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Hepatobiliary rhabdomyosarcoma mimicking choledochal cyst: Lessons learned☆

Mohamed Abd Elwahab[a], Hosam Hamed[a,*], Ahmed Shehta[a], Mahmoud Ali[a], Khaled Zalata[b]

[a] Gastroenterology Surgical Center, General Surgery Department, Mansoura University, Egypt
[b] Pathology Department, Mansoura University, Egypt

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ABSTRACT

INTRODUCTION: The differential diagnosis of hepatic cystic lesions is a challenging process especially in case of hepatic rhabdomyosarcoma (HRMS) presenting as hepatic cyst.
PRESENTATION OF CASE: We introduce our experience with a case of HRMS in a 3-year-old female patient who was misdiagnosed to have type IV-A choledochal cyst and definitive correct diagnosis was reached after the pathological and immunohistochemical examination of the surgically resected lesion. This case presentation is followed by important practical messages to hepatobiliary surgeons regarding HRMS.
DISCUSSION: HRMS is a rare pediatric tumor. Jaundice is the most common presentation of HRMS followed by abdominal pain and vomiting. Great effort is needed to differentiate the tumor from choledochal cyst and infectious hepatitis. Through evaluation using available imaging studies together with clinical anticipation is mandatory for establishing the correct diagnosis.
CONCLUSION: Differentiation of HRMs from choledochal cyst mandates through evaluation and clinical anticipation. HRMS should be suspected in any child with obstructive jaundice. Once diagnosis is established, multidisciplinary treatment is the best management strategy and it has proved better surgical outcome and long term survival.

1. Introduction

Hepatic rhabdomyosarcoma (HRMS) is a rare lesion accounting for about 0.04% of all childhood cancers.[1] Diagnosis of HRMS is usually difficult because of rare prevalence, non-specific clinical and radiological findings and failure of demonstration of biliary origin and intraductal growth of the tumor. We report a case of a surgically managed HRMS that was preoperatively misdiagnosed as type IV-A choledochal cyst, followed by review of literature on hepatobiliary RMS.

2. Presentation of case

A 3-year-old female patient complained of recurrent attacks of jaundice associated with right hypochondrial pain. She was initially treated as infectious hepatitis at the primary care unit in her village. When the condition did not improve, she was referred to Pediatric Hospital, Mansoura University. There she performed liver function tests, complete blood picture, and abdominal sonography. Provisional diagnosis of choledochal cyst was made and the patient was referred to Mansoura Gastrointestinal Surgical Center, Mansoura University for further evaluation and management.

The patient was admitted to our center on March 2013. She was on oral anticonvulsant therapy for treatment of epilepsy. Otherwise, her past and family history was non-contributory to the condition. On examination, icterus was present. The liver was one finger palpable below the costal margin with mild right hypochondrial tenderness. There was no splenomegally, ascites, lower limb edema, lymphadenopathy or skin rash. Liver function tests showed borderline albumin level (3.3 g/dl), conjugated hyperbilirubinaemia (11.3/8.5 mg/dl), mild elevation in liver transaminases (SGOT: 99 IU/l–SGPT: 89 IU/l) and normal prothrombin concentration. Otherwise laboratory investigations were within normal range. Abdominal ultrasound showed distended gall bladder (GB), cystic dilatation of common bile duct (CBD = 13 mm), common hepatic duct (CHD) and intrahepatic biliary radicals (IHBR).

Magnetic resonance cholangiopancreatography (MRCP) showed cystic dilatation of the CBD (3.5 cm), fusiform dilatation of the right posterior sectorial duct draining into the trifurcation of the biliary confluence with intraductal inspissated mud, dilated CHD (10 mm) and mild dilatation of the rest of IHBR. Thus, provisional diagnosis of type IV-A choledochal cyst was established. As complete excision is

* Corresponding author at: Gastroenterology Surgical Center, Mansoura University, 35516, Egypt. Fax: +20 502243220; mobile: +20 1006178599.
E-mail address: hosam-eldin@hotmail.com (H. Hamed).

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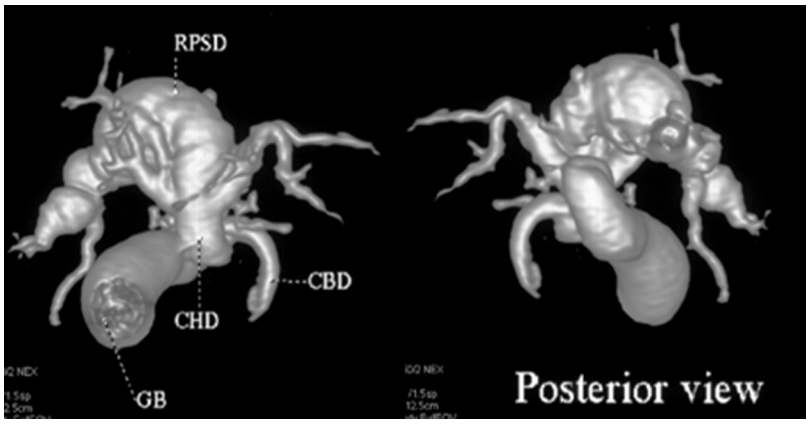

Fig. 1. Magnetic resonance cholangiopancreatography showing dilatation of common bile duct and right biliary system. GB: gall bladder, CBD: common bile duct, CHD: common hepatic duct, RPSD: right posterior sectorial duct.

the best management strategy of choledochal cyst, the patient was evaluated for right hepatectomy with extrahepatic biliary resection (Fig. 1).

Computerized tomographic volumetry of the liver revealed right lobe volume with large vein to segment V and VI draining to middle hepatic vein (MHV) is 550 cc and left liver lobe volume with MHV is 300 cc. Computerized tomographic portography revealed compressed portal vein carina and right portal branch by the cystic lesion in the right lobe (Fig. 2).

On surgical exploration, there was enlarged liver especially right lobe, dilated CBD (30 mm) (Fig. 3). Intraoperative cholangiogram was done and revealed dilated biliary system, fusiform dilatation along CHD and right posterior hepatic duct (Fig. 4). The decision was to do right hepatectomy and left hepaticojejunostomy Roux-en-Y. Postoperative period was uneventful apart from transient

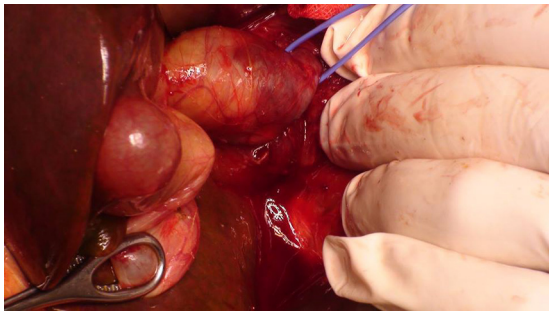

Fig. 3. Operative photo showing cystic dilatation in the extrahepatic bile duct.

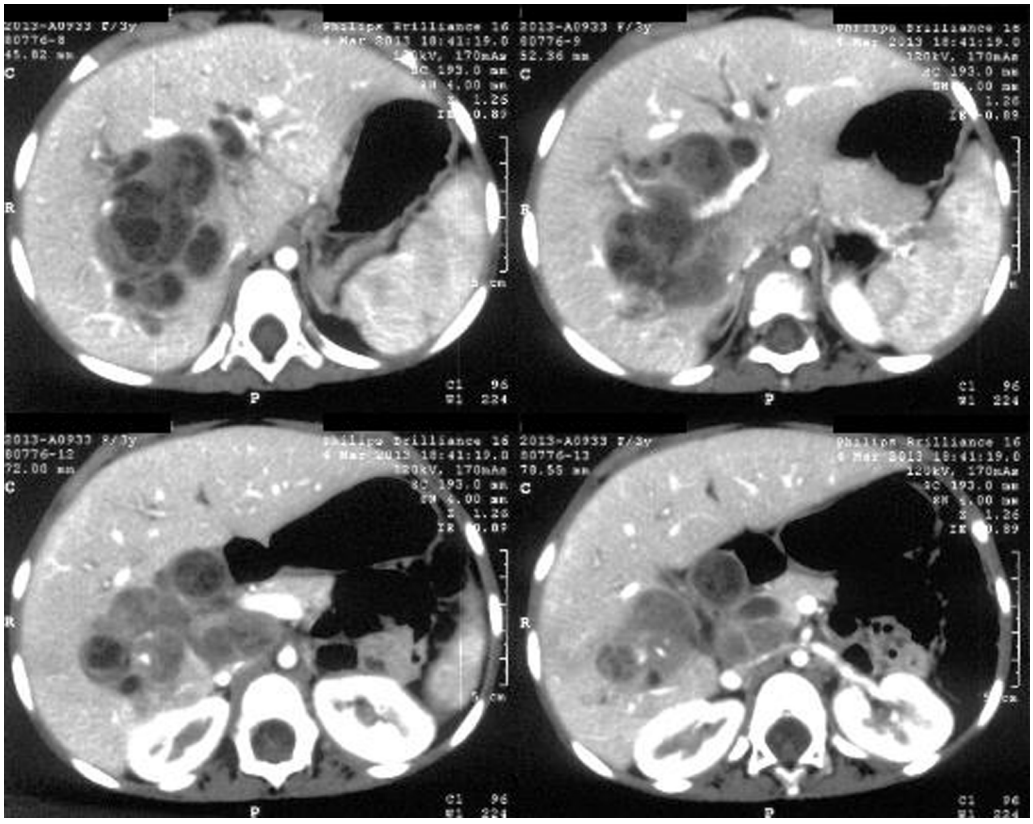

Fig. 2. Computed tomography showing dilated biliary system on the right side with compressed portal vein carina and right portal branch by the cystic lesion in the right lobe.

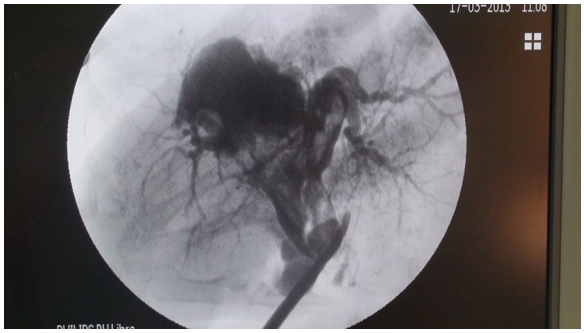

Fig. 4. Intraoperative cholangiogram showing cystic dilatation in the extrahepatic bile duct and the intrahepatic biliary tree in the right hemiliver.

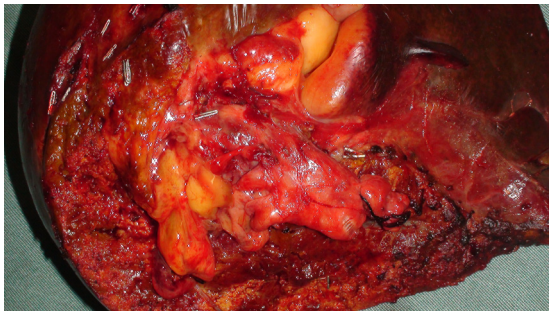

Fig. 5. The surgical specimen showing the resected right hemiliver measuring 15 cm × 12 cm × 5 cm and extrahepatic biliary tree.

hypoalbuminaemia (2.2 g/dl) corrected by three days of albumin infusion therapy. The patient was discharged on 7th postoperative day.

On pathological examination, the tumor tissue is formed of densely arranged short spindle cell proliferation with focal myxoid areas, as well areas of necrosis. Neoplastic cells exhibit moderate atypia with moderate mitotic activity. The tumor tissue is seen infiltrating adjacent liver parenchyma as well large portal areas. By immunohistochemistry, the tumor is focally positive for desmin, as well focal positive nuclear reaction to myogenin. S-100, synaptophysin and chromogranin are negative (Figs. 5–8). It is diagnosed as hepatic rhabdomyosarcoma. The patient was scheduled for evaluation by oncologists for chemotherapy.

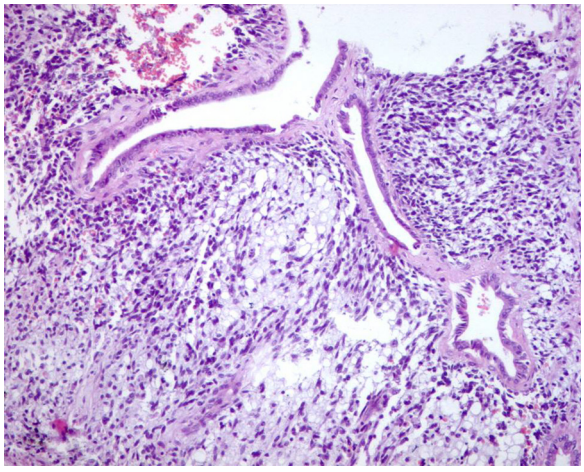

Fig. 6. The tumor tissue is seen infiltrating large portal area (H&Ex 100).

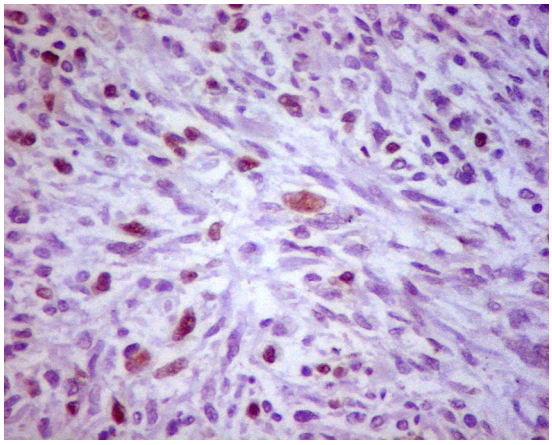

Fig. 7. The tumor tissue is focally positive to desmin (Peroxidasex200).

3. Discussion

Tumors that arise from the biliary tree in early life are generally very rare.[2] HRMS represents about 0.5% of pediatric rhabdomyosarcoma and about 0.04% of all pediatric malignancy.[3] Since Wilks and Moxon reported the first case of HRMS on 1889, there were about 100 cases reported in the available literature till now.[2,4,5] Common presentation of HRMS is jaundice associated with abdominal pain, vomiting and fever. This non-specific presentation makes differential diagnosis of HRMS entails a long list with infectious hepatitis and choledochal cyst on its top.

Most reported cases spent a while treated as infectious hepatitis, especially when they showed initial improvement, until they are referred to specialized centers when the condition is refractory to treatment. This partially justifies the usual long duration between the onset of symptoms and definitive diagnosis and treatment of HRMS in most reported cases.[3] Moderate to marked conjugated hyperbilirubinemia associated with mild elevation in liver transaminases together with the widespread utility of abdominal ultrasound allows the exclusion of infectious hepatitis which is usually the initial suspect.[6]

Most of reported cases in recent literature were misdiagnosed as choledochal cysts until tissue diagnosis is obtained.[7–9] Differentiation between HRMS and choledochal cyst may be difficult even during pathological examination of the surgical specimen. Clinical and laboratory findings in both diseases are the same with no significant difference. Differentiation by diagnostic radiology is difficult when there is no visible mass and detection of intraductal growth cannot be achieved.[10] Also, botryoid RMS, the histological

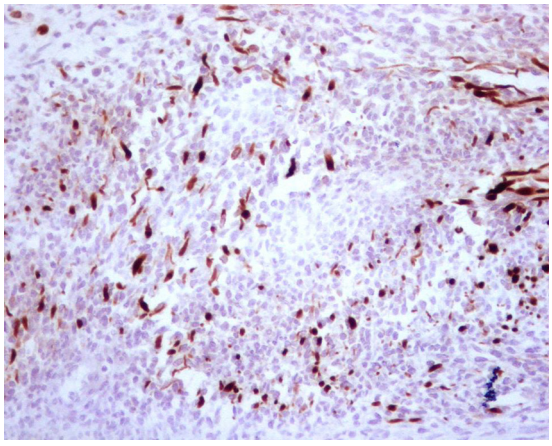

Fig. 8. The tumor cell nuclei are focally positive to myogenin (Peroxidasex200).

variant of HRMS together with embryonal type, may initially be misdiagnosed as benign lesion as developmental anomaly or chronic inflammation.[11]

Differentiation of HRMS from choledochal cyst is of paramount importance as it may help avoiding unwarranted surgery in case of HRMS.[12] HRMS must be put in consideration in any child with obstructive jaundice especially when a mass, intraductal growth or distant metastasis are detected.[3] Uniform dilatation of the biliary system including the gall bladder makes the diagnosis of choledochal cyst, which is typically dilatation of the biliary system, doubtful.[13] Lastly, detection of solid masses during surgical exploration for choledochal cyst can be sent for frozen section and when rhabdomyosarcoma is suspected, immunohistochemical staining using monoclonal antibody to the myogenic regulatory protein "MyoD1" can be used on frozen tissue.[10] Nevertheless, our case is an example of how differentiation between HRMS and choledochal cyst can be difficult even after surgical exploration and intraoperative cholangiography were performed and no intraductal growth or hepatic masses were detected.

Chemotherapy using Vincristine, dactinomycin, and cyclophosphamide can be used in case of extensive disease or R1 resection followed by second look surgery to evaluate resectability. Radiotherapy is indicated when chemotherapy fails or when there is postoperative residual tumor. Palliative treatment of obstructive jaundice is done by endoscopically placed biliary stent.[6]

The concept of operability in HRMS is different from other hepatic tumors. Some authors find it unnecessary to perform major surgical resection with available treatment alternatives especially when the long term survival is not affected by R0 resection.[12] Large tumors involving both liver lobes is not a contraindication for surgery because the tumor is usually localized to the bile ducts and complete excision is not mandatory for long term survival. Also, second look surgery can be performed after chemotherapy for extensive and residual tumors.[10]

4. Conclusion

Hepatic rhabdomyosarcoma must be considered in the differential diagnosis of a child with obstructive jaundice. Great effort is needed to differentiate the tumor from choledochal cyst. Multidisciplinary treatment is the best management strategy and it has proved better surgical outcome and long term survival.

Conflict of interest statement

None.

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None.

Ethical approval

Written informed consent was obtained from the patient parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Mohammed Abdel Wahab: study concept and revision of the draft.

Hosam Hamed, Ahmed Shehta, Mahmoud Ali: data collection and writing the paper.

Khaled Zalata: writing the part relevant to pathology from the paper.

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