Case Report

Congenital Benign Intrahepatic Cyst with Cystobiliary Communication: An Intraoperative Diagnostic Enigma

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Received: November, 2017. Accepted: February, 2018.

INTRODUCTION

Congenital benign liver cystic lesions are rare, heterogeneous in etiopathogenesis, and preoperative radiology may not always clinche the precise diagnosis. This manuscript discusses a 15-month-old male explored with preoperative imaging diagnosis of mesenchymal hamartoma. The operative findings of the cyst with frank bile and a large cystobiliary communication, however, presented a diagnostic challenge. The stepwise intraoperative approach to reaching a rare diagnosis and the operative procedure with literature review are discussed.

CASE REPORT

A 15-month-old male presented with the history of a right upper quadrant mass noticed at 3-month age with rapid increase in size, decreased feeding, and intermittent vomiting for 1 month. Clinical examination revealed a liver mass occupying right hypochondrium, epigastrium, and right iliac fossa. The mass was nontender with smooth surface, rounded margins, and firm consistency.

Liver function tests revealed raised transaminases (aspartate transaminase/alanine transaminase: 319/204 u/L) and raised gamma-glutamyl transpeptidase (GGT: 687 u/L) though serum bilirubin, liver synthetic functions, and serum alfa-fetoprotein levels were normal. Triple-phase computed tomography demonstrated the following: Hypodense cystic lesion (size 15 cm × 10 cm × 11 cm) occupying segments 4, 5, and 8; irregular margins in the cranial segments; few peripheral septations with poor enhancement; and no calcification [Figure 1a]. The left portal vein and left hepatic duct were compressed with consequent left lateral segment atrophy and left intrahepatic biliary dilatation. On T2-weighted magnetic resonance imaging (MRI), the cyst was uniformly hyperintense [Figure 1b]. The child was explored with preoperative diagnosis of a mesenchymal hamartoma.

Surgical exploration revealed a subcapsular cystic lesion occupying segments 4, 5, and 8 [Figure 2]. Cyst decompression yielded one liter of frank bile. A subsequent transcholecystostomy intraoperative cholangiogram confirmed a cystobiliary communication with right posterior sectoral bile duct. Cyst deroofing revealed a 0.5-cm well-epithelialized bile duct opening into the base of the cyst at the hilum. Frozen section histopathology of cyst wall was reported as stratified squamous and ciliated columnar epithelium with underlying connective tissue and smooth muscle layer, thereby suggesting the diagnosis of ciliated hepatic foregut cyst (CHFC). A right extended hepatectomy was precluded because of the atrophic left lateral segment (volume of segments 2 and 3: 23 ml). A left extended hepatectomy was technically precarious because the...

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Quick Response Code:

Website: www.jiaps.com

DOI: 10.4103/jiaps.JIAPS_50_17

How to cite this article: Kishore JS, Lal R, Chinya A, Nijagal Mutt JB. Congenital benign intrahepatic cyst with cystobiliary communication: An intraoperative diagnostic enigma. J Indian Assoc Pediatr Surg 2018;23:93-5.
right hepatic vein was very closely abutting along the entire course, the segment 7 portal vein branch was seen to join the right anterior portal vein on preoperative imaging, and the preoperative cholangiogram suggested a cystobiliary communication with the right posterior sectoral duct. Hence, in view of a benign lesion, subtotal cyst excision was performed leaving behind cyst wall remnant which shared a close interface with the right hepatic vein. The residual mucosa was ablated and a Roux-en-Y loop of jejunum was anastomosed to the bile duct communicating with the cyst at the hilum.

At 1-year follow-up, the baby is asymptomatic with normal liver enzymes and resolution of intrahepatic biliary dilatation on ultrasound imaging.

**Discussion**

The differential diagnosis of a congenital cystic liver lesion in children is as follows: Simple hepatic cyst, cystic mesenchymal hamartoma, epidermoid cyst, intrahepatic choledochal cyst, CHFC, and lymphangioma. A cystobiliary communication narrows down the differential diagnosis further to a CHFC; an intrahepatic choledochal cyst and an epidermoid cyst.[1]

CHFC is rare with just over 100 reported cases reported (median age: 52 years). Literature search revealed only 12 pediatric cases including the case presented here. The clinical details of reported pediatric cases are summarized in Table 1. Five were diagnosed prenatally. A cystobiliary communication was demonstrated in 5 reported CHFC cases.

CHFCs are derived from the embryonic foregut; possibly detached outpouching from hepatic diverticulum. The most common site is segment 4 of liver because segment 4 constitutes major liver bulk at 5th–8th week of embryogenesis. The four classic histological features of CHFC are inner lining of columnar epithelium often

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**Table 1: Clinical details of reported pediatric cases (<12 years) diagnosed with ciliated hepatic foregut cyst**

| Study               | Age/sex     | Cyst size           | Clinical presentation                      | Surgical management               |
|---------------------|-------------|---------------------|-------------------------------------------|-----------------------------------|
| Rogers et al.[1]    | 10 months/male | 10 cm × 7.5 cm × 5 cm | Prenatal diagnosis. Postnatal gross abdominal distension | Extended right hepatectomy         |
| Khoddami et al.[2]  | 3.5 years/male | 3.7 cm × 2.8 cm     | Prenatal diagnosis. Postnatal pain abdomen | Cyst excision                      |
| Khoddami et al.[2]  | 16 months/female | 3.6 cm × 3.2 cm    | Postnatal incidental detection            | Cyst resection                     |
| Betalli et al.[3]   | 11 months/male | 4 cm × 5 cm         | Prenatal diagnosis. Asymptomatic           | Segment 4 cyst excision            |
| Guérin et al.[4]    | 14 months/female | 8.8 cm × 8.3 cm × 6.1 cm | Prenatal detection, asymptomatic          | Central hepatectomy for segment, IV and V cyst, Roux-en-Y HJ |
| Guérin et al.[4]    | 10 years/male | 2 cm subcapsular    | Incidental postnatal detection. Asymptomatic | Central hepatectomy for segment, IV and V cyst, Roux-en-Y HJ |
| Guérin et al.[4]    | 10 years/female | 6 mm subcapsular    | Asymptomatic. Detected at deceased donor hepatectomy | Segment 4 wedge resection          |
| Kim et al.[6]       | 2 years/female | 11 cm × 10 cm × 7 cm | Postnatal detection, symptomatic           | Partial enucleation, marsupialization |
| Carciner et al.[7]  | 3 years/male | 2 cm × 1.5 cm       | Asymptomatic                               | Cyst excision                      |
| Carciner et al.[7]  | 5 years/female | 2 cm × 1.3 cm       | Asymptomatic. Incidental detection         | USG-guided aspiration only         |

USG: Ultrasound, HJ: Hepaticojejunostomy

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**Figure 1:** (a) Contrast-enhanced computed tomography of the abdomen showing solitary cystic liver lesion with few peripheral septa. (b) Magnetic resonance imaging picture showing a solitary hyperintense cyst in the liver in a T2-weighted image.

**Figure 2:** Intraoperative appearance of the ciliated hepatic foregut cyst. Note the central (segments 4, 5, and 8) and subcapsular location of the cyst. GB: Gallbladder
with squamous metaplasia, loose connective tissue, smooth muscle, and fibrous capsule.

The spectrum of reported clinical presentation is as follows: Asymptomatic lesion detected on prenatal or postnatal ultrasound, intracystic hemorrhage resulting in rapidly enlarging painful mass, portal hypertension, and jaundice because of compression effect and squamous cell carcinoma (3%). While the reported median size at diagnosis is only 3 cm, a rapid increase in size could signify hemorrhage or a cystobiliary communication as in our case.\(^\text{[2,8]}\)

Although preoperative radiology is not pathognomonic, CHFC appears as well-defined anechoic or hypoechoic unilocular solitary cystic lesion, may have irregular margins, typically central (segments 4, 5, and 8) and subcapsular in location with poor enhancement on contrast-enhanced computed tomography scan and a high signal intensity on T2-weighted MRI images. Cyst biliary communication, if suspected, could be documented on a preoperative hepatobiliary scintigraphy.\(^\text{[3,9]}\)

Fine-needle aspiration cytology (FNAC) has been reported to demonstrate pathognomonic ciliated columnar cells in a mucoid background which is specific but not sensitive. Intraoperative frozen section is helpful when CHFC is encountered as an intraoperative enigma as exemplified by our case.\(^\text{[10]}\)

The reported surgical procedures are anatomical liver resection and enucleation. A subtotal cyst excision with a Roux-en-Y biliary-enteric anastomosis was performed in this case because an anatomical liver resection would have been technically precarious on account of complex liver anatomy. Nonetheless, stringent long-term follow-up with serial imaging is planned.\(^\text{[4]}\)

**Conclusion**

This case report is clinically relevant because large CHFC with a cystobiliary communication in pediatric age group is extremely rare and therefore rarely suspected preoperatively. Although rare, one should entertain this differential diagnosis for a unilocular solitary centrally located subcapsular cystic lesion which is characteristically hypoechoic, poorly enhancing, and hyperintense on T2-weighted imaging. A preoperative FNAC could substantiate the diagnosis. Frozen section histopathology is a useful guide when encountered as an intraoperative enigma.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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