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Case Report
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A Case Report and Literature Review: Intrahepatic Multicystic Biliary Hamartoma

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Abstract

Background: Multicystic Biliary Hamartoma (MCHB) is a rare hamartomatous nodule of the liver, which has recently been described as a new category of hepatic nodular cystic lesion. Most of them are clinically benign. The imaging findings are similar to those of many other hepatic cystic lesions, but MCBH also has some notable features, such as large cysts, smooth cyst walls, and lack of communication with the hepatic duct. Due to the non-specific radiology, preoperative diagnosis is difficult, and is usually diagnosed by postoperative pathology. Complete resection is the best treatment option, and the postoperative prognosis is good.

Case Report: This article reports a case of a patient with MCBH. The main points of the patient were the patient had no other obvious symptoms except abdominal distension, presenting as a space-occupying lesion. The Computed Tomography (CT) scan found liver lesion. The patient underwent laparoscopic resection of the right liver tumor. The postoperative pathology and immunohistochemistry were both diagnosed as MCBH. The cyst was excised and the patient was treated with anti-infection, liver care, analgesia, nutritional support and dressing change.

Conclusion: When the patients have MCBH, they require a combination of imaging and pathology for diagnosis. Under normal circumstances, the prognosis of MCBH is good. When we find abdominal distension, regardless of age, we should go to the hospital for related auxiliary examination to determine the cause, and give treatment to prevent the disease from developing and worsening.

Keywords: Liver; Bile duct; Multicystic biliary hamartoma; Immunohistochemistry

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Introduction

Bile duct hamartoma was described for the first time by von Meyenburg in a pathology report. Subsequently, it was depicted on contrast-enhanced magnetic resonance imaging. In 1975, McLoughlin and Phillips first described its angiographic characteristics. Later, others described the Computed Tomography (CT) and ultrasound characteristics of MCBH [2]. As for the etiology of MCBH, biliary plate malformations may be one of the pathological causes, which can be ascribed to abnormal embryogenesis of the biliary ductal system. As a type of fibrous polycystic liver diseases, MCBH needs to be differentiated from the others include congenital liver fibrosis, autosomal dominant polycystic liver disease, Caroli's disease and choledochal cyst. It is worthy to make an early diagnosis and differential diagnosis of MCBH.

However, only a few cases have been published in the world literature [1]. The previous literatures have never reported patient aged lower than 30 years old with MCBH, nevertheless, the age of this case is only 14 years old, and apart from this, the large size of the cyst is unprecedented. Here we describe this unique case report and the literature review of MCBH.

Case Presentation

A 14-year-old boy experienced upper abdominal distension. Physical examination showed abnormal lumps in the upper abdomen. Further consultation and physical examination revealed no evidence of the following: Headache and dizziness, dyspnea and shortness of breath, diarrhea and melena, and frequent and/or urgent micturition. No relevant past interventions had been carried out. No significant clinical or family history or relevant genetic information. An abdominal

Table 1: Summary of reports of the case worldwide.

Case No.	Authors	Patients' age (years)/Gender	Location	Size (cm)	Treatment
1	Kobayashi et al. [4]	30/M	Seg. VI	3.6	Partial resection
2	Zen et al. [5]	59/M	Seg. IV	4.2	Left hepatectomy
3		70/F	Seg. III	1.8	Segmentectomy
4		69/F	Seg. III	2.8	Segmentectomy
5	Kai et al. [6]	55/M	Seg. IV	5	Partial resection
6	Rye et al [11]	45/M	Seg. VII	2.0-3.5 (Case nos 6-8)	Partial resection
7		58/M	Seg. III		Partial resection
8		55/F	Seg. VI, VII		Partial resection
9	Song et al. [1]	52/M	Seg. III	2.7	Partial resection
10	Beard et al. [8]	48/F	Seg. VII	4.7	Extended right hepatectomy
11	Yoh et al. [9]	69/M	Seg. III	3	Left hepatectomy
12	Fernandez-Carrion et al. [10]	60/F	Seg. VI	5	Partial hepatectomy
13	Tominaga et al. [7]	26/M	Seg. V, VI	10	Right hepatectomy
14	Morinaga et al. [12]	53/M	left lobe of the liver	12	Left hepatectomy
15	Ogura et al. [13]	77/F	Seg. III	12	Partial hepatectomy
16	Wentao et al. [14]	37/M	Seg. VI	8	Laparoscopic Partial Resection
Present case		14/M	Seg. III	17	Laparoscopic Partial Resection

M: Male; F: Female

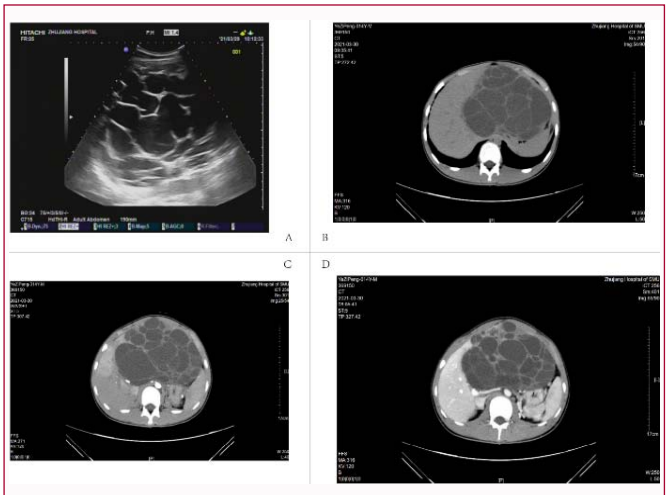

Figure 1: (A) Liver ultrasound; (B) CT plain scan; (C) The arterial phase of enhanced CT scans; (D) The portal phase of enhanced CT scans.

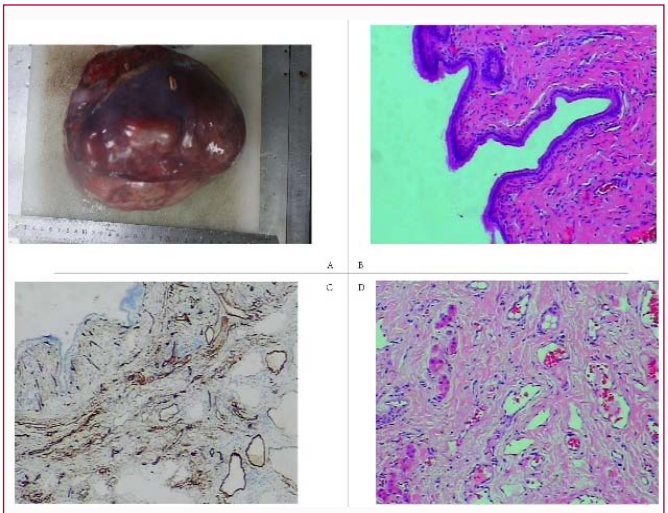

Figure 2: (A) Gross specimen of right liver tumor; (B) Microscopic image of right liver tumor; (C) Immunohistochemical staining of cyst gland duct epithelium (CK+); (D) Immunohistochemical vascular staining (CD34+).

Computed Tomography (CT) (Figure 1) showed a large, cystic, space-occupying lesion, with blurred outline with respect to adjacent liver structures. Because of this lesion, he was admitted for further treatment. No abnormality was observed via routine thoracic plain CT. The patient underwent several laboratory tests, which posed a further burden on the family's finances. During the present admission, the following results of laboratory tests were recorded: Total Bilirubin (TBil), 9.7 μmol/L; direct bilirubin, 4.5 μmol/L; Albumin (Alb), 39.9 g/L; Alanine Aminotransferase (ALT), 8 IU/L; Aspartate Aminotransferase (AST), 12 IU/L; Alpha-Fetoprotein (AFP), 1.2 μg/L; HBsAb, 24.100 IU/L; HBeAb, 1.590COI; and HBcAb, 2.360COI. Further CT imaging revealed a multicystic lesion located in the upper abdomen, suggestive of a cystadenoma or lymphangioma. Enhanced CT indicated asymmetrical intensification in the right lobe during the arterial phase, and demonstrated homogeneous enhancement in the right lobe during the portal vein phase and delayed phase, which was considered to be attributed to abnormal perfusion. At ultrasonography, the lesion appeared as a polycystic mass with unclear origin and needed further confirmation.

After admission, the patient underwent liver biopsy, and pathological assessment revealed hamartoma. On further examination, no other lesions were found. After discussion, we advised surgical treatment for the patient. The patient and his legal guardians provided assent and consent, respectively, for the operation, with the aim of removing the tumor, performing drainage, and enhancing the quality of life. After routine preoperative preparation and anesthesia, a trocar was placed to explore the abdominal cavity. After removal of the tumor by ultrasonic knife, clear cystic fluid spurted out and subsequently, the tumor slightly collapsed. Because the tumor was too large to be clearly exposed, we decided to perform

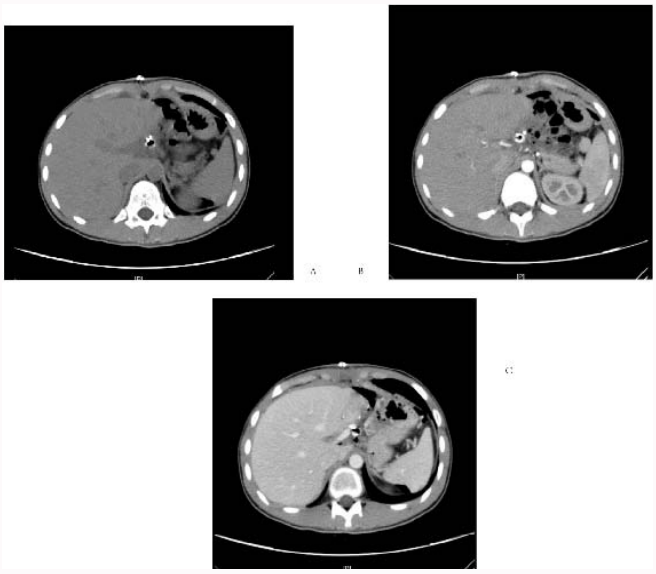

Figure 3: (A) CT plain scan after operation; (B) The arterial phase of enhanced CT scan after operation; (C) The portal phase of enhanced CT scan after operation.

hepatic left lateral lobectomy and enterolysis, and an in-dwelling drainage tube was placed. The size of the resected tumor was 17 cm × 16 cm × 15 cm. Postoperatively, a definitive diagnosis of polycystic bile duct hamartoma was established based on paraffin section analysis and immunohistochemical analysis (Figure 2, 3). The patient received postoperative antibiotic, hepatoprotective, and analgesic therapy, along with nutritional support and wound dressing. The postoperative recovery was good, and the CT reexamination showed good prognosis.

Discussion

MCBH is a rare benign fibrocystic liver disease, which may be related to the malformations of the bile duct plate [3]. Currently, there are very limited published reports of MCBH worldwide. Since the first description of MCBHs in 2005, there have only been 16 published cases worldwide (Table 1). MCBHs can occur at any age, although it is more common in patients aged 30 to 70 years, and is two times more likely to affect men than women, with a male-to-female ratio of 10:6. Of note, compared with the former cases, the age is younger and the size is larger of the patients with MCBH in this report, revealing the importance of considering diagnosis of MCBH in suspected cases in lower age groups.

Most of the patients come to see a doctor complain of abdominal pain, while some of them have no obvious clinical manifestations, and their cysts are usually discovered incidentally. Also, some may suffer from obstructive jaundice and abdominal discomfort. The patient in this case came to our hospital because of abdominal distension, without any specific symptom. Consequently, patients should be diagnosed after auxiliary examination, especially pathological examination.

According to the current published reports, the cysts are usually large and often larger than 2 cm (17 cm × 16 cm × 15 cm in our case), which can be differentiated from biliary microhamartomas; further, MCBH is prone to develop in segment 3 in the left lobe and in segment 6 in the right lobe underneath the hepatic capsule [1]. In our case, MCBH was characterized by a space-occupying lesion with unclear boundary in the right lobe.

Consistent with the features of the present case, the cystic wall of MCBHs are generally smooth without solid protrusions, and can be distinguished from cystic lesions with solid protrusions, for example, intraductal papillary neoplasm of the bile duct. MCBHs are mostly benign, and there is no report yet of distant metastases occurring in MCBH [11]. The cancer tissue is mainly composed of fibrous connective tissue, and not only in the glands that surround the biliary ducts but also in the areas between the dilated bile ducts [1]. Moreover, MCBHs are made up of bile ducts, peribiliary glands, and fibrous connective tissue that are rich in blood vessels histologically, but they are typically not connected to the hepatic ducts or biliary tree [7], consistent with the presently discussed cases. This is a distinguishing feature of MCBHs, which is different from Caroli's disease, multiple hepatic cysts, and polycystic livers.

After MCBH was first described by von Meyenburg in the pathologic literature, the diagnostic standard for MCBHs remains contentious. Considering that the imaging features of MCBH are similar to those of other hepatic cystic diseases, making a differential diagnosis very challenging, we faced difficulties in establishing a preoperative diagnosis. However, MCBHs are characterized by the following distinctive features: Caroli's disease often gives rise to atrophy of the relevant liver lobes and compensative hypertrophy of the rest of the hepatic lobes, but MCBHs will not cause such changes. Multiple hepatic cysts and polycystic livers are distributed randomly, with uneven distribution in the liver; whereas, MCBHs are distributed diffusely, mostly characterized by a distribution along the biliary tree. On MRI, MCBHs are hyperintense on T2-weighted imaging with multicystic and honeycomb-like nodules, while they are hypointense on T1-weighted imaging. On ultrasound, the most concordant findings in MCBHs are a combination of uneven hypoechoic masses with hyperechoic cystic walls, which are usually revealed as low density with septate enhancement on CT.

In the case, spiral CT showed that the original upper abdominal mass disappeared after excision of the tumor. A dense linear image was seen in the left edge of the liver, and scattered pneumatosis was found in the abdomen, with mild effusion in the perihepatic and surgical area. An indwelling drainage tube was placed in the patient's abdominal cavity after the surgery. Neither abnormal density lesions nor abnormal enhancing lesions were found in the liver parenchyma. Choledoch and bile ducts, both intrahepatic and extrahepatic, were not found to be dilated, with clear structures around the hepatic portal. The gallbladder was depicted clearly, within which there were no calculi showing high-density imaging; further, the gallbladder wall thickening and enhancement could be noticed. The patient showed good intervention adherence, surgical tolerance, and postoperative therapy. From the patient's point of view, perceived physical discomfort was significantly eased, after treatment. The overall prognosis was satisfactory, and reexamination was recommended.

Author Contributions

HGL & LT provides cases and constructs a framework for the report. WCY sorts out the context of the case, summarizes the characteristics of the case, and conducts a literature review. SFY, HWF & TY organized the images in the article and participated in the literature review.

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