Case report

Primary non-Hodgkin’s lymphoma of the common bile duct presenting as obstructive jaundice

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Primary non-Hodgkin’s lymphoma of the extrahepatic bile duct presenting as obstructive jaundice is an extremely rare disease. At this writing, a review of the medical literature disclosed 17 reported cases of primary non-Hodgkin’s lymphoma arising from the extrahepatic bile duct. We, herein, report an additional case of obstructive jaundice caused by primary non-Hodgkin’s lymphoma of the common bile duct, in a 21-year-old woman. Our patient showed clinical evidence of obstructive jaundice, and endoscopic retrograde cholangiopancreatography and abdominal magnetic resonance imaging demonstrated a long strictured segment of the common bile duct with proximal bile duct dilatation. These clinical and radiological findings resembled those of cholangiocarcinoma. Resection of the common bile duct tumor, cholecystectomy, lymph node dissection, and Roux-en-Y hepaticojejunostomy were carried out. Histology and immunohistochemistry of the resected specimen confirmed a diffuse large B-cell-type malignant lymphoma involving the common bile duct. She received four courses of combination chemotherapy, including cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP), and 3060 cGy external irradiation. She has been well, without evidence of tumor recurrence, 17 months after the surgery. In summary, first, primary non-Hodgkin’s lymphoma of the extrahepatic bile duct, despite its rarity, should be considered in the differential diagnosis of causes of obstructive jaundice. Second, an accurate histopathologic diagnosis and surgical resection, if feasible, combined with chemotherapy with or without radiotherapy may be the approach to offer a chance for cure.

Key words: non-Hodgkin’s lymphoma, bile duct, obstructive jaundice

Introduction

Hepatobiliary involvement by malignant lymphoma is usually a secondary manifestation of systemic lymphoma.¹⁻⁶ In contrast to secondary involvement, primary malignant lymphomas arising from the hepatobiliary tree are extremely rare, and obstructive jaundice is seldom a presenting symptom of malignant lymphoma.¹⁻⁶ The occurrence of primary non-Hodgkin’s lymphoma arising from the extrahepatic bile duct was first described by Nguyen,⁷ in 1982. Subsequently, 16 cases of primary non-Hodgkin’s lymphomas arising from the extrahepatic bile duct have been documented in the literature.⁸⁻²³ We, herein, report an additional case of obstructive jaundice caused by primary non-Hodgkin’s lymphoma of the common bile duct, in a 21-year-old woman. We also review the literature pertaining to this condition.

Case report

A 21-year-old woman was admitted with a 1-month history of progressive jaundice. She complained of pruritus and dark urine, but had no weight loss, fever, or chills. She denied any previous history of travel, use of hepatotoxic medications, any alcohol ingestion, or hepatobiliary disease. On admission, she was afebrile, and her blood pressure and pulse were normal. Her right upper abdomen was tender to direct palpation, but there was no rebound tenderness or Murphy’s sign. The liver and spleen were not palpable, and there were no palpable superficial lymph nodes. Laboratory evaluation revealed white blood cell count, 6000/mm³; hemoglobin, 11.7 g/dl; platelet count, 349000/mm³; serum albumin, 3.4 g/dl; aspartate aminotransferase, 171 U/l; alanine aminotransferase, 428 U/l; alkaline phosphatase, 280 U/l; and gamma-glutamyl-transpeptidase, 428 U/l. Total bilirubin was 5.1 mg/dl,
with 3.5 mg/dl direct fraction. Coagulation profiles were within normal limits. The serologic tests for hepatitis viruses A, B, and C were negative. Abdominal sonography revealed that an echogenic, nonshadowing mass was present within the dilated common bile duct (Fig. 1).

Abdominal computed tomography revealed a well-circumscribed heterogeneous enhancing mass in the midportion of the common bile duct, with proximal bile duct dilatation, but there was no evidence of enlarged lymph node or hepatosplenomegaly. Endoscopic cholangiopancreatography revealed a long strictured segment in the midportion of the common bile duct, with proximal bile duct dilatation (Fig. 2). A coronal T2-weighted magnetic resonance image revealed dilatation of the proximal common bile duct and both intrahepatic bile ducts, and tapered narrowing of the distal common bile duct (Fig. 3).

The first diagnosis was cholangiocarcinoma. Exploratory laparotomy was performed. Grossly, the common bile duct was diffusely thickened and a firm tumor was found in the midportion of the common bile duct. However, there was no definite lymph node enlargement near the common bile duct. During laparotomy, multiple frozen sections of the common bile duct and lymph nodes showed no evidence of cholangiocarcinoma. Resection of the common bile duct tumor, cholecystectomy, lymph node dissection, and Roux-en-Y hepaticojejunostomy were carried out. The resected specimen disclosed diffuse infiltration of large cells with vesicular nuclei, prominent nucleoli, abundant cytoplasm, and numerous mitoses (Fig. 4). Immunohistochemical studies were positive for leukocyte common antigen and B-cell marker (CD 20) and negative for