RABDOMYOSARCOMA OF THE BILIARY TREE*

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Rabdomyosarcoma of the biliary tree is one of the rare causes of biliary tract obstruction in childhood. Nevertheless it is the most common cause of obstructive jaundice due to neoplastic biliary obstruction. We present a two-year-old child with obstructive jaundice secondary to an embryonal rhabdomyosarcoma of the biliary tree. She underwent surgery and, after total excision of the mass, a hepaticojejunostomy and Roux-en-Y anastomosis were performed. She was referred to the Pediatric Oncology Group for follow-up. Rhabdomyosarcoma of the bilary tree, although rare, must be considered in the etiology of obstructive jaundice in children.
Key words: rhabdomyosarcoma, biliary tree.

Rabdomyosarcoma of the biliary tree is an uncommon disorder of the bile ducts and is also a rare cause fo biliary tract obstruction in childhood[1,2]. However, it is the most common cause of neoplastic biliary tract obstruction in children[1]. The tumor, which originates from the biliary ducts, is often polypoid and embryonal botyroid types. It either obstructs the biliary lumen and causes obstructive jaundice or causes complications like cholangitis and hepatic abscess[3]. Herein we present a patient with rhabdomyosarcoma of the biliary tree whom we diagnosed and operated on in our department. Rhabdomyosarcoma of the biliary tree must be considered in the differential diagnosis of obstructive jaundice in children.

Case Report

A two-year-old female was admitted with a one-month history of jaundice, acholic defecation, loss of appetite and weight loss. On physical examination the patient was icteric with hepatomegaly and acholic stool.

Her liver function tests were as follows: aspartate aminotransferase 220 U/L (normal 2 to 35 U/L), alkaline phosphatase 4000 U/L (normal 70 to 350 U/L), and total bilirubin 7.6 mg/dl (direct 6.4 mg/dl). Tumor markers including alpha-fetoprotein, beta human chorionic gonadotropin and carcinoembryonic antigen were normal. Abdominal ultrasonography (USG) and computed tomography (CT)

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confirmed a cystic, lobular-shaped heterogneous mass 8 cm in diameter between the head of the pancreas and the porta hepatis (Fig. 1). Radiological and radioisotopic investigations revealed no metastases.

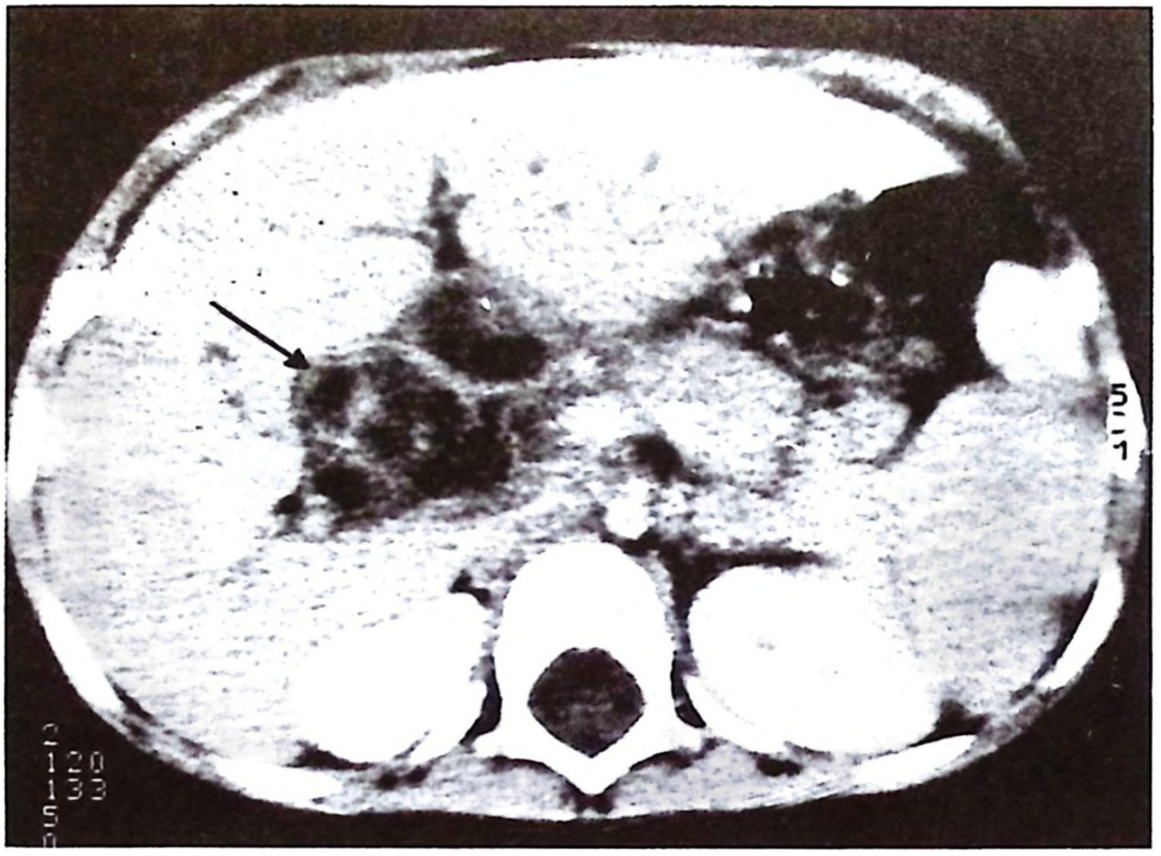

Fig. 1: Computed tomography CT scan of the abdomen at the level of T12-L1 levels the mass between the porta hepatis and the head of the pancreas.

The patient underwent an exploratory laparotomy which revealed a mass 8x3x2 cm in diameter arising from the choledochus. The mass extended to the ductus cysticus and common hepatic bile duct but did not penetrate the head of the pancreas or right-left hepatic ducts. The obstructed choledochus was opened and botryoid nodules and gelatinous material spontaneously extruded from the cyst cavity. Frozen section evaluation revealed rahbdomyosarcoma.

The tumor was considered surgically resectable and had no metastasis or vascular invasion. After total excision of the tumor, biliary draniage was achieved by hepaticojejunostomy and Roux-en-Y anastomosis. Final pathology on the specimen confirmed an embryonal rhabdomyosarcoma stemming from the choledochus. Microscopic residual tumor was found at the proximal and distal bile duct margins (Fig. 2). Lymph node from the porta hepatis was free of disease. Based on the above, the patient was given stage III protocol of the Intergroup Rhabdomyosarcoma Study Group, and was transferred to Pediatric Oncology

uneventfully on the 7th postoperative day. Two weeks later, a course of chemotherapy including vincristine, actinomycin D, cyclophosphamide and etoposide was started. Eleven courses of chemotherapy have been adiminstered to the patient to date. Radiation therapy was not applied. The patient is healthy and has had no relapse over the 18 months since the time of diagnosis.

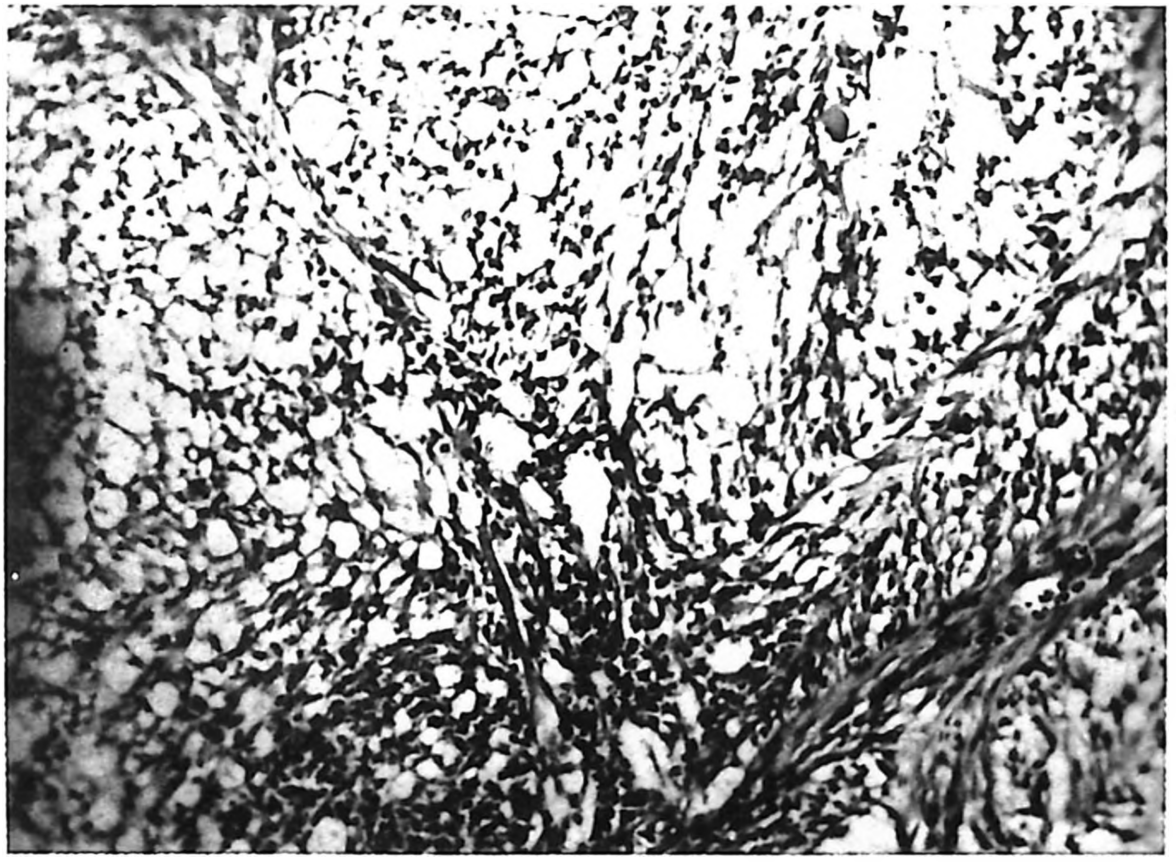

Fig. 2: Microscopic sections illustrate atypical striated rhabdomyoblasts in stroma (H&E, X 200).

Discussion

Although rhabdomyosarcoma is the most frequently seen soft tissue sarcoma in infants and children, its ratio in the biliary system is a reported three percent[5, 6]. rhabdomyosarcoma of the biliary tree usually stems from the choledochus, common hepatic bile duct or ampulla of Vater, but there are reports in the literature which present rhabdomysarcoma stemming from the intrahepatic bile ducts. The tumor as a rule extends into the liver and/or the ampulla of Vater. Because of this extension, total resection of the tumor is unfortunately impossible in most cases[3, 6]. Obstructive jaundice is the most common presenting symptom of this tumor[4]. But other symptoms such as abdominal pain, fever, itching and weight loss are also seen; cholangitis and liver abscess may complicate the

clinical presentation[3]. Most patients with rhabdomyosarcoma develop metastatic disease usually via the bloodstream and less commonly via the lymphatics to the regional lymph nodes, lung, bone, marrow, bone, liver and brain[5].

In our patient, the tumor did not extend to the junction of the right and left hepatic ducts or to the ampulla of Vater. The gallbladder was free of tumor. Obstructive jaundice and weight loss were the main symptoms of our patient; she was free of metastases after radiological and radioisotopic evaluation.

Ultrasonography, USG, CT and/or magnetic resonance imaging, endoscopic retrograde cholangio-pancreatography, or percutaneous transhepatic cholangiography can be used for diagnosis[6]. The diagnosis is unfortunately usually established during exploration. Ultrasonography and CT revealed the tumor between the head of the pancreas and the porta hepatis in our patient, but the diagnosis was established during exploration.

Currently, a multidisciplinary regimen is the recommended therapy for rhabdomyosarcoma of the bile ducts. Surgery, chemotherapy anad radiotherapy combination has been suggested. Chemotherapy must be multiagent. Radiotherapy, when used properly, should reduce the incidence of local recurrence at the primary tumor site[1,3]. Prognosis for rhabdomyosarcoma of the biliary tree is very poor, due to its strategic anatomical location, propensity for liver invasion and the frequent delay in diagnosis[1]. Patients with metastatic disease at the time of diagnosis also have a poor outcome[3].

In conclusion, rhabdomyosarcoma of the biliary tree, although rare, must be considered in the etiology of obstructive jaundice in children.

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