Rhabdomyosarcoma of the common bile duct: an unusual cause of obstructive jaundice in a child

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Biliary rhabdomyosarcoma (RMS) is a rare malignancy of childhood. The radiological findings and clinical presentation of the tumor can mimic an entirely different pathology. The incidence of RMS has impeded the development of a standardized form of treatment. A four-year-old child with botryoid RMS in the common bile duct is reported herein to emphasize the role of surgery in the small-sized tumor and the role of endoscopic retrograde cholangiopancreatography (ERCP) in the diagnosis and relief of biliary obstruction before total excision.

Key words: biliary rhabdomyosarcoma, jaundice, endoscopic retrograde cholangiopancreatography, botryoid rhabdomyosarcoma, embryonal rhabdomyosarcoma.

Rhabdomyosarcoma (RMS), which originates from a variety of sites, is the most common soft tissue sarcoma in childhood[1]. Although RMS is the most common tumor of the biliary tree in childhood, it is a rare lesion, accounting for about 1% of all RMS in children[2]. Preoperative radiological and clinical studies may be insufficient to define the lesion due to the nature of the tumor, and initial diagnostic impression can be completely different[3].

Localized disease is curable with a combined modality therapy, with a five-year survival rate of more than 75%, but the low frequency of RMS of the extrahepatic biliary tree has impeded the development of a standardized form of treatment[2-4].

We present herein a child with RMS of the biliary tree in whom endoscopic retrograde cholangiopancreatography (ERCP) was used for both tissue diagnosis and relief of obstruction. Total excision of the tumor with Roux-en-Y hepaticojejunostomy was the initial treatment of choice.

Case Report

A four-year-old boy was admitted to our unit with complaints of jaundice and acholic stool. Serum total/direct bilirubin was 15.00 mg/dl/12.21 mg/dl (N: 0.10-1.20/0.0-0.30 mg/dl), alkaline phosphatase 1022 U/L (N: <269 U/L), gamma glutamyl transpeptidase 653 U/L (N: <18 U/L), aspartate aminotransferase 370 U/L (N: <52 U/L), and alanine aminotransferase 360 U/L (N: <39 U/L). Hepatitis markers were negative.

Ultrasonography (US) showed mild enlargement of the liver, hydropic gallbladder with normal lumen and wall, and significant dilatation of the intrahepatic biliary tract and common hepatic duct. US also showed a hyperechoic tubular solid mass lesion with a diameter of approximately 34 mm in length and 1 cm in width in the dilated lumen of the common bile duct. Magnetic resonance cholangiopancreatography (MRCP) showed the polypoid solid lesion in the lumen of the common bile duct (Fig. 1A). The pancreatic duct was normal. This lesion was initially interpreted as common bile duct polyp or parasitic infestation. Empirical treatment with praziquantel for ascaris was commenced.

Endoscopic retrograde cholangiopancreatography (ERCP) showed a filling defect in the common bile duct (Fig. 1B). After

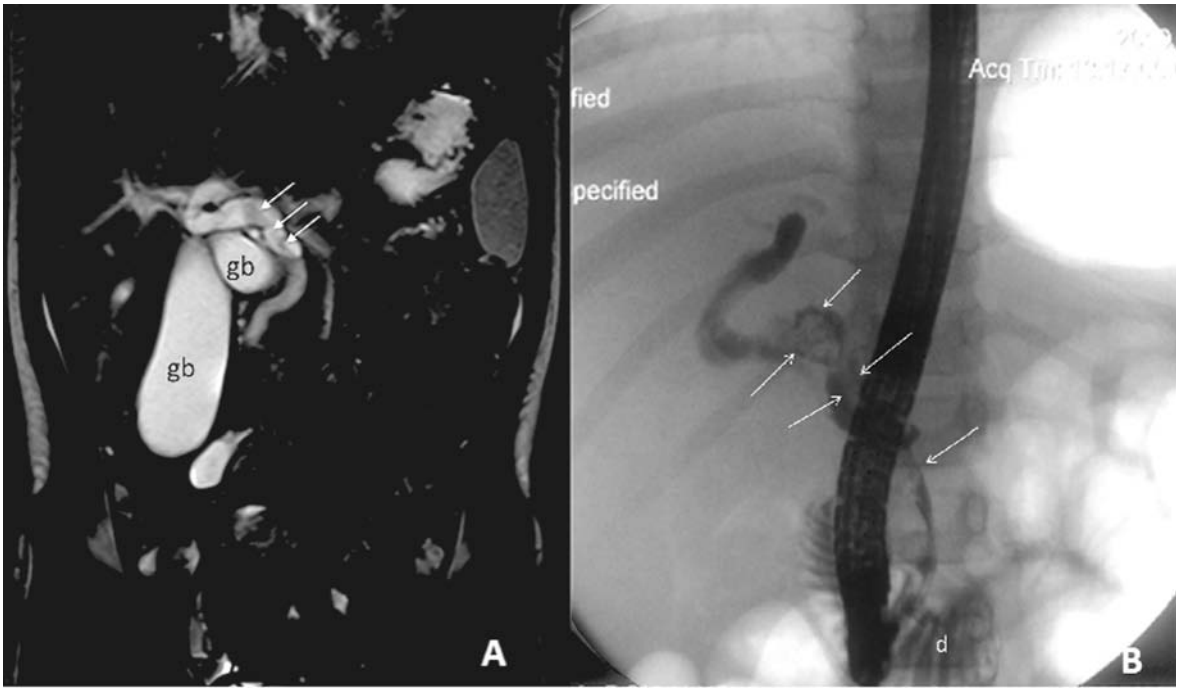

Fig. 1A. Coronal T2-weighted image of MRCP shows hypointense solid lesion (arrows) in the lumen of the common bile duct. gb: gallbladder **1B.** ERCP shows filling defect in the lumen of the common bile duct (arrows). d: duodenum

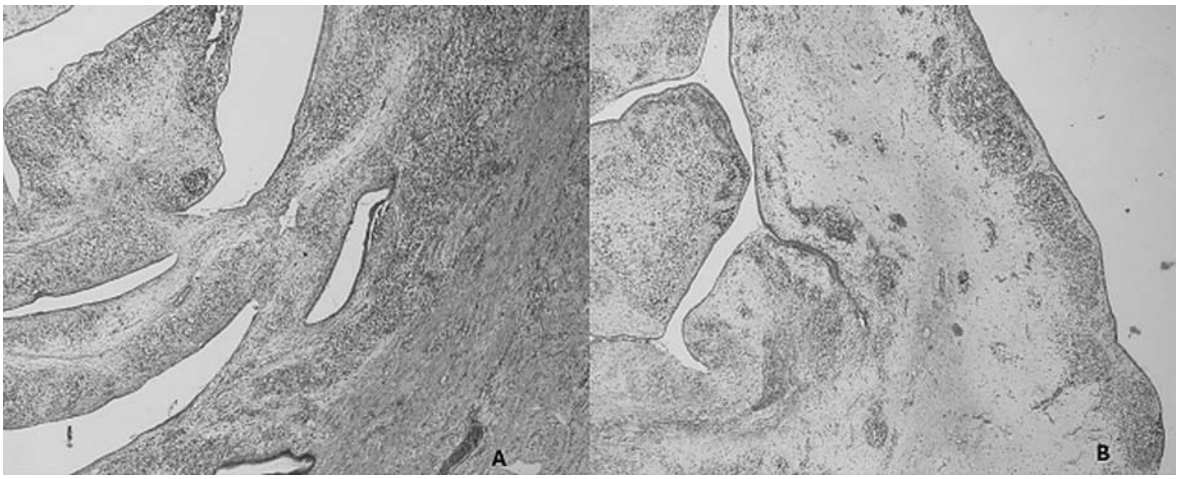

Fig. 2A, B. Tumor originating from the wall of the common bile duct as a polypoid mass protruding to the lumen. Under the epithelium, the cambium layer composed of tumor cells is seen and the stroma is loose. (hematoxylin&eosin [H&E], original magnification X4).

sphincterotomy, a part of the solid lesion was removed with the help of gallstone balloon, and a 7 French plastic stent (Huibregtse flap endoprosthesis; PBN Medicals, Denmark) was propelled into the right hepatic duct and left in the common bile duct. The serum total/direct bilirubin levels were 4.95 mg/dl/2.82 mg/dl after biliary stenting.

Histopathological examination of the ERCP specimen was compatible with botryoid type embryonal RMS, with a cambium layer characterized by subepithelial condensed desmin-positive tumor cells under an intact epithelium and loose stroma.

A solid intraluminal tumor mass was found located in the common bile duct during

laparotomy. The distal parts of the right and left hepatic ducts, main hepatic duct, gallbladder, and common bile duct together within the tumor were resected. An enlarged portal lymph node was excised, and then right and left hepatic ducts were anastomosed to the Roux-en-Y jejunal segment. The postoperative course was complicated by massive delayed upper gastrointestinal bleeding. Surgical exploration revealed hemorrhage from the sphincterotomy site.

On macroscopic examination, the diameter of the common bile duct was 3.5x1.5 cm, and its cross-section revealed a yellow-white colored mass completely filling the lumen. The gallbladder was unremarkable. Microscopic examination of the tumor specimen revealed a polypoid mass with a loose stroma protruding from the common bile duct wall to the lumen, covered with cubical epithelium and sheets of spindle or round tumor cells under the epithelium (Fig. 2A, 2B). Tumor cells were desmin- and myogenin-positive by immunocytochemistry. These findings confirmed the diagnosis of botryoid-type embryonal RMS. The tumor was positive in the distal margin. The gallbladder was normal microscopically. Lymph nodes (2x1x0.8 to 0.9x0.5x0.3 cm) excised from the portal hilus were tumor-free.

Discussion

Rhabdomyosarcoma (RMS) of the biliary tree is a rare mesenchymal neoplasm that affects children at a median age of three years with a slight male predominance. Common presenting symptoms are jaundice and abdominal pain often associated with abdominal distension, vomiting and fever[4]. Mild elevations of conjugated bilirubin and hepatic transaminases are signs for diagnosis[5].

Jaundice, acholic stools and elevated bilirubin levels were the symptoms in our patient. Biliary stenting was ameliorative in the present case.

Abdominal US is the first and simplest examination method. However, RMS may not be considered initially in the differential diagnosis. Additionally, an embryonal RMS mass can mimic the radiological appearance of a choledochal cyst because of its combined cystic and solid nature[6,7]. MRCP provides detailed information about the biliary tree, location and extension of the mass and the anatomy of the biliopancreatic junction. MRCP determined the common bile duct location of the mass and the anatomy of the biliary tree in the present case. However, in our case, US and MRCP were not diagnostic for RMS, nor could they exclude the possibility of parasite.

Endoscopic retrograde cholangio-pancreatography (ERCP) can be performed to relieve biliary obstruction, obtain biopsy, visualize the biliary tree, and place the stent[6]. ERCP was very helpful in our case. First, it showed that the ampulla of Vater was normal; second, sphincterotomy revealed the protruding tumor and provided the biopsy; third, complete visualization of the biliary tree was performed; and fourth, a biliary stent was placed to relieve obstruction. The diagnosis of RMS can be made by biopsy during ERCP.

It has been reported previously that there is no standardized treatment approach for biliary RMS in children. A previous cumulative report of the Intergroup Rhabdomyosarcoma Study (IRS) emphasized that the tumor was usually increased in size (>6 cm), locally invasive and not suitable for total excision. Additionally, the course was usually complicated by biliary catheter-related sepsis[1]. In recent years, new reports have mostly suggested adjuvant chemotherapy and conservative surgery[3,4].

In our case, we preferred a surgical approach as a first step because of the small size of the tumor, which was suitable for total excision. Additionally, outcome for the patients with residual disease after initial surgery is excellent[1]. In embryonal RMS, tumor extension to the surrounding tissues may render total resection impossible. As a result, aggressive resections and intraoperative cholangiography for definition of tumor extension were recommended in earlier reports[3]. In our patient, absence of local invasion allowed total excision, and we did not perform cholangiography during the surgery because we had ERCP images. We preferred endoscopic stent placement for biliary drainage at the time of diagnosis, and we removed the stent during the surgery. During Roux-en-Y hepaticojejunostomy, left and right hepatic ducts were anastomosed separately after spatulation in order to avoid the risk of stenosis development.

This type of RMS responds well to chemotherapy

and radiotherapy. Neoadjuvant chemotherapy following biopsy can be a treatment option in cases not suitable for surgery. Our patient received chemotherapy postoperatively, and no recurrent disease had developed at one year of follow-up.

Outcome in RMS appears to have improved over the last several decades except in patients with distant metastases at diagnosis[1]. Gross total resection is rarely possible but outcome is good despite residual disease after surgery.

We think that surgery should be considered among the initial treatment options in suitable patients with biliary RMS. ERCP provides detailed visualization of the biliary tree, and biliary stenting relieves obstructive jaundice.

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