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ORIGINAL ARTICLE – HEPATOBILIARY TUMORS

Total Laparoscopic Resection of an Extrahepatic Mucinous Biliary Cystadenoma with Liver Involvement (with Video)

Huanwei Chen[1,2], Fa Luo, MD[1,2], Ying Liu, MD[1], Fengjie Wang, MD[1], Qiucheng Lei, MD[1], Wai I. Ho, PhD[1], and Wan Yee Lau[1,3]

[1]Department of Liver and Pancreas Surgery, The First People's Hospital of Foshan, Foshan, China; [2]Guangdong Medical University, Zhan Jiang, China; [3]Faculty of Medicine, The Chinese University of Hong Kong, Shatin, New Territories, Hong Kong SAR, China

ABSTRACT

Background. Intrahepatic mucinous biliary cystadenoma is rare, and extrahepatic MBC is even rarer. To our knowledge, total laparoscopic resection of an extrahepatic MBC that had extended intrahepatically has never been reported.

Patients and Methods. A 28-year-old female presented to our hospital with upper abdomen pain. Radiological investigations demonstrated a 7-cm multiloculated cystic lesion arising from the left hepatic bile duct extending to involve the extrahepatic biliary system down to and posterior to the back of the head of pancreas. The entire extrahepatic bile duct was involved, except for the gallbladder. Laparoscopic surgery was carried out using a five-port approach. A gourd-shaped well-defined multiloculated cyst was found extending from the extrahepatic biliary system proximally to involve the left hepatic duct intrahepatically. After cholecystectomy, the gourd-shaped cyst was opened at its narrowest part at the hepatic hilus to facilitate subsequent resectional surgery. The distal sac was dissected to the distal bile duct end at the duodenal wall and transected. The proximal sac was dissected and resected en bloc with the bifurcation of the right/left hepatic ducts, combined with left hepatectomy plus caudate lobectomy. The reconstruction was done by anastomosing the right anterior and posterior sectional bile ducts to a Roux-en-Y jejunal loop. Multiple intraoperative frozen sections demonstrated the lesion to be a benign MBC.

Results. The patient was discharged home 12 days after surgery. She was well on follow-up 24 months after surgery.

Conclusion. Total laparoscopic resection is technically feasible to treat an extrahepatic MBC with intrahepatic extension.

Mucinous biliary cystadenoma (MBC) is rare, and in the majority of cases, the lesions are within the liver. Over 170 years have elapsed since the first report on this lesion by Henter and colleagues in 1887.[1] Up to now, less than 500 cases have been reported in the medical literature.[2–4]

Extrahepatic MBC is extremely rare. Of the 70 cases of MBC reported by Devaney and colleagues, there were only 9 cases of extrahepatic MBC.[5] Of the 15 cases of MBC reported by Lewis and colleagues, there was only 1 case of extrahepatic MBC.[6] However, there are still occasional isolated case reports on this pathology.[7,8] It is important to distinguish extrahepatic MBC from the less rare intrahepatic MBC with extension of the lesion to involve the extrahepatic biliary system.[9–12] In this current case report, the main tumor arose from the extrahepatic biliary system with extension to involve the intrahepatic left duct. Thus, this is a very rare tumor with an uncommon extensive involvement that was still surgically resectable.

The treatment of MBC, irrespective of whether it is intrahepatic, extrahepatic, or both extra- and intrahepatic, is complete surgical resection if technically feasible because of the potential of this tumor to become malignant.

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H. Chen
e-mail: chwei_fsyyy@163.com

W. Y. Lau
e-mail: josephlau@cuhk.edu.hk

To our knowledge, this is the first reported case using totally laparoscopic surgery to resect an extrahepatic MBC that had extended intrahepatically into the left duct.

PATIENTS AND METHODS

A 28-year-old Asian female presented with upper abdominal distending pain. Abdominal ultrasound showed a large multicystic lesion in the head of pancreas. Abdominal computed tomography showed a multiloculated cyst, 7.0 × 6.4 × 4.1 cm, extending from the left hepatic duct to involve the extrahepatic biliary system, posteriorly to the head of pancreas, giving a differential diagnosis of intraductal papillary mucinous neoplasm and pancreatic mucinous cystadenoma (Fig. 1A, D). Magnetic resonance cholangiopancreatography (MRCP) showed the lesion to be connected to the common bile duct, suggesting a diagnosis of choledochel cyst, or a pancreatic cystadenocarcinoma with invasion into the biliary system (Fig. 2). Laboratory blood tests showed normal liver functions and a CA 19-9 level of 55.12 U/ml, and the indocyanine green retention at 15 min was 2.8%. Three-

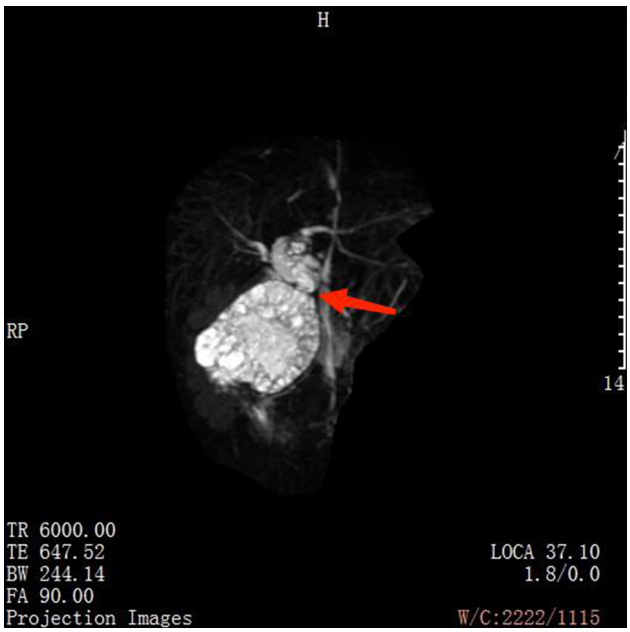

FIG. 2 Preoperative MRCP showing a gourd-shaped multiloculated cyst

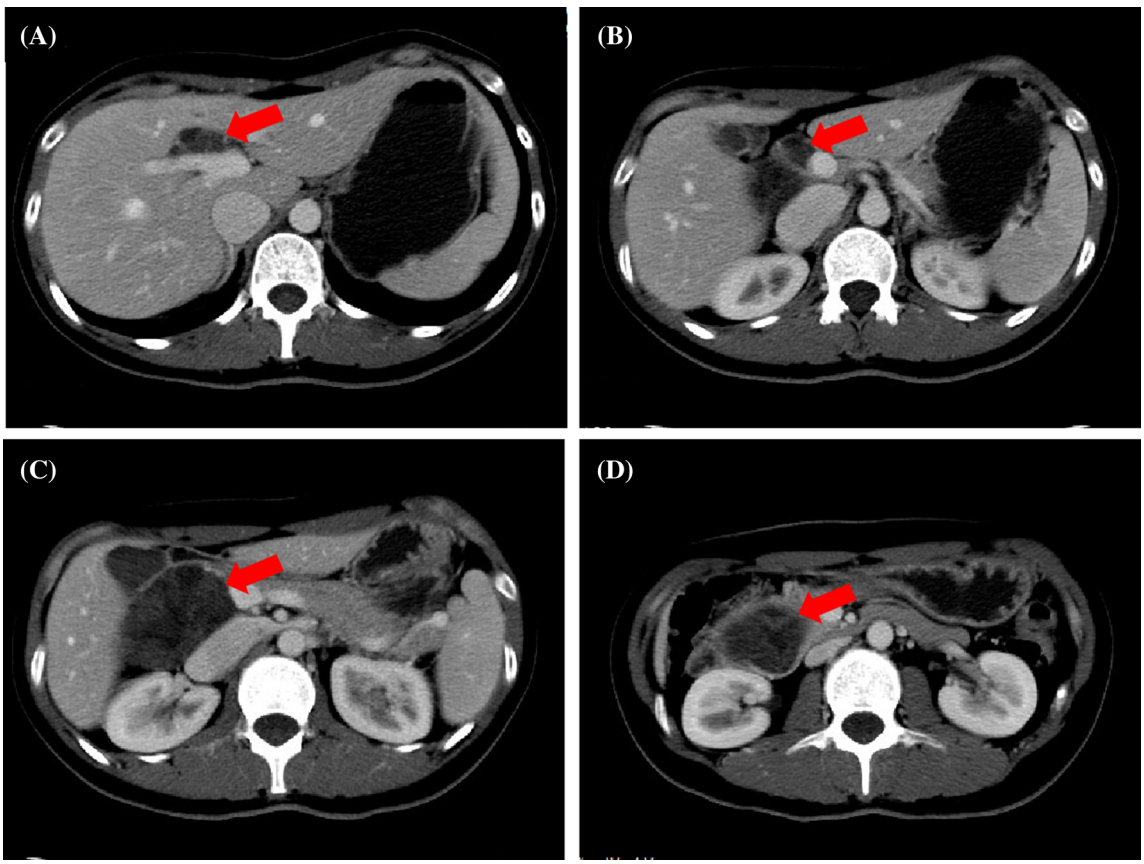

FIG. 1 Preoperative portal phase of CT showing a multiloculatel cyst from the hepatic hilus to pancreatic head region, with a thick wall and honeycomb shape, 7.0 × 6.4 × 4.1 cm in size, involving the left hepatic duct

dimensional computed tomography (CT) reconstruction showed the right hepatic artery to arise from the superior mesenteric artery. The tumor was close to the middle and left hepatic arteries and the common bile duct. The lesion had extended from the common bile duct to involve the bifurcation of the right and left hepatic ducts and then intrahepatically into the left hepatic duct. The distal part of the tumor extended inferiorly to the descending duodenum, without involving the duodenal wall (Fig. 3A, D).

Approval was obtained from our hospital institutional review board to carry out laparoscopic resection of extrahepatic MBC, and informed consent was obtained from the patient and her relatives after a very detailed discussion was carried out between them and the surgical team. Laparoscopic surgery was carried out using a five-port approach (Fig. 4). The tumor was found intraoperatively to involve the extrahepatic biliary system with intrahepatic extension into the left duct. The gallbladder was not involved by the tumor, and there was no evidence to suggest that the tumor extended out of the bile duct walls. After cholecystectomy, kocherization, and division of the right gastric artery, the gastroduodenal artery (which derived from the common hepatic artery), the proper hepatic artery (which derived from the superior mesenteric artery), and the left hepatic artery were identified (Fig. 5A). The cyst was opened at its narrowest part at the hepatic hilus. Multiple frozen sections established the diagnosis and benignity of the lesion. The cyst was transected at its narrowest part to facilitate subsequent resectional surgery.

The distal cyst was dissected close to the bile duct wall to the point where the common bile duct entered the second part of duodenum. It was then transected.

The proximal cyst was dissected in a cranial direction, ending with en bloc resection of the common hepatic duct, bifurcation of right/left hepatic ducts, followed by combined left hepatectomy with caudate lobectomy (Fig. 5B, E).

After achieving hemostasis and removal of the specimens with specimen bags through a small incision in the midline of upper abdomen, the right anterior and posterior sectional bile ducts were anastomosed to a Roux-en-Y jejunal loop (Fig. 5E), and two plastic stents were used to stent the anastomoses, followed by a side-to-side

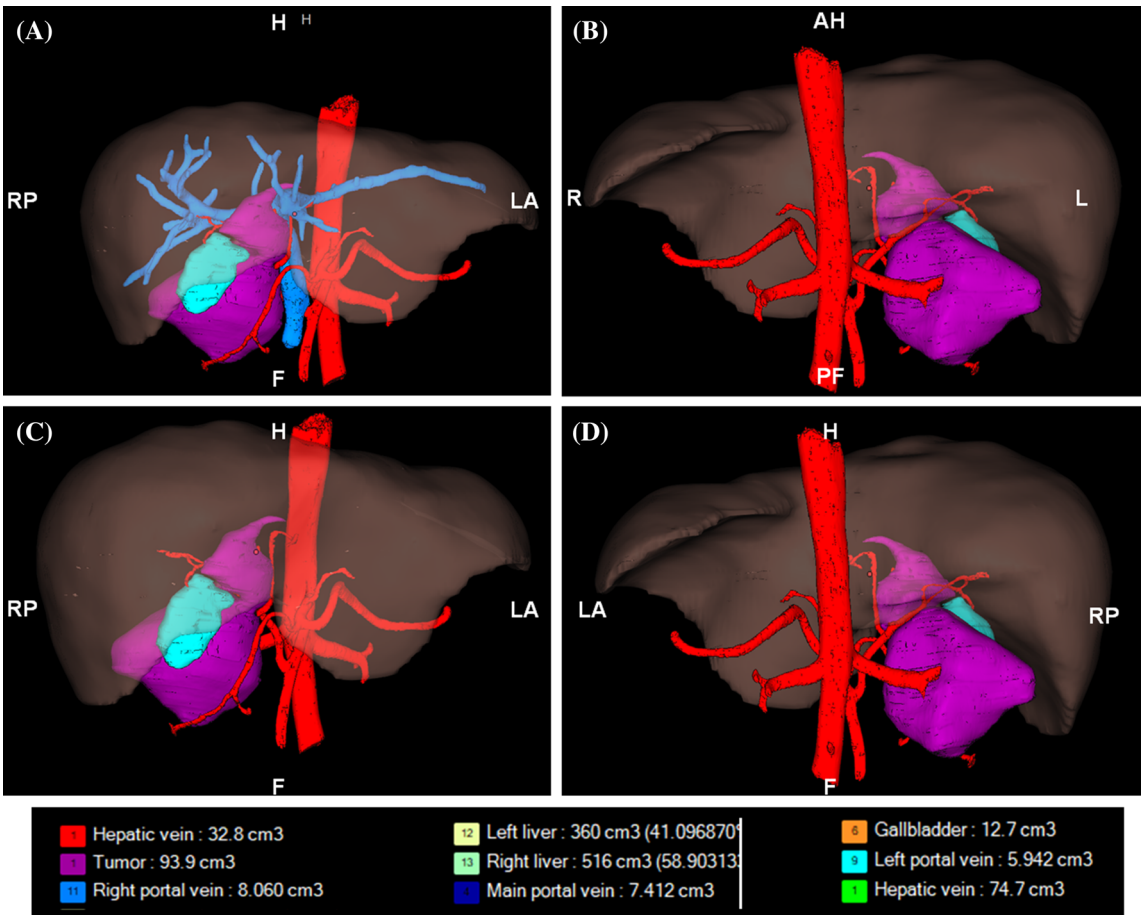

FIG. 3 Preoperative three-dimensional CT reconstruction showing the extent of the lesion and its anatomical relationship with the hepatic arteries and portal vein

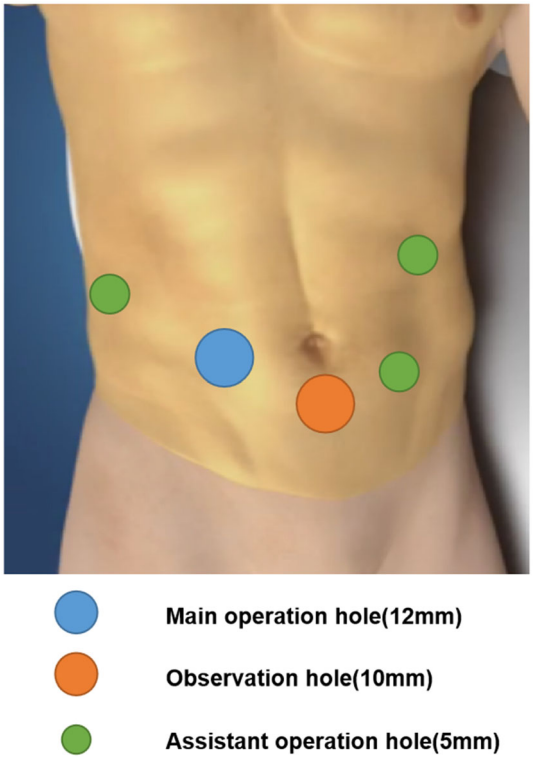

FIG. 4 Trocars' layout

jejunojejunostomy (Fig. 5F). The resected specimens are shown in Fig. 6A, B. The operation ended with drainage tubes put on the two sides of the dissected area.

The operation took 540 min. The blood loss was 550 ml. There was no need for any blood transfusion. Except for postoperative ileus, which required nasogastric intubation for 3 days, there were no other significant complications, including bleeding and biliary fistula. The patient was discharged home 12 days after surgery. She was well on follow-up 24 months after surgery.

DISCUSSION

An accurate preoperative diagnosis of MBC can be difficult because of its rarity, and the lesion can be misdiagnosed as other more common types of liver cysts. MBC can be seen at any age, with the majority of patients being female and more than half of them presenting with symptoms, the most common of which is pain. However, patients can present with abdominal masses, abnormal liver function tests, jaundice, fever, and weight loss,[1–6,9,12,13] Most MBCs present as multiloculated cysts, rarely as uniloculated cysts. Intrahepatic MBCs are much more common than extrahepatic MBCs. For intrahepatic MBCs, the incidences of the lesion in the left or the right liver are almost equal. The levels of CA 19-9 in serum or cyst fluid are usually high, with significantly higher CA 19-9 levels in cyst fluids than in serum.[14] CT imaging usually shows a solitary multiloculated cyst in the liver. Within the loculated chambers, nodules, papillary protrusions, and calcification of the cyst wall may be seen with different degrees of contrast enhancements. Magnetic resonance can

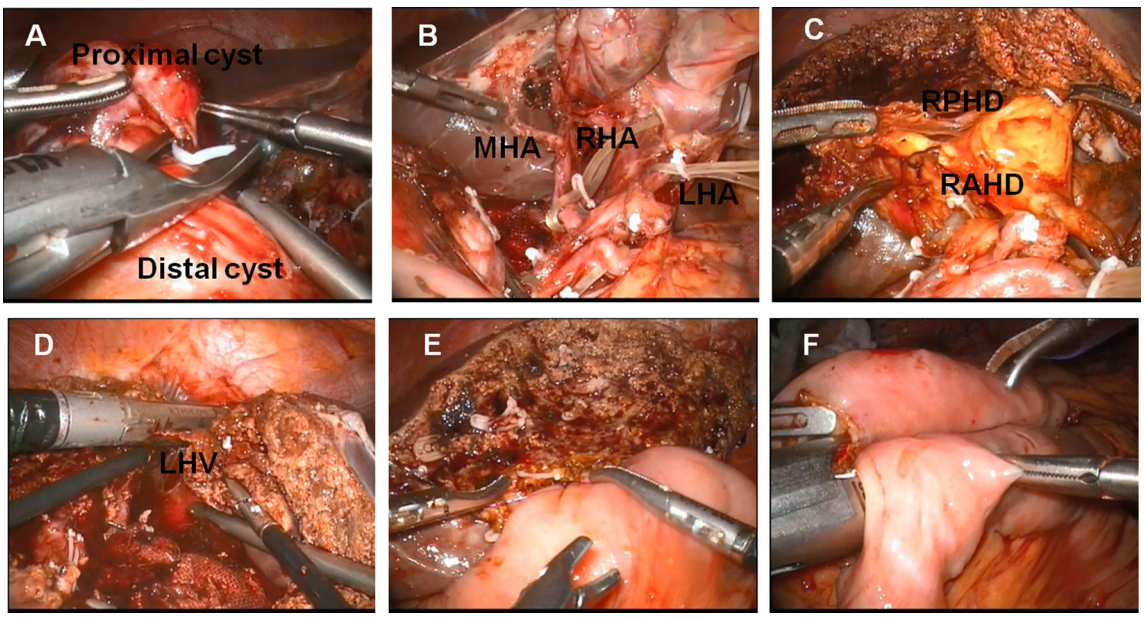

FIG. 5 The main steps of the operation: **A** the cyst was transected at its narrowest part; **B** showing the left hepatic artery behind the tumor and the anomalous right hepatic artery, the middle hepatic artery had been severed; **C** disconnecting the right anterior and right posterior bile ducts; **D** left hepatectomy and caudate lobectomy; **E** right anterior and posterior bile ducts anastomosed to jejunum; **F** side-to-side jejunojejunostomy

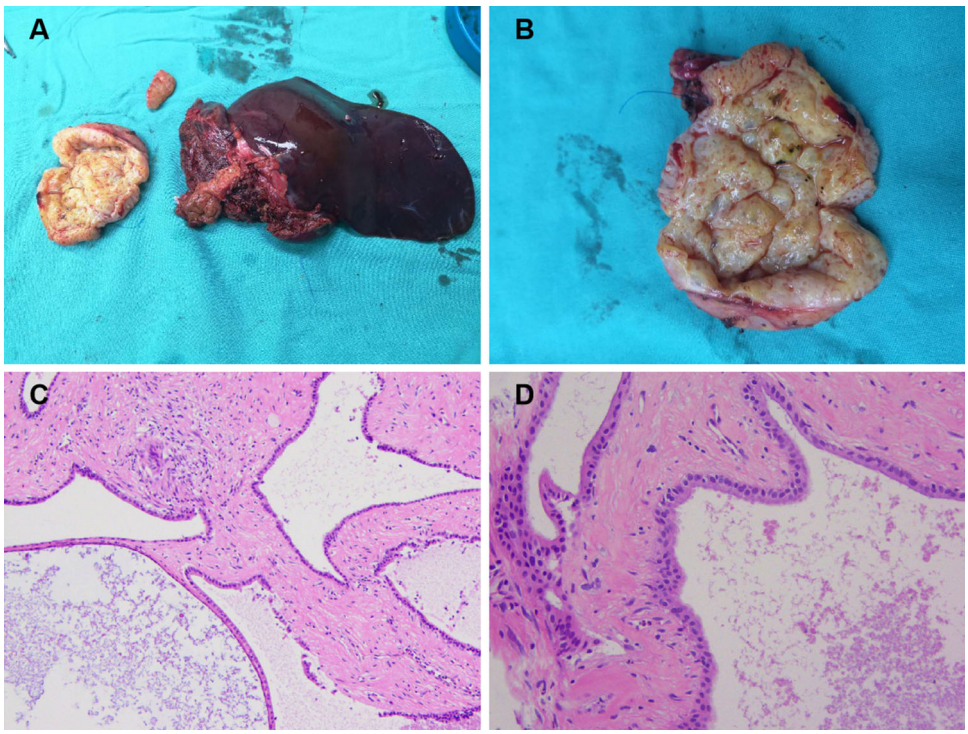

FIG. 6 Macroscopic and microscopic findings of the resected specimens: **A** proximal specimen with the proximal part of the cyst en bloc with left hepatectomy and caudate lobectomy; **B** distal specimen of the resected cyst. Histopathology study confirmed the lesion to be a biliary mucinous cystadenoma. **C** Low-power field and **D** high-power field

show the relations between tumor and blood vessels/bile ducts more clearly to provide more information to surgeons to determine the most appropriate surgical treatment.[15] MRCP shows the biliary system even more clearly to allow reliable surgical planning. Many treatments have been used to treat MBC, including cyst puncture and aspiration, injection of sclerosing agents, and laparoscopic fenestration. However, current opinion is that complete surgical removal of tumor is the only curative treatment for MBC, as this is a neoplastic lesion.[14,16,17] Furthermore, as MBC is a benign tumor, there is no need to achieve a wide resection margin for intrahepatic MBC,[18] and a resection margin is impossible for extrahepatic MBC, as in this case report, since the extrahepatic bile ducts containing the MBC were surrounded by vital vascular structures that cannot be sacrificed. However, for intrahepatic mucinous cystadenocarcinoma, there are still controversies on whether the liver resection margins should be 1 cm or 2 cm.[4,19,20]

CONCLUSION

Extrahepatic MBC is rare. This is the first report on a successful total laparoscopic resection in a patient with extrahepatic MBC that had extended intrahepatically to involve the left hepatic duct.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1245/s10434-022-12150-7.

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DISCLOSURE Huanwei Chen, Fa Luo, Ying Liu, Fengjie Wang, Qiucheng Lei, Wai I Ho, and Wan Yee Lau have no conflicts of interest or financial ties to disclose.

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