Case Report

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Carcinoma of the Cystic Duct Leading to Obstructive Jaundice

A Case Report and Review of the Literature

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Surgical treatment

Abstract
Only 34 cases of primary cystic duct carcinoma have previously been published in the literature. Most of these cases presented with upper abdominal pain and a palpable mass in the right upper quadrant due to gallbladder hydrops or cholecystitis. We report a case of cystic duct carcinoma with the clinical presentation of obstructive jaundice. The patient was treated by cholecystectomy, resection of the common bile duct and a Roux-en-Y hepaticojejunostomy. An extended lymph node dissection was not performed. Fourteen months after the operation the patient died with local carcinoma recurrence. A literature review comparing clinical signs, surgical treatment, and outcome of 14 Japanese and 21 reported Western cases, including ours, was performed. Extended lymph node dissection in addition to combined resection of the gallbladder and ductus hepaticocholedochus appears to offer a better prognosis and larger survival, including the chance of potential cure.

Introduction

Only carcinomas fulfilling the strict criterias defined by Farrar [1] represent cystic duct cancers. Therefore cancer of the cystic duct includes: (1) restricted tumor growth to the cystic duct; (2) no evidence of tumor in the gallbladder, hepatic or common bile duct, and (3) histological proof or carcinoma [1].

The worldwide incidence of primary carcinoma of the cystic duct is very low, being reported to fall between 2.6 and 3.3% of all bile duct cancers [2, 3].

Cystic duct cancer in most cases presents without clinical signs of obstructive jaundice. Since the first description of cystic duct carcinoma in 1941 [4] only 11 of the 34 described cases caused obstructive jaundice [4–13]. We describe here a cystic duct carcinoma which presented with obstructive jaundice, and we review the literature to identify the surgical procedure with the most favorable outcome. Compared to other malignant biliary tumors the prognosis of cystic duct carcinoma seems more favorable, always depending on the tumor stage and on the surgical procedure.

Patients and Methods

In a retrospective single-center series of 1,759 cholecystectomies performed at our institution between January 1986 to March 1995, only 1 case of carcinoma of the cystic duct was found as defined by Farrar [1], resulting in an extremely rare incidence rate of 0.6‰.

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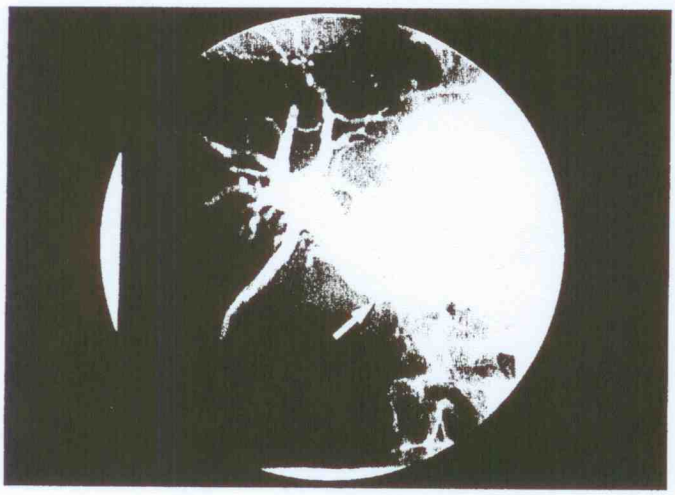

Fig. 1. Percutaneous transhepatic cholangiography revealing pronounced dilatation of the intrahepatic bile ducts with a filiform narrowing down to a complete obstruction of the common hepatic duct (white arrow) caused by cystic duct carcinoma.

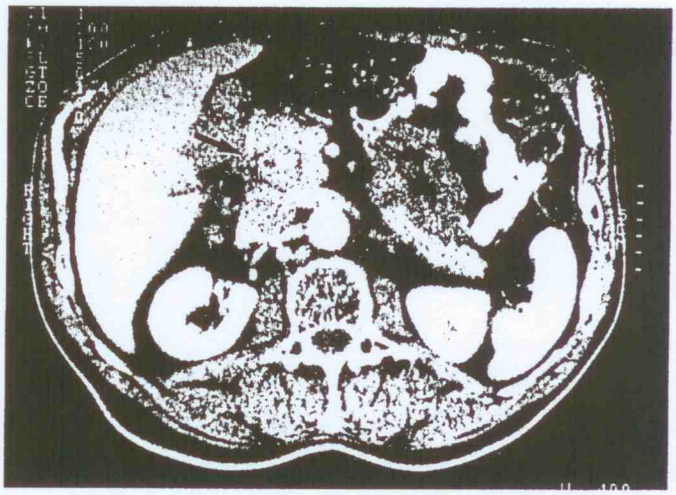

Fig. 2. Computed tomography revealing tumor recurrence and lymph node metastases (black arrow) 11 months postoperatively.

Case Report

A 74-year-old male was admitted to our department with a 3-week history of jaundice and pruritus. During the previous 2 months, he reported a weight loss of 8 kg and intermittent episodes of epigastric colicky pain. On clinical examination the patient appeared deeply jaundiced, but otherwise in good general condition and afebrile. Abdominal palpation revealed a light tenderness in the right upper quadrant. The liver was not enlarged, nontender, with a smooth surface. On laboratory blood tests the following parameters were pathologic (normal values in parentheses): total bilirubin 696 µmol/l (<20); direct bilirubin 346 µmol/l (<5.1); SGOT 26 U/l (<18); SGPT 46 U/l (<22), and alkaline phosphatase 620 U/l (<170). Tumor markers were also elevated with 3.9 ng/ml (<1.5) CEA and 210 kU/l (<33) CA 19-9.

Ultrasonography of the upper abdomen showed a 2.5 × 1 cm solid mass in the region of the distal bile duct with proximal bile duct dilatation, cystic duct obstruction and a moderately dilated gallbladder. Duplex sonography (DS) showed intact vascularity without tumor involvement of the portal vein and hepatic artery. Percutaneous transhepatic cholangiography revealed a marked dilatation of the intrahepatic bile ducts with a filiform narrowing down to a complete obstruction of the common hepatic duct (fig. 1). Cholestasis was treated preoperatively by percutaneous biliary drainage for 6 days. With the presumptive diagnosis of obstructive jaundice due to a bile duct tumor in the region of the confluence of the cystic duct, a laparotomy was performed. At surgery, a 2-cm mass was palpable at the junction of the cystic duct with the common bile duct, extending to the portal vein. Lymph nodes in the hepatoduodenal ligament were suspicious for tumor metastases. The liver was free of tumors on inspection and bimanual palpation, and confirmed by liver biopsy results. A cholecystectomy was performed with en-bloc resection of the cystic duct and the hepaticocholedochus, followed by Roux-en-Y hepaticojejunostomy reconstruction. The gallbladder was filled with thick bile, but without gallstones. At the distal margin of the gallbladder a 2.5-cm-long stenosis was found. The cystic duct itself could not be identified. Histological examination showed a moderately to well-differentiated papilliform adenocarcinoma in the cystic duct with infiltration through the cystic wall to the subserosa and periductal fatty tissue. The final histological preparation of the specimen showed no evidence of adenocarcinoma in the gallbladder, hepatic duct and common bile duct, thus fulfilling all the cystic duct tumor criteria defined by Farrar [1]. Two lymph nodes in the hepatoduodenal ligament were infiltrated by tumor cells confirming a tumor stage pT3pN1pM0 according to the UICC classification [14]. The postoperative course was uneventful, and the laboratory parameters returned to normal. The patient was readmitted 11 months postoperatively, again with epigastric pain and increased tumor markers (5.6 ng/ml CEA, 312 kU/l CA 19-9). At ultrasonography, there was an indication of local tumor recurrence with multiple lymph node metastases at the biliary anastomosis which was confirmed by computed tomography (fig. 2). Three months later the patient died. At autopsy, local tumor recurrence with lymph node metastases was found.

Discussion

Common bile duct and hepatic duct tumors account for more than 95% of extrahepatic bile duct tumors [15]. Cystic duct tumors that fit Farrar's [1] strict definition are exceedingly rare, accounting for only 2.6–3.3% of all bile duct cancers [2, 3]. Twenty-one of 35 cases including ours have been treated surgically in Western countries, and 14 patients were treated in Japan [16] (table 1). Comparing these 2 groups the following remarks can be made. The mean age was similar in both groups (63.2 years, range 38–80 years). The male to female ratio was 2.5:1 and 1:1