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.

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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 $3.8 \times 5.2 \times 3.4$ 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...
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).

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, gall-bladder, and ventral pancreas [25,26]. A small lumen is formed
inside the primitive duodenum during the 6th week. In the 7th and 8th week, duodenum increases in diameter and the vacuoles coalesce and begin to restore the intraluminal patency [27]. The coalescence of vacuoles forms two channels in the duodenum which appears in the developing biliary system [27,28]. The common hepatic duct opens into the gallbladder and cystic duct. Cystic duct continues in place of the absent common bile duct to open into the duodenum [29]. Although the two organs are embryologically intertwined during their development, the chance of duodenal duplication cyst or diverticulum communicating with the biliary tree or liver is very low.

Alimentary tract duplications have a potential to develop infection, bleeding, intestinal obstruction or volvulus, and malignancy. Therefore it should be treated by early complete resection [30]. In our case, complete resection of the intrahepatic cyst was impossible without a massive liver resection or transplantation due to its location. We consider Roux-en-Y cystojejunostomy to be the most appropriate management in case of the duodenal duplication cyst communicating to the biliary tree considering its sophisticated location.

This report is the second case report of an intrahepatic duodenal duplication cyst and the first case report in a pediatric setting.

Fig. 3. A) Schematic anatomy of the intrahepatic duplication cyst. B) Schematic anatomy after Roux-en-Y cystojejunostomy.

Fig. 4. Histologic appearance of the stalk. A) Whole layer of the stalk. B) Duodenal mucosa with paneth cell. C) Smooth muscle hyperplasia.
patient. The patient showed no abdominal pain, jaundice or any other signs of complications during her 7 months of follow up since the operation.

3. Conclusion

Intrahepatic duodenal duplication cyst is a very rare case and we want to share our successful treatment experience.

References