CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 5 (2014) 919–923

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Multicystic biliary hamartoma: A report of a rare entity and a review of the literature

Rachel E. Beard[a,*], Eric U. Yee[b], Koenraad J. Mortele[c], Khalid Khwaja[a]

[a] Department of Surgery, Beth Israel Deaconess Medical Center, 110 Francis Street, Boston, MA 02215, United States
[b] Department of Pathology, Beth Israel Deaconess Medical Center, 330 Brookline Avenue, Boston, MA 02215, United States
[c] Department of Radiology, Beth Israel Deaconess Medical Center, 330 Brookline Avenue, Boston, MA 02215, United States

ARTICLE INFO

Article history:
Received 10 April 2014
Received in revised form 9 September 2014
Accepted 8 October 2014
Available online 17 October 2014

Keywords:
Hamartoma
Biliary neoplasm
Periductal
Cystic
Honeycomb

ABSTRACT

INTRODUCTION: Multicystic biliary hamartoma is a rare liver tumor that was first described in 2005. Only nine cases are reported in the literature and all of them originate from Eastern patient populations, specifically Japan and Korea.
PRESENTATION OF CASE: Herein we report the occurrence of the tenth multicystic biliary hamartoma reported to date, arising in a Caucasian American woman initially presenting with abdominal pain. At 4.7 cm this is the second largest tumor reported to date and the only one arising in a Western patient population.
DISCUSSION: The patient underwent multimodality imaging and the tumor was biopsied preoperatively, but the diagnosis remained unclear. An extended right hepatectomy was performed for resection of her tumor, and the tumor was definitively diagnosed based on the surgically resected specimen. As all nine of the previously reported cases also underwent resection, the natural history of this lesion remains unknown. The lack of both recurrence and tumor spread in the previously reported cases indicates that this may be a benign lesion not requiring surgical resection unless symptomatic.
CONCLUSION: Multicystic biliary hamartoma is an extremely rare tumor. Increased awareness of the radiologic and pathologic features will likely lead to the diagnoses of further cases in both Western and Eastern populations and could potentially assist with preoperative diagnosis. The natural history and optimal management of this tumor remain uncertain.

1. Introduction

Multicystic biliary hamartoma (MBH) is a rare liver lesion that has been described in the last decade as a distinct entity from other previously classified hepatobiliary cystic lesions.[1] There are currently nine cases reported in the literature, with all previous reports originating from Japan or Korea (Table 1).[1–5] Herein we report a case of a MBH occurring in a Caucasian woman presenting with abdominal pain. At 4.7 cm it is the second largest lesion reported in the literature, as well as the only one reported from a Western population to date.

2. Presentation of case

In January 2014, we evaluated a 48-year-old woman for a newly diagnosed right liver mass. She was symptomatic with epigastric and right upper quadrant abdominal pain. Her past medical history was remarkable for recently diagnosed hepatitis C. She had undergone an open cholecystectomy 20 years previously for biliary colic and a hysterectomy 10 years earlier for abnormal uterine bleeding. She also reported that her paternal aunt had passed away in the 1980s from a very rare liver tumor.

Her workup prior to presentation at our hospital included an abdominal ultrasound demonstrating an echogenic 5.7 cm mass in the right lobe of the liver. CT scan showed a 5.6 × 3.4 cm lobulated subcapsular mass in segment 8 (Fig. 1). Two additional subcentimeter cystic lesions, consistent with microhamartomas, were noted in segment 7 (Fig. 1b). MRI highlighted its tubulocystic composition and intermingled normal hepatic tissue (Fig. 2). She had undergone a needle biopsy of this lesion at an outside hospital that on microscopic exam demonstrated thick, dense fibrous tissue containing cytologically bland, large caliber bile ducts with intermingled benign hepatocytes (Fig. 4c). Given the multicystic nature on imaging and microscopic findings, this was initially diagnosed as a possible biliary adenofibroma and the patient was referred to our institution for surgical evaluation. Complete laboratory

* Corresponding author. Tel.: +1 253 279 7046; fax: +1 617 632 7424.
E-mail address: rbeard@bidmc.harvard.edu (R.E. Beard).

http://dx.doi.org/10.1016/j.ijscr.2014.10.014

Table 1
Multicystic biliary hamartomas cases reported to date.

Reference	Country of origin	Cases reported	Largest tumor measurement (cm)	Patients' ages and genders	Surgical resection performed	Patient presentation	Co-existing liver disease
Song et al. [2]	Korea	1	2.7	52M	Not stated	Abdominal pain	None
Ryu et al.[a] [1]	Japan	3	2.0–3.5[b]	45M	Partial resection	Incidental finding on US	None
				58M	Partial resection	for routine checkup	None
				55F	Lateral segmentectomy	Incidental finding on US for routine checkup Elevated LFTs	None
Kai et al. [3]	Japan	1	5.0	55M	Partial resection of posterior segment	Finding on US during workup for Hepatitis B	Hepatitis B
Zen et al. [4]	Japan	3	1.8	59M	Left hepatectomy	Right-sided abdominal	None
			2.8	69F	Resection of medial segment	pain	HCV cirrhosis
			4.2	70F	Left segmentectomy	Incidental on imaging Elevated LFTs	None
Kobayashi et al. [5]	Japan	1	3.6	30M	Partial hepatectomy	Incidental finding on imaging	None

[a] Ryu et al. actually reported the imaging findings of four cases, however one of the cases, that of the 70-year-old female, had been previously reported in the group's 2006 paper, Zen et al., so only the other three cases are summarized in this table.
[b] A range of tumor sizes was provided in this paper but specific measurements of each tumor were not.

studies including CBC, LFTs, CA19-9, CEA, AFP and hepatitis serologies were all within normal limits, aside from a HCV antibody that was positive. HCV RNA and viral load were non-detectable.

Given the possible diagnosis of biliary adenomafibroma and the potential for malignancy, the patient underwent an extended right hepatectomy. Intraoperative ultrasound demonstrated tumor extension into segment 4A and involved part of the middle hepatic vein. During the dissection, note was made of a sizeable replaced left hepatic artery and a portal vein that lacked a right common trunk but instead gave direct rise to the right anterior and posterior branches at the bifurcation. The patient recovered without complication and was discharged from the hospital the following week. She is now eight months post-surgery and has yet to undergo any repeat abdominal imaging.

On pathologic examination, the lesion measured $4.7 \times 4.3 \times 4.0$ cm and consisted of a subcapsular, intrahepatic, well-circumscribed mass with solid and cystic cut surfaces (Fig. 3). Microscopic exam revealed large bile ducts with varying degrees of cystic dilation, periductal glands surrounded by fibrous tissue with interspersed islands of benign hepatocytes, and bile-like material observed within some of the large ducts (Fig 4b and d). These features were consistent with a diagnosis of MBH.[1,4] The background non-lesional liver demonstrated only mild macrovesicular steatosis without significant fibrosis.

3. Discussion

The diagnosis of MBH proved challenging, as demonstrated by the inability to preoperatively diagnose this lesion based on the radiographic studies and needle biopsies. In a series of four cases diagnosed by examination of surgically resected specimens from 1998 to 2007, two cases were originally diagnosed as unusual hepatobiliary hamartomas; only after retrospective review, in light of the original descriptions of the histologic findings published in 2006, were these diagnoses corrected.[1] To our knowledge, our case is the first reported that demonstrates findings from preoperative needle core biopsies.

The nine previously reported cases in the literature all originated from Eastern patient populations, but beyond that commonality the patients' characteristics and initial clinical presentations varied widely (Table 1). Patients' ages at time of diagnosis ranged

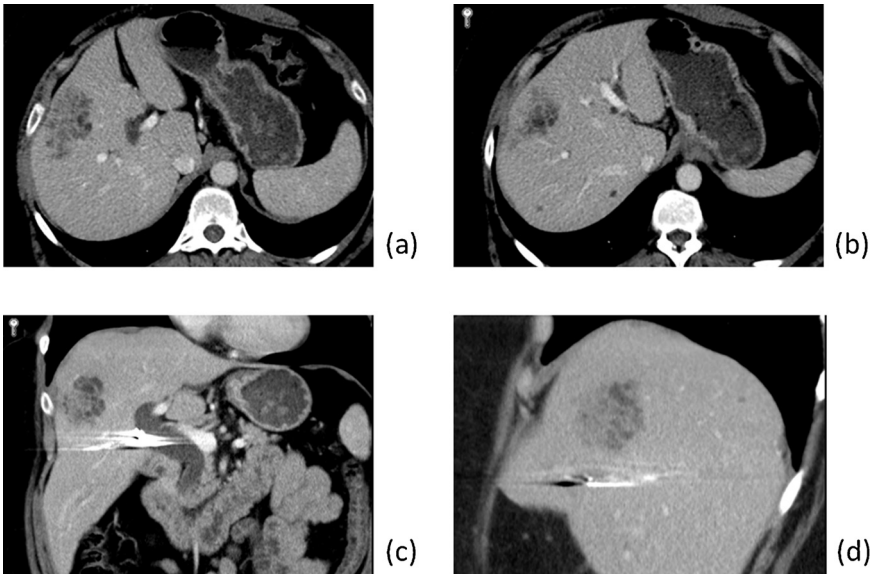

Fig. 1. Axial (a and b), coronal (c) and sagittal (d) contrast-enhanced CT images through the liver demonstrate a 5.6×3.4 cm ill-defined lobulated mass located subcapsular in segment 8 of the liver. The lesion is predominantly composed of tubulocystic structures intermingled with strands of hepatic parenchyma. Two small additional microhamartomas are identified in segment 7 (b).