DDC: duodenal duplication cyst, GB: gallbladder, MRCP: magnetic resonance cholangiopancreatography.

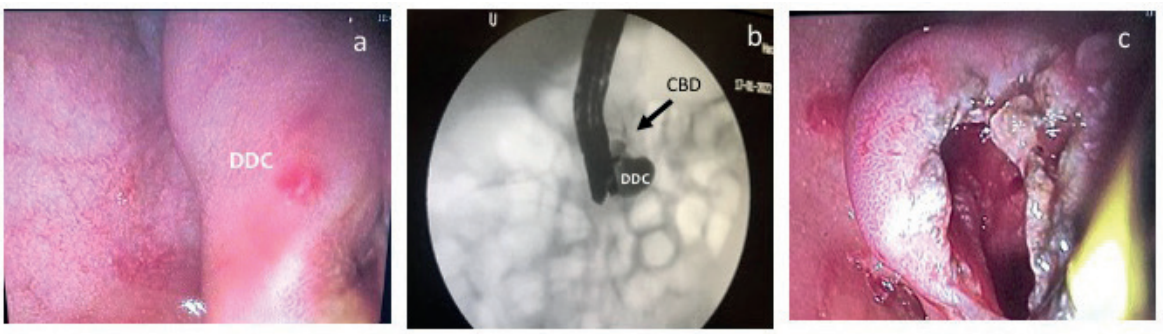

Fig. 3. Endoscopic diagnosis and treatment of periampullary DDC: (a) DDC bulging into the lumen of second part of the duodenum. (b) The contrast material injected to the cyst confirms the communication of the cyst with the common bile duct. (c) Unroofing of the cystic wall with endoscopic cautery. CBD: common bile duct, DDC: duodenal duplication cyst.

PDDC and only 50% of them were managed endoscopically. The median age of the patients was 14 years and the youngest patient was 3 years-old.[3,4] Herein, we report the youngest case of successful endoscopic management of PDDC in children.

Abdominal pain, vomiting and clinical findings of pancreatitis are common presentations of PDDC in children. In addition to the localization and size of the cysts, the patient may have a variety of symptoms due to anatomical variations. Pancreatitis may occur due to external obstruction of the papilla or obstruction due to debris or mucus secreted from the cyst. The biliary sludge in the pancreatic duct may also be seen in DDC. Therefore, detailed evaluation of anatomical variation is mandatory to differentiate DDC from other cystic lesions especially from common bile duct cysts.

US is highly suggestive for DDC. An outer hypoechoic muscle layer with an internal echoic inner layer suggests a duplication cyst. Although US is used as the first diagnostic tool, more comprehensive radiologic evaluation is needed to define the anatomical variations.[5] Preferably, MRCP can be used to define the communication between the pancreatic duct and the cyst.[6] Therefore, we performed MRCP and suspected that both the common bile duct and pancreatic duct were draining into the cyst.

Moreover, endoscopic evaluation is also useful to define the major papilla and obtain an endoscopic retrograde cholangiopancreatography (ERCP). In patients with large cysts, filling the lumen of the duodenum, the major papilla cannot be defined and it may not possible to obtain an ERCP. Therefore, we suggest that endoscopic injection of contrast material is a good option

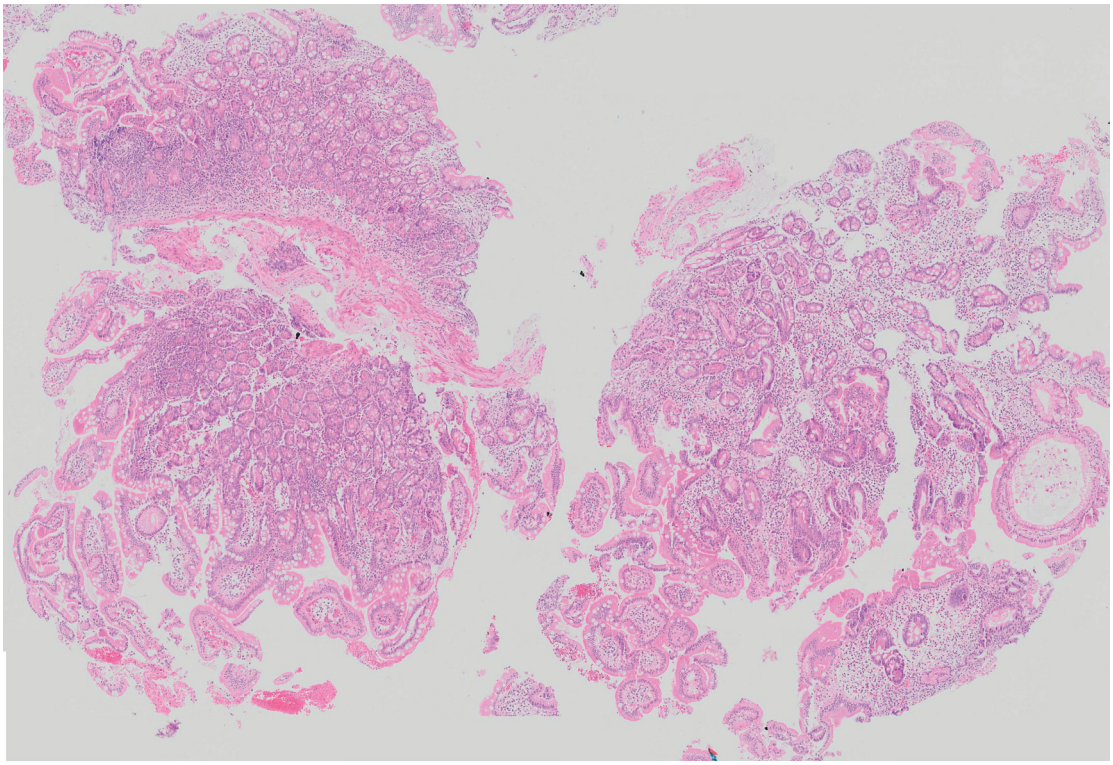

Fig. 4. The biopsy obtained from the cyst wall revealed duodenal epithelium (H&E stain).

to confirm the communication of the DDC with the biliary and pancreatic duct.

The surgical resection of PDDC can be demanding and may cause severe complications in small children. Endoscopic treatment consists of incision of the cyst wall, marsupialization, unroofing and sphincterotomy if needed.[4] Needle-knife, sphincterotome and endoscopic cautery can be used for cyst incision.[4] Although removing the cystic mucosa is recommended to prevent malignant transformation, endoscopic treatment of PDDC is highly effective for relieving symptoms. Endoscopy has several advantages over surgical excision. It enables direct visualization of the cystic mass in the duodenum and provides accurate localization of the papilla.[3] Endoscopic excision is well tolerated even in small children, with better cosmetic results and fewer complications. Although endoscopic incision of PPDC in small infants is not technically demanding, puncture of the bulging cyst may be needed in small children to have a wider working space. Finally, it has no visible scars and can be performed on an outpatient basis. The most important disadvantage of endoscopic treatment is that it leaves gastric and pancreatic mucosa with the potential for malignant complications.[7] Therefore, all parents should be informed about the risk of malignant transformation and close follow-up with endoscopic surveillance and/or radiologic imaging should be recommended.

Bleeding is an important but very rare complication of the endoscopic incision of the PDDC. It can be treated with metallic clips and blood transfusion.[3] This complication can be easily prevented by using endoscopic cautery, and in most of the cases early oral feeding can be initiated.

The differential diagnosis of PDDC from other cystic lesions, particularly choledochocele, is critical. Although some authors consider biopsy of the cyst wall to be the gold standard, intestinal mucosa has also been reported in

choledochocele.[8] However, a biopsy of the cyst wall should be routinely performed for histologic confirmation. Moreover, Antaki et al. reported that 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.[9] In our patient, the papilla was on the proximal side of the cystic protrusion, which suggests a PDDC more than a choledochocele.

Although, DDCs are benign lesions, a few cases of malignant transformation have been reported.[7,10] Several years after endoscopic treatment, it has been reported that asymptomatic adult patients have undergone malignant transformation. Therefore, long-term follow-up is recommended, particularly in patients treated endoscopically.[3] Despite the fact that there is no clear recommendation about follow-up periods in endoscopically treated patients, asymptomatic cases can be evaluated by US, whereas endoscopic evaluation is needed in symptomatic cases.

In conclusion, endoscopic treatment of PDDC with various anatomical variants can be considered an alternative to surgical excision in children. It is an easy and safe procedure, even for young patients. Long-term follow-up is mandatory for endoscopically treated patients because of the potential risk of malignancy.

Ethical approval

Informed consent was obtained from the parents.

Author contribution

The authors confirm contribution to the paper as follows: study conception and design: TS, EP, data collection: ÖB, HBA; analysis and interpretation of results: BO, DO; draft manuscript preparation: TS, EP. All authors reviewed the results and approved the final version of the manuscript.

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Conflict of interest

The authors declare that there is no conflict of interest.

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