

病例报告  
CASE REPORT

## Rhabdomyosarcoma of the common bile duct mimicking choledochal cyst: a rare cause of obstructive jaundice

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**Abstract:** Extrahepatic biliary tract tumors are rare and among them rhabdomyosarcoma is most common. Rhabdomyosarcoma is a soft tissue malignant musculoskeletal tumor and is a very rare malignancy of the common bile duct in children. It usually presents as obstructive jaundice and/or pruritus. If there is no local invasion to the adjacent tissues, the radiological appearance of the tumor lesion is like a choledochal cyst. So the diagnosis is usually made at surgery or by preoperative biopsy. It is important to diagnose early and differentiate it from choledochal cyst and start treatment as early as possible for long time survival of the patient. This case report presented a case of a 10-year-old boy with recurrent onset of obstructive jaundice and fever preoperatively who was diagnosed as choledochal cyst and postoperatively as embryonal rhabdomyosarcoma of the common bile duct. After surgical resection and postoperative chemotherapy, the child had a good prognosis. So it is crucial to know that this rare tumor can mimic congenital choledochal cyst and it should be considered in the differential diagnosis of obstructive jaundice in children.

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**Key words:** Biliary rhabdomyosarcoma; Obstructive jaundice; Choledochal cyst; Common bile duct; Child

### 类似胆总管囊肿的胆总管横纹肌肉瘤：阻塞性黄疸的罕见原因

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**【摘要】** 肝外胆道肿瘤罕见，其中以横纹肌肉瘤最常见。横纹肌肉瘤是一种软组织恶性肌肉骨骼肿瘤，是儿童非常罕见的一种胆总管恶性肿瘤。它通常表现为阻塞性黄疸和/或瘙痒。如果肿瘤没有局部浸润邻近组织，其影像学表现则类似胆总管囊肿。因此，确诊通常是在手术时或术前通过活检。早期诊断、与胆总管囊肿鉴别、尽早开始治疗对延长患者生存时间有重要意义。该文报道 1 例 10 岁男童，反复阻塞性黄疸和发热，术前诊断为胆总管囊肿，术后确诊为胆总管胚胎性横纹肌肉瘤。经手术切除、术后化疗，该患儿预后良好。故必须了解这种罕见的肿瘤可以类似先天性胆总管囊肿，并且在儿童阻塞性黄疸的鉴别诊断时应加以考虑。

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**【关键词】** 胆道横纹肌肉瘤；阻塞性黄疸；胆总管囊肿；胆总管；儿童

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## 1 Introduction

Obstructive jaundice in children beyond the neonatal period can be caused by choledochal cyst, congenital biliary tract abnormalities, gallstones, strictures due to chronic cholangitis and uncommonly by neoplastic biliary obstruction due to rhabdomyosarcoma (RMS)<sup>[1-2]</sup>. RMS, a skeletal muscle malignancy, is the soft tissue tumour most common among children aged 0-14 years, accounting for 1% of all malignancies<sup>[1,3-4]</sup>. Median age is about 3 years for the occurrence of hepatobiliary RMS in children<sup>[1]</sup>. This uncommon tumour was originally reported by Wilks and Moxon in 1875<sup>[5]</sup>. Because of its low incidence, diagnosis of biliary RMS is very challenging, needing a high degree of suspicion and the use of new and appropriate imaging techniques<sup>[6]</sup>. As embryonal RMS is both solid and cystic, preoperative imaging may not suffice to define the lesion exactly<sup>[7]</sup>. Due to newer radiological techniques and available combined therapy modalities, treatment and prognosis of biliary RMS are frequently changing<sup>[8]</sup>.

## 2 Case report

A 10-year-old boy, the only child of non-consanguineous parents, immunized according to the Expanded Programme on Immunization (EPI) schedule, presented to the Paediatric Gastroenterology and Nutrition Department, Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, Bangladesh with high-grade intermittent fever, occasionally associated with chills, rigors and jaundice, accompanying by anorexia, but no nausea and vomiting. He had occasional abdominal pain, mild and diffuse, involving mostly the right upper abdomen. There was a history of similar attack two months back. He had no history of pruritus, coagulopathy or encephalopathy. On examination, he was conscious, oriented, cooperative, mildly pale, icteric and febrile, with stable vital signs, no stigmata

of chronic liver disease and having normal growth. Systemic examination revealed a soft abdomen, no palpable lump, nontender just palpable liver. And absence of splenomegaly, ascites, skin rash or cervical lymphadenopathy.

There was a neutrophilic leucocytosis with a high erythrocyte sedimentation rate. The liver function tests were abnormal. Regarding viral markers HBsAg, anti-HAV IgM, anti-HCV IgM were negative except antibodies to herpes simplex virus type 1 (anti-HSV1) IgM were positive (Table 1). In imaging findings, ultrasonogram of the abdomen showed slightly enlarged liver with homogenous echotexture, slightly dilated intrahepatic biliary channels, and fusiform dilatation (3.8 cm × 5.2 cm × 5.8 cm) of the common bile duct (CBD) with soft tissue extending into the distal part of CBD compressing the head of the pancreas, which is suggestive of a choledochal cyst (infected or hemorrhagic – type IVb). An irregular hypoechoic soft tissue mass (5.3 cm × 3.8 cm) near the head of the pancreas and flow within the mass with Resistance Index (RI) = 0.51 is seen in the Doppler study of the portal vein, hepatic veins, and inferior vena cava. Magnetic resonance cholangiopancreatogram (MRCP) revealed gross fusiform dilatation of CBD and common hepatic duct (CHD) producing a sac-like mass 4.3 cm × 2.7 cm in size, slightly dilated intrahepatic biliary channels, and low signal intensity within the dilated CBD and these features suggestive of a choledochal cyst (Type IVb) with sludge and secondary dilatation of intrahepatic biliary radicles (Figure 1).

Intraoperatively, the CBD was widely dilated, and its wall was thin and oedematous. The cystic dilatation of the CBD contained soft tissue, necrotic and tumorous tissue up to intrahepatic bile duct (Figure 2).

Choledochotomy was done. Soft, necrotic and tumorous tissue was cleared off as far as possible from the CBD and sent for histopathology. Proximal and distal patency of the CBD was ensured and a T-tube placed *in situ* before closure. No evidence of metastasis or lymph node involvement was found.

Histopathology report was suggestive of malignant small round blue cell tumour (Figure 3A) followed by immunohistochemistry revealed tumour cells were positive for desmin, myogenin and vimentin whereas negative for pancytokeratin and leukocyte common antigen which were compatible with embryonal rhabdomyosarcoma (Figure 3 B-D).

Table 1 Laboratory indexes

Indexes	Results	Normal range
Complete blood count		
Haemoglobin (g/dL)	11.6	13-17
Erythrocyte sedimentation rate (mm in 1st hr)	77	0-10
White blood cell count (/mm <sup>3</sup> )	17 500	4 500-11 000
Neutrophil leucocytes (%)	79	40-80
Lymphocytes (%)	18	20-40
Platelet count (/mm <sup>3</sup> )	340 000	150 000-400 000
Liver function tests		
Total bilirubin (mg/dL)	10.39	0.3-1.0
Direct bilirubin (mg/dL)	4.5	1-4
Serum albumin (g/L)	34	35-50
Prothrombin time (s)	23.2	12-16
International normalized ratio	1.89	<1.4
Alanine aminotransferase (U/L)	302	35-50
Aspartate aminotransferase (U/L)	455	10-34
Alkaline phosphatase (U/L)	878	128-356
Gamma glutamyl transferase (U/L)	358	9-85
Viral markers		
Antibodies to herpes simplex virus type 1 (anti-HSV1) IgM	Positive	



Figure 1 Results of coronal magnetic resonance cholangiopancreatogram Gross fusiform dilation of the CBD (red arrow) and slightly dilated intrahepatic biliary channel.



Figure 2 Intraoperative findings of the CBD Soft tissue, necrotic tissue, and tumorous tissue are observed in the CBD.

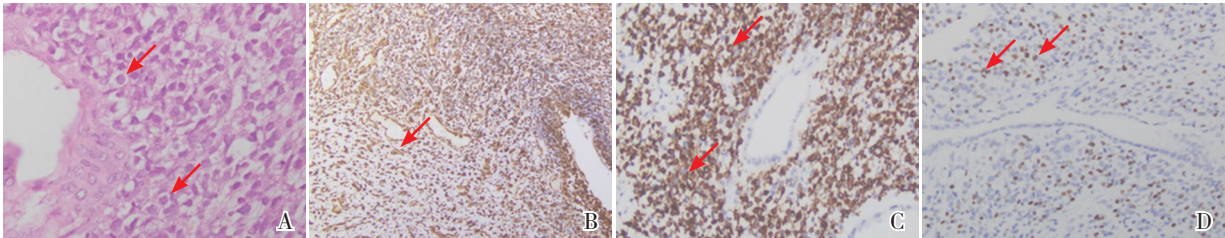


Figure 3 Results of histopathology A: Perivascular condensations of malignant cells have hyperchromatic round nuclei and contain generous amounts of eosinophilic cytoplasm. The stroma is loose and myxoid (red arrow, Hematoxylin and Eosin, ×200). B: Tumor cells and stroma show diffuse positive immunoreactivity (red arrow, Vimentin, ×100). C: Tumor cells show diffuse positive immunoreactivity (red arrow, Desmin, ×100). D: Most of the tumor cells show positive immunoreactivity (red arrow, Myogenin, ×100).

Postoperatively, patient received vincristine, adriamycin and cyclophosphamide (VAC) chemotherapy. There were no apparent distant

metastasis, as chest X-ray and bone marrow study of the patient were normal. During three months of follow up, there were no findings of recurrence, and

the obstructive jaundice resolved. Close follow up is being continued.

### 3 Discussion

Common causes of obstructive jaundice in children include hepatitis, drug-induced jaundice, and benign, recurrent cholestasis<sup>[9]</sup>. Surgical causes of obstructive jaundice include choledochal cysts, choledocholithiasis, strictures, congenital anomalies, and rarely RMS<sup>[6]</sup>. For the exclusion of this etiology, the first investigation should be ultrasonogram. In this case, the ultrasound scan demonstrated fusiform dilatation of the CBD with soft tissue structure resultant obstructive jaundice. Congenital anomalies like stricture and choledocholithiasis were excluded by these initial imaging findings leaving the imaging differential diagnosis as choledochal cyst filled with sludge and primary biliary neoplasm or primary liver tumor with metastasis to the biliary tree. However, the hepatic neoplasm was excluded due to normal hepatic architecture.

Histological subtypes of RMS are embryonal, botryoid, alveolar, spindle cell variant, and RMS not otherwise specified<sup>[10]</sup>. Among them, the biliary tract has neoplasms of only embryonal or botryoid histology. Embryonal variety is three times more common than other varieties but the botryoid subtype has the best prognosis<sup>[11]</sup>. Embryonal RMS in children mainly presents in the head and neck, genitourinary tract, extremities, parameningeal area, retroperitoneum, and rarely biliary tract<sup>[12]</sup>. It may arise in any part of the biliary channel<sup>[13]</sup> or within a choledochal cyst<sup>[14]</sup>. In this case, RMS occurred in the wall of the CBD. RMS of the biliary tree is a disease of young children, 1-9 years of age with a slight male predominance<sup>[10,15-16]</sup>.

RMS presents with obstructive jaundice in 60%-80% of cases<sup>[17]</sup>. Other less common presenting features are abdominal pain, abdominal distension, anorexia, nausea, vomiting, and fever<sup>[6,16,18-19]</sup>. In this case, the patient presented with recurrent obstructive

jaundice, fever, abdominal pain, anorexia, and hepatomegaly. Conjugated hyperbilirubinemia and raised liver enzymes are commonly seen as in our case. In our case, leucocytosis was due to cholangitis.

The initial imaging modality for diagnosis should be the ultrasonogram. Ultrasonogram with color Doppler is the optimal method of assessing the response to preoperative and postoperative chemotherapy<sup>[6]</sup>. The characteristic sonographic finding of biliary RMS is a biliary system filled with solid cystic content giving a lace-like appearance and the presence of intralesional vascularity by color Doppler confirms the possibility<sup>[20-21]</sup>. A preoperative sonogram can show biliary cystic dilatation, which may suggest a congenital choledochal cyst. Because of bile stasis and filling of the lumen by soft, inspissated debris, both solid and cystic areas may present within an embryonal RMS and for that reason, an embryonal RMS can mimic the radiological features of a choledochal cyst that occurred in our case<sup>[7,22-23]</sup>. Embryonal RMS may occur within a choledochal cyst in the literature but histopathologically it is not possible to confirm, whether it is primary RMS of the CBD or within the choledochal cyst<sup>[24]</sup>.

For better evaluation of the biliary channel, we did MRCP that revealed gross fusiform dilatation of the CBD producing sac-like mass and intrahepatic biliary channels slightly dilated, suggestive of choledochal cyst with sludge and secondary dilatation of intrahepatic biliary radicles. Other imaging techniques may be computed tomography (CT) scan or magnetic resonance imaging (MRI). The identification of the solid nature of the tumor by CT scan can sometimes aid differentiation from a choledochal cyst<sup>[19]</sup>. CT scan and MRI are also helpful to see the extent and staging of the disease<sup>[6]</sup>.

Embryonal RMS is usually confirmed postoperatively using histology and immuno-histochemistry. Myoblastic differentiation and expression of skeletal muscle markers, such as desmin, myogenin, and/or myoD1 found in RMS by histopathology<sup>[25]</sup>. Over 90% of embryonal RMS are



positive for desmin<sup>[26]</sup>, but this is non-specific. Recently it was shown that myogenin is a specific immune-histochemical marker for embryonal RMS<sup>[17,26]</sup>. In our case, histopathology was positive for malignant small round blue cell tumor and immune-histochemical staining was positive for desmin, vimentin, and myogenin.

Over the past 30 years, embryonal RMS therapy has greatly improved and the cure rate increased 3-fold<sup>[27]</sup>. Surgical removal, chemotherapy, and radiotherapy are the current modalities of treatment<sup>[27]</sup>. The Intergroup Rhabdomyosarcoma Study IV recommends a combination of vincristine, actinomycin D and cyclophosphamide, ifosfamide, or etoposide as chemotherapy for embryonal RMS<sup>[12]</sup>. Besides, most children with embryonal RMS also need surgery and/or radiotherapy to achieve local tumor control<sup>[17]</sup>. In our case, after surgical resection, VAC chemotherapy is being given. Although gross total excision is not always possible, the prognosis remains relatively good which is probably owing to the favorable histology. Neoadjuvant chemotherapy followed by resection of the residual tumor has been associated with good outcomes<sup>[25]</sup>. The absence of distant metastasis, embryonal variety, and small size  $\leq 5$  cm, place our patient in the intermediate-risk group according to Children's Oncology Group<sup>[10]</sup>.

In conclusion, biliary RMS presents with obstructive jaundice and in cystic tumors with central necrosis can mimic a choledochal cyst. Thus, embryonal RMS should be considered in the differential diagnosis of children presenting with obstructive jaundice and a cystic or soft tissue mass within the CBD.

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