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A case of prenatally detected hepatic cyst communicating with the hepatic duct☆

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ABSTRACT

Here, we describe the case of a 9-year-old girl. During the patient's prenatal period, her mother had suffered domestic violence perpetrated by the patient's father. On maternal ultrasonography and magnetic resonance imaging, an intraabdominal cystic lesion was detected in the fetus at around the 30th prenatal week. The patient was delivered normally, and there were no evident anomalies on her body. Computed tomography with drip infusion cholangiography and percutaneous retrograde transhepatic cholangiography demonstrated an intrahepatic cyst of approximately 3 cm in diameter, which was located at S_5–S_8 and communicated with the confluence of the bilateral hepatic ducts. The cyst is clinically conjectured to be a solitary intrahepatic biliary cyst. However, it remains possible that the cyst is a ciliated hepatic foregut cyst or indicates hepatic injury that may have occurred as a result of domestic violence to the mother. Careful, long-term observation of the patient will be continued to ensure that any malignant transformation is not missed.

The use of ultrasonography or magnetic resonance imaging (MRI) can prenatally visualize various congenital diseases as intraabdominal cystic lesions. These congenital diseases include congenital biliary dilatation, hydronephrosis, renal cyst, ovarian cyst, mesenteric cyst, intestinal atresia, and meconium peritonitis. However, the incidence of congenital hepatic cysts is considered to be very low (0.01%) [1]. In this report, we describe an extremely rare case of a prenatally detected hepatic cyst communicating with the hepatic duct.

1. Case report

The patient is a girl who is currently 9 years old. During the patient's prenatal period, her mother had suffered domestic violence perpetrated by the patient's father. In the 23rd gestational week, an epigastric cystic lesion was detected in the fetal abdomen using maternal ultrasonography. In the 30th gestational week, MRI revealed an intrahepatic or extrahepatic cystic lesion in the fetus (Fig. 1). On the sixth day of the 38th gestational week, the patient was born by scheduled cesarean section, which was performed because the mother had previously undergone cesarean delivery.

The baby girl weighed 2675 g at birth and had an Apgar score of 9 points at both 1 and 5 min after birth. There were no apparent anomalies on her body. Her respiratory and circulation systems functioned normally. Her abdomen was soft and flat, without any palpable masses, and her extremities were normal, in both appearance and function.

All blood data were consistent with a normal neonate at birth, and there was no evidence of jaundice, liver dysfunction, or inflammation (Table 1).

After birth, an abdominal ultrasound of the patient revealed an irregular cystic lesion that measured approximately 3 cm in diameter. The lesion was located in segments 5–8 (S_5–S_8) of the liver, had a clear border, showed posterior echo enhancement, and appeared to contain biliary sludge. We observed mild dilatation of the intrahepatic bile ducts near the porta hepatis. The cyst was adjacent to both the gallbladder and the middle hepatic vein, which was mildly displaced to the left on Doppler ultrasonography. No blood flow was detectable in the cyst, and no dilation of the extrahepatic bile duct was evident (Fig. 2).

All authors had access to the data and a role in writing the manuscript.

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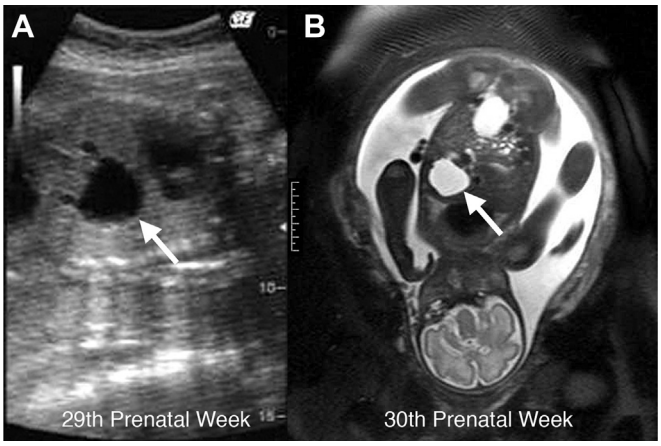

Fig. 1. Maternal ultrasonography and magnetic resonance imaging before the birth of the patient. An intrahepatic or extrahepatic cystic lesion is apparent in the fetus (arrows). Hydramnios is not seen.

We first suspected a congenital biliary dilatation. However, this possibility was soon rejected for the following reasons: (1) the cyst was intrahepatic and adjacent to the middle hepatic vein, (2) no dilatation was observed in the extrahepatic bile duct, (3) the patient did not have jaundice or liver dysfunction, and (4) the patient did not have hyperamylasemia or inflammations. Accordingly, we made preparations for further examination: computed tomography (CT), upper gastrointestinal series, hepatobiliary scintigraphy, magnetic resonance cholangiopancreatography (MRCP), and percutaneous antegrade transhepatic cholangiography (PTC).

On drip infusion cholangiography CT (DIC-CT), influxes of contrast medium were observed in the cyst and the gallbladder. No other lesions were detected in the liver, and no dilation of the extrahepatic bile duct was observed. A three-dimensional DIC-CT image reconstruction displayed the enhanced cyst, located just between the bilateral hepatic ducts. The cyst had an enhanced density that was almost identical to all major components of the bile duct system, including the bilateral hepatic ducts, cystic duct, and common bile duct, except the gallbladder (Fig. 3).

Upper gastrointestinal series showed good passage through the upper gastrointestinal tract and no enlargement of the duodenal C-loop.

Hepatobiliary scintigraphy with technetium-99m Sn-N-pyridoxyl-5-methyltryptophan (99mTc-PMT) demonstrated normal accumulation of radioisotope in the porta hepatis, a cold area nearby and normal excretion of radioisotope to the duodenum (Fig. 4).

Table 1
Laboratory data at birth.

WBC	12,200/mm^3
RBC	390 × 10^4/mm^3
Hb	14.7 g/dl
Ht	43.7%
Plt	36.5 × 10^4/mm^3
CRP	0.01 mg/dl
TP	5.6 g/dl
Alb	3.7 g/dl
T.Bil	2.44 mg/dl
D.Bil	0.56 mg/dl
AST	26 IU/L
ALT	6 IU/L
ALP	544 IU/L
LDH	258 IU/L
AMY	10 IU/L
Lipase	7 mg/dl

MRCP demonstrated an irregular cystic mass, which had an intensity that was almost identical to the intensities of the gallbladder and the common bile duct. The connections between the cyst and bile ducts were not obvious (Fig. 5).

PTC was performed under general endotracheal anesthesia 2 months after the patient's birth. A greenish bilious fluid (7 mL) was aspirated from the cyst; it was not mucinous. Although the contrast medium sequentially revealed the cyst, the bilateral main hepatic ducts, gallbladder, common bile duct, and duodenum, some branches of the main bile ducts could not be identified. The presence of a very narrow communication between the cyst and the confluence of the bilateral hepatic ducts was confirmed by PTC (Fig. 6).

Biochemical investigation of the fluid aspirated during PTC did not indicate reflux of pancreatic juice. Instead, results indicated the presence of bile in the cyst (Table 2). The day after PTC was performed, ultrasonography revealed that the cyst had entirely recovered its original size and configuration.

Because the patient did not manifest any symptoms and blood panels did not suggest any disorders, no additional testing was performed. At present, the patient is healthy and displays normal growth and development. She has no clinical symptoms or abnormal characteristics (for example, jaundice, abdominal pain, or liver dysfunction). The cyst has not changed in size, although the patient has grown considerably, and the intrahepatic duct dilatation has already disappeared from ultrasound results. Currently, the patient is thought to have a variety of congenital hepatic cyst communicating with the bile ducts. She has been carefully followed up as an outpatient.

2. Discussion

Although congenital intraabdominal cystic lesions occasionally arise in fetuses or neonates, most of these lesions are extrahepatic and, indeed, lesions in the liver are extremely rare [1]. We suggest three possible etiologies for our case.

First, this case appears to be consistent with a solitary intrahepatic biliary cyst (SIBC). Nonparasitic liver cysts are classified into solitary or multiple cysts [2], and the solitary liver cysts (SLCs) are further categorized as simple solitary liver cysts or SIBCs [3] (Fig. 7). A few etiologies have been suggested for SLCs, namely, they may result from the obstruction of an aberrant bile duct caused by infection, vascular disruption of the fetus, or overdevelopment of the intralobular bile ducts [2–8]. In addition, Soyer et al. [2] hypothesized that insufficient drainage of the hypoplastic bile ducts accompanied by stenosis in some regions may cause dilatation of the bile pool, which results in an SIBC.

SLCs are usually asymptomatic—their symptoms do not manifest until patients are in their forties or fifties [2] and neonatal cases are extremely rare [4]. The antero-inferior segment of the right hepatic lobe is the most frequent site of SLC [2,4], which is consistent with the findings in our case. Furthermore, it is possible that SLCs develop connections to the biliary tracts.

Soyer et al. [2] reported a case of a newborn with SIBC that required an urgent laparotomy because of its progressive enlargement. They were able to demonstrate the presence of a connection between the cyst and the biliary tree on retrograde cholangiography via the common bile duct or anterograde cavitography; however, we found that anterograde cavitography could accurately determine that the cyst was near the bilateral hepatic ducts and, indeed, had a short connection with their confluence. In our case, DIC-CT could also indicate the connection. These findings suggest that SIBC may originate from the biliary system.

Alternatively, the patient's lesion may be a ciliated hepatic foregut cyst (CHFC), which is an extremely rare congenital lesion of