INTRODUCTION

Carcinoid tumors of the biliary tract are rare and account for 0.2%-2% of all GI carcinoids (1). Intrahepatic biliary carcinoid is exceedingly uncommon. These are usually slow-growing neoplasms with a low malignant potential, and hence are amenable to aggressive surgical management. Preoperative diagnosis is difficult because they mimic the signs and symptoms of choledocholithiasis and/or cholangiocarcinoma. The present case serves to highlight that intrahepatic biliary carcinoids constitute a rare but identifiable subset of bile duct tumors.

CLINICAL CASE

A 60 y/o male initially presented with clinical jaundice and elevated transaminases. MRI and MRI-cholangiography identified a mass suspicious for cholangiocarcinoma (Klatskin’s tumor) (Figs. 1 and 2). The patient was initially managed with a percutaneous transhepatic cholangiography tube. At the time of surgery (August 2005), a successful removal of a firm nodular mass in the ductal bifurcation area was achieved, and biliary continuity was reestablished with a Roux-en-Y hepatic-jejunostomy using multiple hepatic-enteral segmental anatomoses. Pathology revealed a carcinoid tumor (Figs. 3 and 4) of the bile duct with no tumor-positive lymph nodes. The patient received no adjuvant radiation or chemotherapy. Patient status is currently normal.

Carcinoid tumors in extrahepatic bile ducts are a rare form of biliary obstruction, with fewer than 34 cases reported in the English literature (2). The most common anatomic sites were CBD (58%), perihilar region (28%), cystic duct (11%), and common hepatic duct (3%). Unlike cholangiocarcinoma, biliary carcinoids occur more commonly in younger patients and women. Aggressive local invasion by the primary tumor is rare, and metastases occur in less than one-third of patients. Surgical resection is recommended. The final diagnosis is usually confirmed by immuno-histochemistry studies (3).
COMMENTS

Median survival time for patients suffering from carcinoid tumors was 102 months, whereas it was 33 months for patients suffering from cholangiocarcinomas in one series (1).

Available information suggests that patients with biliary carcinoid have an overall favorable prognosis after aggressive surgical management (3). The mean disease-free follow up after resection in another study was 32 months (range, 3 months-20 years) (4). Hence surgery with curative intention should be considered wherever possible.

This case serves to highlight that biliary carcinoids constitute a rare but identifiable subset of bile duct tumors. Their diagnostic workup should include US, CT, and cholangiography. Surgical exploration is universally indicated in physiologically fit patients, with operative management to include resection and a restoration of biliary continuity. Data on adjuvant therapy remain investigational; however, available information suggests that patients with biliary carcinoid have an overall good prognosis after aggressive surgical management (5).

REFERENCES