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A large intrahepatic duodenal duplication cyst in a 3 year-old girl

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ABSTRACT

Duodenal duplication cyst consists 6% of alimentary tract duplications and the prevalence is estimated to be less than 1 per 100,000 live births. This report is the first case report in a pediatric patient. A 19-month-old female patient was detected with a 5.2 cm sized intrahepatic cystic lesion in her follow-up ultrasonography examination for hydronephrosis. In her regular check-up, findings suggestive of food materials were detected inside the cyst. Further evaluation showed a 7 cm sized cyst located inside segment IV of the liver, connected to the duodenal bulb with a possibility of biliary communication. On exploration, duodenal duplication cyst with a 1.5 cm long stalk starting from the duodenal bulb to the hilum of the liver was identified. Due to its sophisticated location and possible communication with the biliary tree, Roux-en-Y cystojejunostomy was performed after resecting the stalk. The histopathologic finding of the stalk showed serosa, muscle, submucosa and duodenal mucosa which suggested its duodenal origin. The patient has been followed up without any complications for 7 months.

Alimentary tract duplication is a rare congenital disease which can occur anywhere from the oropharynx to the anus [1]. Duodenal duplication cyst consists 6% of alimentary tract duplications and the prevalence is estimated to be less than 1 per 100,000 live births [2–4], According to a meta-analysis published in 2010, 38 studies which included 47 cases of duodenal duplication cyst were accessible during the period of 1999–2009 [5]. Symptoms of duodenal duplication cyst include vague abdominal pain with or without nausea and vomiting (80%), early satiety, pancreatitis, cholestasis, gastrointestinal bleeding and failure to thrive [5].

First case of an intrahepatic duodenal duplication cyst in a 44 year-old female patient was published in 1977 [6]. Our report is the second report after the case reported in 1977 and the first case of an intrahepatic duodenal duplication cyst in a pediatric patient.

The purpose of this report is to introduce a very rare case of intrahepatic duodenal duplication cyst and share our successful treatment experience.

1. Case report

The patient was a girl born at 40 weeks gestation by normal vaginal delivery at an outside hospital. Birth weight was 3.15 kg. Although a cystic lesion in the biliary tract was suspected in prenatal ultrasonography, the follow up exam that was taken after birth showed no abnormal findings except mild hydronephrosis. The patient showed normal growth with no abdominal symptoms. At the age of 19 months, intrahepatic cystic lesion was detected during a follow up ultrasonography exam for the hydronephrosis. The cyst was measured 3.8 × 5.2 × 3.4 cm in abdominal magnetic resonance imaging (MRI). However, the patient showed no abnormal signs and symptoms such as jaundice or abdominal pain and no other invasive procedure were undertaken. The patient was followed up with an impression of benign hepatic cyst. However on ultrasonography that was taken at the age of three, there were findings suspected to be food material inside the intrahepatic cystic lesion. She was referred to our center for further evaluation. The patient showed no abnormal symptoms and there was no abnormal sign on physical examination. Height and weight were both 50 percentile for her age. Laboratory values were all within normal range. Plain abdominal radiography showed a radiolucent air-fluid level in the right upper quadrant. In the abdominal ultrasonography, a huge intrahepatic cystic lesion which was bearing materials suspected to be undigested food was measured to be 6.6 cm in diameter and

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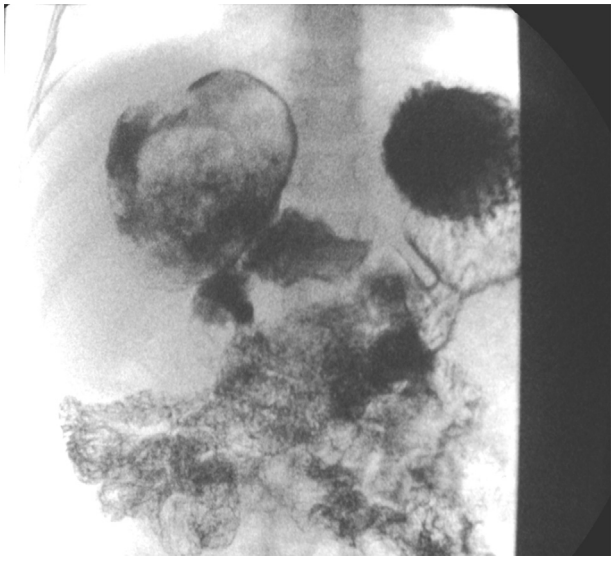

Fig. 1. Upper gastrointestinal series demonstrates a large duodenal duplication cyst above the duodenal bulb.

a preduodenal portal vein was also detected. In the upper gastrointestinal series and abdominal MRI, the cyst was found to be located in segment IV of the liver with a size of $7 \times 4.1 \times 3.1$ cm communicating to the duodenal bulb (Figs. 1 and 2). Hepatobiliary (DISIDA) scan was taken to rule out the possibility of communication with the biliary tree, which showed radioactive tracers inside the cyst. An exploratory laparotomy was undertaken.

A supraumbilical transverse incision was made. After identifying the duodenum, duodenal duplication cyst possessing a long stalk starting from the duodenal bulb to the hilum of the liver was identified (Fig. 3A). The gap between the hilum and the duodenal bulb was approximately 1.5 cm. The width of the stalk was measured 2 cm. The liver was not cirrhotic, located in a normal position with a hypertrophied left lobe. Duplication cyst was located within segment IV of the liver. A preduodenal portal vein was communicating with the splenic vein through an intervening vein, and passed above the duodenal bulb anteriorly. The common bile duct was on the right side of the preduodenal portal vein. The right and left hepatic ducts joined together at the level of the duodenal 1st portion. After joining with the cystic duct, the bile duct went into the pancreas at the level of the distal part of 2nd portion of duodenum. Cholecystectomy was performed and stalk of the duodenal cyst and right and left hepatic ducts were dissected cautiously. Injury of the anterior wall of the stalk was inevitable in trying to avoid injury of the anteriorly located bile duct. The stalk was transected after complete exposure of the duodenal bulb. Food materials were found undigested in the cyst. Operative cholangiography showed no additional information due to the remaining barium contrast left inside the duplication cyst after the upper gastrointestinal series. Food materials were removed and the inner layer mucosa of the intrahepatic cyst was identified. Primary closure of the stalk opening was done in the duodenal bulb. Roux-en-Y cystojejunostomy was performed using the proximal jejunum (Fig. 3B).

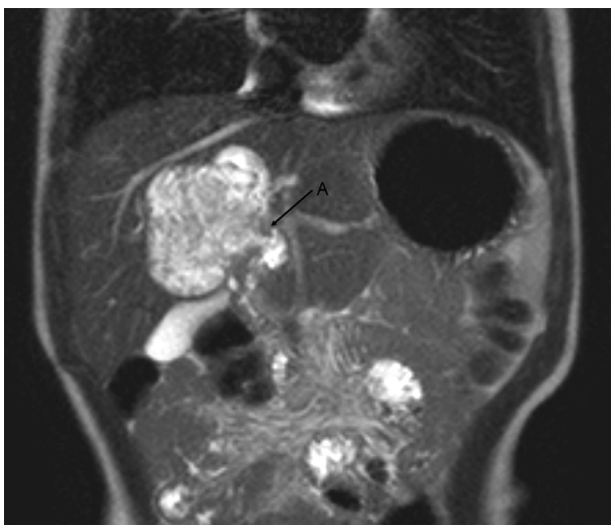

Fig. 2. Abdominal MRI shows an intrahepatic cyst mass with a stalk (A).

The histopathologic finding of the stalk showed serosa, muscle, submucosa and duodenal mucosa which suggested its duodenal origin. Submucosal chronic inflammation with hypertrophy of proper muscle was observed in the pathology (Fig. 4).

The patient began oral diet on the 5th postoperative day and was discharged with no other complication on the 7th postoperative day.

2. Discussion

Alimentary tract duplication is a tubular or cystic mass-like lesion which can occur at any location of the gastrointestinal tract. Duplications were described in various terms such as the followings; giant diverticula, enterogenous cysts, ileum or jejunal duplex, giant thoracic cyst, duplications and unusual Meckel diverticula [7–10]. In 1937, William E. Ladd proposed the term “duplications of alimentary tract” [11] and Gross et al. pointed out the features as the followings: (1) the presence of a coat of smooth muscle, (2) an epithelial lining representing some type of intestinal tract mucosa, and (3) intimate anatomic association with some portion of the alimentary tract [12].

In a meta-analysis published in 1997, duodenal duplication consist 6% of all alimentary tract duplications [2], while other literature report a proportion of 2–12% [3,13–17]. While most duodenal duplications have communications with the pancreatic or biliary duct, only a few cases showed communication to the native duodenum [5,18]. The majority of duplications are diagnosed within the age of 2 years [19]. However, duodenal duplications showed abdominal symptoms in only 40.4% until 10 years and diagnosis rate until 20 years was 61.7% [5].

Congenital hepatic cyst is also comparably rare while the prenatal diagnosis rate is increasing [20]. Simple hepatic cysts are considered to arise from aberrant biliary tree, which gets obstructed from the main biliary system [21]. Most simple hepatic cysts do not require treatment due to its spontaneously regressing nature [22]. Simple hepatic cysts produce clear fluid showing no communication to the intrahepatic biliary system [23].

Embryologically, the duodenum develops from the caudal aspect of the primitive foregut and the cranial aspect of the primitive midgut during the start of the 4th week [24]. At the same time, the hepatic diverticulum buds from the ventral wall of the primitive midgut. The diverticulum becomes the anlage for the development of the future liver, extrahepatic biliary ducts, gallbladder, and ventral pancreas [25,26]. A small lumen is formed