



Case report

Laparoscopic excision of a ciliated hepatic foregut cyst in a child: A case report and review of the literature



Nicholas E. Bruns*, Sofya H. Asfaw, Kathryn A. Stackhouse, Gavin A. Falk, David K. Magnuson, Federico G. Seifarth

Department of Pediatric Surgery, Cleveland Clinic Children's Hospital, 9500 Euclid Ave, A120, Cleveland, OH, 44195, USA

HIGHLIGHTS

- Ciliated hepatic foregut cyst (CHFC) is a rare congenital hepatic lesion.
- Indications for excision include large size, symptoms and LFT abnormalities.
- Laparoscopic excision is safe for CHFC and should be considered.

ARTICLE INFO

Article history:

Received 24 August 2015

Received in revised form

27 October 2015

Accepted 27 October 2015

Keywords:

Ciliated hepatic foregut cyst

Hepatic cyst

Laparoscopy

Pediatric

ABSTRACT

Introduction: Ciliated hepatic foregut cysts (CHFC) are rare congenital hepatic lesions derived from the embryonic foregut. Because of potential transformation to squamous cell carcinoma in adulthood, the mainstay of therapy is surgical resection. To our knowledge, we report the first case of CHFC in a child that was successfully excised laparoscopically.

Presentation of case: We report a case of a 4-year-old boy that was diagnosed with an asymptomatic 5-cm liver cyst. After surveillance for 3 years, the cyst grew to 7 cm at which time it was successfully resected laparoscopically. The pathology was consistent with CHFC.

Discussion: There have been few previous reports of CHFCs in children, all of which described excision via a laparotomy. This is the first case report of laparoscopic resection of CHFC in a child.

Conclusion: This case report suggests that laparoscopy may be safe and effective for resection of CHFCs with favorable anatomy such as peripheral location and noninvolvement of key vascular and biliary structures.

© 2015 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Limited. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Ciliated hepatic foregut cysts (CHFC) are rare congenital hepatic lesions derived from the embryonic foregut. Since their description by Friedrich in 1857, there have been approximately 100 reported cases with only 14 cases reported in the pediatric population in English literature [1–6]. CHFCs are often asymptomatic and are frequently discovered incidentally on imaging or during surgical exploration [2]. Due to risk of transformation to squamous cell carcinoma, surgical excision is indicated. To our knowledge, we report the first case of CHFC in a child that was successfully excised laparoscopically.

This work has been reported in line with the CARE criteria for

Abbreviations: CHFC, congenital hepatic foregut cyst; MRI, magnetic resonance imaging; US, ultrasound.

* Corresponding author.

E-mail address: nickebruns@gmail.com (N.E. Bruns).

case reports [7].

2. Presentation of case

A 4-year-old boy presented with a liver cyst discovered incidentally on an abdominal ultrasound (US) performed for uncomplicated congenital hydronephrosis. He had no history of hepatic disease, biliary obstruction, infection or pain. At this time, magnetic resonance imaging (MRI) showed a $5.3 \times 4.8 \times 2.2$ cm minimally complex partially exophytic right hepatic lobe cyst with minimally enhancing septations. Laboratory studies, including a CBC, AFP, PT, hepatic function panel and *Echinococcus* antibody titers, were within normal limits. Surveillance US was performed annually to monitor the cyst size and characteristics. After 3 years the cyst was noted to increase in size. At this time, an MRI demonstrated an interval increase in size to $7.4 \times 6.1 \times 3.0$ cm (Fig. 1). The patient remained asymptomatic.

<http://dx.doi.org/10.1016/j.amsu.2015.10.017>

2049-0801/© 2015 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Limited. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

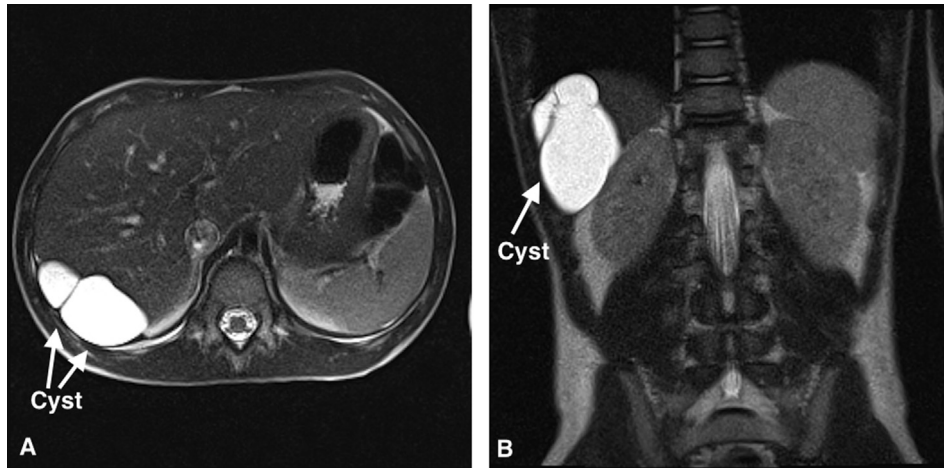


Fig. 1. Preoperative MRI images. T2-weighted axial cross section (A) and coronal cross section (B) MRI shows a 7.4 × 6.1 × 3.0 cm partially exophytic cyst in the right hepatic lobe (Couinaud segment VII) with thin septations.

Due to increasing size, the patient underwent laparoscopic resection of the hepatic cyst. An 11-mm port was placed in the umbilicus to accommodate a 10-mm 30-degree laparoscope. The abdomen was insufflated with carbon dioxide to a pressure of 15 mm Hg. Three additional 5-mm ports were placed, one in the right flank, right lower abdomen and left flank. A 5-mm fan liver retractor was used to reflect the right liver edge. The mass was identified involving the right hemidiaphragm, retroperitoneum and Glisson's capsule overlying segment VII of the liver, compressing and abutting the posterior and lateral aspects of segments VI and VII of the liver without direct involvement of the hepatic parenchyma. Due to the peripheral location of the mass, there was no need for intraoperative ultrasound or vascular control. To completely remove the mass, dissection was carried into the retroperitoneum and diaphragm. Using a combination of Harmonic Scalpel (Ethicon Endo-Surgery, Inc., Cincinnati, Ohio) and electrocautery, the mass was circumscribed and safely resected (Fig. 2). The integrity of the cyst wall had been violated during the dissection, at which point the cyst was aspirated with minimal intra-peritoneal spillage of its contents. The specimen was extracted through the umbilical port. Estimated blood loss was 10 mL.

The patient's postoperative course was uneventful. He was discharged home on postoperative day two. At one-month follow-up, he remained without complications. Microscopic examination was consistent with CHFC without evidence of malignancy.

3. Discussion

CHFCs are rare, typically solitary, unilocular congenital lesions of the liver. They are composed of four layers: ciliated pseudostratified mucin-secreting columnar epithelium, subepithelial loose

connective tissue, an incomplete layer of smooth muscle fibers, and a fibrous outer rim [8]. The presence of ciliated columnar cells in a liver lesion is pathognomonic for CHFC. These cysts are histologically similar to esophageal and bronchogenic cysts, suggesting a common derivation from the embryonic foregut. Esophageal and bronchogenic cysts can be distinguished from CHFCs by the presence of two distinct smooth muscle layers or mural cartilage, respectively [1,9].

CHFCs are most often located in the left lobe of the liver, usually in segment IVb [2,10]. This may be explained by the fact that the left lobe constitutes the majority of the liver during the 4th to 8th weeks of development. Until the 8th week of development, two pleuroperitoneal canals are patent, possibly trapping abnormal foregut buds. In contrast to CHFCs, simple liver cysts are more commonly located in the right hepatic lobe [9].

As in this case report, the most common presentation of a CHFC is an asymptomatic lesion found incidentally on radiographic imaging [9]. As a result, it is difficult to ascertain the true incidence of CHFCs. If symptoms are present, they have been reported to include abdominal pain, nausea, and vomiting [9]. Patients may also present with obstructive jaundice, portal hypertension, and malignancy [2,6,11–13]. In neonates, CHFCs may be detected on antenatal imaging [3]. The differential diagnosis for CHFC includes other unilocular hepatic cysts such as a simple hepatic cyst, parasitic cyst, epidermoid cyst, pyogenic abscess, intrahepatic choledochal cyst, mesenchymal hamartoma, hypovascular solid tumor, and hepatobiliary cystadenoma or cystadenocarcinoma. Imaging alone is non-diagnostic as CHFC is a histologic diagnosis [14]. Hence, cases remain undiagnosed until after aspiration, biopsy or surgical excision.

CHFCs are typically considered benign processes. However, over

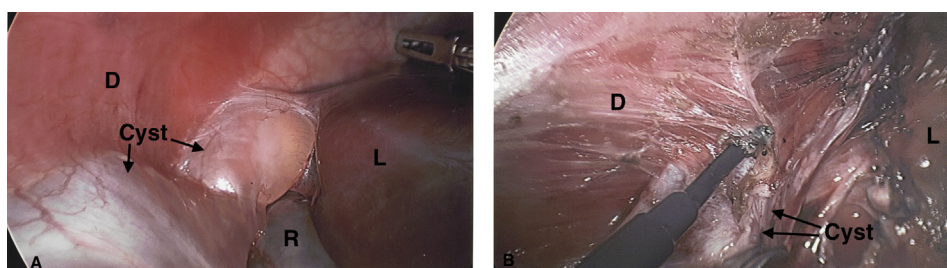


Fig. 2. Laparoscopic images of the dissection. Laparoscopic images of the mass display involvement with the retroperitoneum and right diaphragm (A). The diaphragmatic bed is clear after complete resection (B). L = liver; D = diaphragm; R = retroperitoneum.

the last two decades there have been three reports of malignant transformation to squamous cell carcinoma and extensive squamous metaplasia, resulting in a 4–5% rate of malignancy over that time period [11–13,15,16]. These malignancies were aggressive with reported survival of 2 and 9 months. The presence of dysplasia associated with squamous cell carcinoma may suggest a stepwise progression from non-dysplastic epithelium to dysplasia to carcinoma. The only identified risk factor for malignant transformation in CHFC is size greater than 12 cm [16]. Laboratory markers for malignancy like CA19-9 levels may be misleading, as elevated levels have been associated with benign CHFC [9].

Due to the potential for malignant transformation, most authors agree that surgical resection should be the mainstay of therapy. There has been one case report describing US-guided aspiration followed by 1-year-long event-free observation in a 5-year-old child without long-term follow-up [5]. Suggested indications for surgery include increasing size, size greater than 4 cm, clinical symptoms, unexplained abnormal liver function tests or cyst wall abnormalities on imaging [11,14]. As many CHFCs are not diagnosed until postoperative pathologic evaluation, surgical excision may be diagnostic as well as therapeutic.

Laparoscopy has been adopted for a wide variety of procedures in pediatric surgery due to improved visualization, decreased postoperative pain, quicker recovery and improved cosmetic result [17]. Several reports of laparoscopic excision of a CHFC have been reported in the adult literature [9,15,16,18]. In the pediatric population, there have been two cases describing laparoscopic approaches that were converted to laparotomy, but, to our knowledge, none had been completed laparoscopically prior to this case report [4,6]. Successful laparoscopic resection of hepatic cysts, other than CHFC, in the pediatric population has been described [19,20]. This minimally invasive approach may also be ideal for CHFCs: the typical small size and anterior location allow for easy access. In addition, the generally benign nature allows for removal from the hepatic bed without concern for adequate margins, and the thick cyst wall facilitates handling with laparoscopic instruments [9]. Relative contraindications for laparoscopy may include lesions with central or posterior location or involvement of major biliary or vascular structures as these would be more challenging to remove [18]. Due to the slow progression to malignancy, the procedure should be performed electively under optimal conditions. In neonates with a prenatal or antenatal diagnosis of CHFC, surgical excision can be postponed. In this case, laparoscopy proved to be safe and effective with a short postoperative recovery.

4. Conclusions

Due to the risk of malignancy, CHFCs should be surgically excised for increasing size, clinical symptoms or unexplained abnormal liver function. With proper patient selection, laparoscopic resection can be an advantageous and safe approach to the management of CHFCs in the pediatric population.

Ethical approval

Written informed consent was sought from the parents of this patient for publication of this case report, but the parents could not be located with extensive effort. The content of this manuscript and images are completely anonymized.

Sources of funding

None.

Author contribution

NB, SA, KS and FS were involved with the literature review and gathering and interpreting the patient's clinical information. NB created the figures. All authors were involved with drafting and revising the manuscript. All authors read and approved the final manuscript.

Conflicts of interest

All authors have no conflicts of interest to declare.

Guarantor

Nicholas Bruns and Federico Seifarth accept full responsibility for the work and conduct of the study, had access to the data, and controlled the decision to publish.

References

- [1] S. Kim, F.V. White, W. McAlister, R. Shepherd, G. Mychaliska, Ciliated hepatic foregut cyst in a young child, *J. Pediatr. Surg.* 40 (2005) e51–e53, <http://dx.doi.org/10.1016/j.jpedsurg.2005.07.061>.
- [2] D.J. Vick, Z.D. Goodman, M.T. Deavers, J. Cain, K.G. Ishak, Ciliated hepatic foregut cyst: a study of six cases and review of the literature, *Am. J. Surg. Pathol.* 23 (1999) 671–677.
- [3] F. Guérin, R. Hadhri, M. Fabre, D. Pariente, V. Fouquet, H. Martelli, et al., Prenatal and postnatal ciliated hepatic foregut cysts in infants, *J. Pediatr. Surg.* 45 (2010) e9–e14, <http://dx.doi.org/10.1016/j.jpedsurg.2009.12.009>.
- [4] P. Betalli, D. Gobbi, E. Talenti, R. Alaggio, P. Gamba, G.F. Zanon, Ciliated hepatic foregut cyst: from antenatal diagnosis to surgery, *Pediatr. Radiol.* 38 (2008) 230–232, <http://dx.doi.org/10.1007/s00247-007-0648-1>.
- [5] J. Carnicer, C. Durán, L. Donoso, A. Sáez, A. López, Ciliated hepatic foregut cyst, *J. Pediatr. Gastroenterol. Nutr.* 23 (1996) 191–193.
- [6] V. Zaydfudim, M.J. Rosen, L.A. Gillis, H. Correa, H.N. Lovvorn III, C. Wright Pinson, et al., Ciliated hepatic foregut cysts in children, *Pediatr. Surg. Int.* 26 (2010) 753–757, <http://dx.doi.org/10.1007/s00383-009-2468-x>.
- [7] J.J. Gagnier, G. Kienle, D.G. Altman, D. Moher, H. Sox, D. Riley, et al., The CARE guidelines: consensus-based clinical case reporting guideline development, *J. Med. Case Rep.* 7 (2013) 223, <http://dx.doi.org/10.1186/1752-1947-7-223>.
- [8] B. Bogner, G. Hegedüs, Ciliated hepatic foregut cyst, *Pathol. Oncol. Res.* 8 (2002) 278–279.
- [9] M.D. Goodman, G.Z. Mak, J.P. Reynolds, A.D. Tevar, T.A. Pritts, Laparoscopic excision of a ciliated hepatic foregut cyst, *JSLs* 13 (2009) 96–100.
- [10] T. Horii, Ciliated hepatic foregut cyst a report of one case and a review of the literature, *Hepatol. Res.* 26 (2003) 243–248, [http://dx.doi.org/10.1016/S1386-6346\(03\)00089-5](http://dx.doi.org/10.1016/S1386-6346(03)00089-5).
- [11] D.J. Vick, Z.D. Goodman, K.G. Ishak, Squamous cell carcinoma arising in a ciliated hepatic foregut cyst, *Arch. Pathol. Lab. Med.* 123 (1999) 1115–1117.
- [12] A.-S. de Lajarte-Thirouard, N. Rioux-Leclercq, K. Boudjema, Y. Gandon, M.-P. Ramée, B. Turlin, Squamous cell carcinoma arising in a hepatic foregut cyst, *Pathol. Res. Pract.* 198 (2002) 697–700.
- [13] A. Furlanetto, A.P. Dei Tos, Squamous cell carcinoma arising in a ciliated hepatic foregut cyst, *Virchows Arch. Int. J. Pathol.* 441 (2002) 296–298, <http://dx.doi.org/10.1007/s00428-002-0668-z>.
- [14] M. Boumoud, A. Daghfous, H. Maghrebi, S. Gharbi, S. Ayadi, L. Bouallegue, et al., Imaging features of ciliated hepatic foregut cyst, *Diag. Interv. Imaging* 96 (2015) 301–303, <http://dx.doi.org/10.1016/j.diii.2013.07.001>.
- [15] N. Ben Mena, S. Zalinski, M. Svrcek, M. Lewin, J.F. Fléjou, D. Wendum, et al., Ciliated hepatic foregut cyst with extensive squamous metaplasia: report of a case, *Virchows Arch. Int. J. Pathol.* 449 (2006) 730–733, <http://dx.doi.org/10.1007/s00428-006-0320-4>.
- [16] J.D. Jakowski, J.G. Lucas, S. Seth, W.L. Frankel, Ciliated hepatic foregut cyst: A rare but increasingly reported liver cyst, *Ann. Diag. Pathol.* 8 (2004) 342–346, <http://dx.doi.org/10.1053/j.anndiagpath.2004.08.004>.
- [17] P. Mattei, Minimally invasive surgery in the diagnosis and treatment of abdominal pain in children, *Curr. Opin. Pediatr.* 19 (2007) 338–343.
- [18] J. Saravanan, G. Manoharan, S. Jeswanth, P. Ravichandran, Laparoscopic excision of large ciliated hepatic foregut cyst, *J. Minim. Access Surg.* 10 (2014) 151–153, <http://dx.doi.org/10.4103/0972-9941.134879>.
- [19] T. Tuxun, J.-H. Zhang, J.-M. Zhao, Q.-W. Tai, M. Abudurexti, H.-Z. Ma, et al., World review of laparoscopic treatment of liver cystic echinococcosis—914 patients, *Int. J. Infect. Dis. IJID Off. Publ. Int. Soc. Infect. Dis.* 24 (2014) 43–50, <http://dx.doi.org/10.1016/j.ijid.2014.01.012>.
- [20] P. Tabrizian, P.S. Midulla, Laparoscopic excision of a large hepatic cyst, *JSLs J. Soc. Laparoendosc. Surg. Soc. Laparoendosc. Surg.* 14 (2010) 272–274, <http://dx.doi.org/10.4293/108680810X12785289144638>.