KASUISTIK

Congenital bile duct cyst: A premalignant lesion of the biliary tract associated with adenocarcinoma – a case report –

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Summary

The incidence of carcinoma arising in the wall of a congenital bile duct cyst is high and there is no doubt that these lesions represent a precancerous state of the biliary tract. In almost all cases congenital bile duct cysts are related to anomalous arrangements of the pancreaticobiliary duct system which seems to play a crucial role in the development of cystic bile ducts and biliary carcinogenesis. Bile stasis together with reflux of pancreatic juice causing longstanding inflammation and activation of bile acids might be the factors in carcinogenesis of the exposed bile duct epithelium in the cystic wall. In the case of primary or secondary extrahepatic bile duct cysts, primary excision is mandatory because of the high risk to develop biliary cancer with even nowadays poor prognosis despite advantages in biliary surgery during the last years. We report a case of a young woman in which bile duct cancer was found arising in the wall of a congenital bile duct cyst. Despite radical surgery the outcome was poor proving the high malignant potential of bile duct cancer. The question of possible tumor seeding in hepatobiliary surgery is discussed as a way of inducing hepatobiliary metastatic tumors.

Key words: Choledochal cyst – preneoplastic lesion – biliary carcinogenesis – biliary carcinoma – surgical therapy – tumor seeding

Zusammenfassung

(Kongenitale Gallengangszyste: eine prämaligne Läsion der Gallenwege assoziiert mit einem Adenokarzinom)

Die Karzinominzidenz bei kongenitaler zystischer Gallengangspathologie ist hoch, und es steht außer Zweifel, daß diese Pathologie der Gallenwege eine Präkanzerose darstellt. In den meisten Fällen von kongenitalen Gallengangszysten besteht gleichzeitig eine Anomalie der pankreatikobiliären Verbindungsstelle, die für die Zystenentstehung und Karzinogenese eine entscheidende Rolle zu spielen scheint. Lang anhaltende Cholestase sowie Reflux von Pankreassekret mit Aktivierung primärer Gallensäuren werden für die Karzinomentstehung verantwortlich gemacht. Somit besteht bei primär oder sekundär aufgetretenen Gallengangszysten die Indikation zur chirurgischen Exzision, um das Auftreten eines Gallengangkarzinomes mit auch heute noch schlechter Prognose zu verhindern. Wir stellen das Fallbeispiel einer solchen Karzinomentwicklung bei einer jungen Patientin vor, welche trotz radikaler chirurgischer Maßnahmen eine infauste Prognose zeigte. Ferner wird die Frage der Tumoraussaat durch den chirurgischen Eingriff diskutiert.

Schlüsselwörter: Bile duct cyst – biliary carcinogenesis – bile duct cancer

Introduction

Carcinoma of the intra- and extrahepatic biliary tract is a relatively rare disease. In most cases the etiology of these cancers is rather obscure. Depending on the location of the carcinoma there are some risk factors identified, including congenital cystic and dysplastic lesions of the biliary tract. Even if congenital cystic dilatation of the biliary ductal system is a rare abnormality, it is important to be recognized because of its high association with carcinoma arising in the choledochal cyst wall epithelium. For a long time the classical clinical traid of »pain, jaundice and abdominal mass« is not present in most patients, which often causes longterm delay in diagnosis therefore leading to longstanding inflammation of the biliary epithelium and possible development of carcinoma in the cystic wall. Hence primary radical excision of choledochal cysts is mandatory to prevent development of biliary cancer which has a poor prognosis. We present the case of a biliary carcinoma arising in a choledochal cyst in a young woman with a poor outcome despite extensive radical surgery. Early development of peritoneal and subcutaneous abdominal wall metastases prove the high malignant potential of this lesion.

Case report

A 33-year-old female with an unevenful past history was admitted to our department with a recent history of nausea, vomiting and weight loss of 6 kg in the last month. A palpable abdominal mass in the right upper quadrant with little pain was found on physical examination. An infrahepatic cystic mass in continua-

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tion of the hepatoduodenal ligament was demonstrated by abdominal ultrasonography. Intravenous cholangiotomography and abdominal computed tomography revealed a choledochal cyst of 5 x 5 x 5 cm in size (fig. 1 and 2). In accordance to the Todani classification (1) of congenital bile duct cysts the biliary cyst was classified as type 1 a. The lower part of the cystic dilatation showed wall irregularities suspicious for malignant epithelial transformation. Gastroduodenal endoscopy revealed a light compression of the duodenal bulb explaining the clinical symptomatology of nausea and vomiting. ERCP was not performed because there was no doubt about the indication for an operative treatment of this bile duct cyst with suspicion of malignancy. Laboratory blood tests are summarized in tab. 1. Only liver transaminases and bilirubin were slightly elevated indicating cholestasis caused by the bile duct cyst. The tumor marker CEA was at the upper normal limit.

Surgical abdominal exploration was performed and the gallbladder was found to be markedly distended. A large choledochal cyst measuring approximately 6 cm in diameter displacing the duodenum laterally and adhering to the gallbladder and the right side of the transverse colon was found. During exploration and palpation of the cyst, a tumor was found in the distal portion of the choledochal cyst measuring 2 x 2 cm in size causing light stenosis of the distal choledochus. Unfortunately a frozen section of biopsied material from the distal end of the choledochal cyst wall revealed adenocarcinoma. For radical tumor clearance in this young patient we performed an en-bloc partial pancreaticoduodenectomy with excision of the cyst together with the gallbladder and a right hemicolectomy because of the above mentioned suspicious adhesions. The final histological examination (fig. 3) confirmed a moderately differentiated adenocarcinoma of the cystic choledochal wall with invasion of the cystic duct, the papilla of Vater, the pancreatic capsule and the pericolic fatty tissue (stage pT3, pN1, pMO according to UICC) (2). Resection margins were free of tumor and the colon was not infiltrated. After histological examination we considered to have performed a RO-resection.

The postoperative course was uneventful for two months and the patient felt well and regained 2 kg of weight. Two months after the operation and shortly before the first scheduled postoperative follow-up, the patient observed an induration and redness around the right umbilical area. Two weeks later during her first postoperative clinical examination we found a tumor measuring 3 x 3 cm in diameter adherent to the fascia and located at the right of the navel in a site where there was no operative wound or postoperative drainage canal. Additional abdominal computed tomography confirmed a subcutaneous tumor in the abdominal wall suspicious for a metastasis of bile duct cancer (fig. 4). Three months after the first operation the patient underwent surgery again, and the paraumbilical parietal tumor and the adjacent abdominal wall were resected. Unfortunately laparotomy demonstrated diffuse peritoneal metastases in addition to the abdominal wall metastasis confirming a tumor grade 4 B (according to UICC) (2). The abdomen was closed without further intervention. A proposed palliative chemotherapy postoperatively was refused by the patient. She died nine months after the second operation.

Discussion

Bile duct cysts of congenital origin may be intrahepatic or extrahepatic. In the latter location usually solitary and called choledochal cysts are found. In 1977 *Todani* et al. (1) reported 37 cases of congenital bile duct cysts and classified them into six different types. Like in our case over 80% of congenital choledochal cysts can be classified as type 1 according to the *Todani* classification (1, 3). As the cause of choledochal cysts, *Babbit*

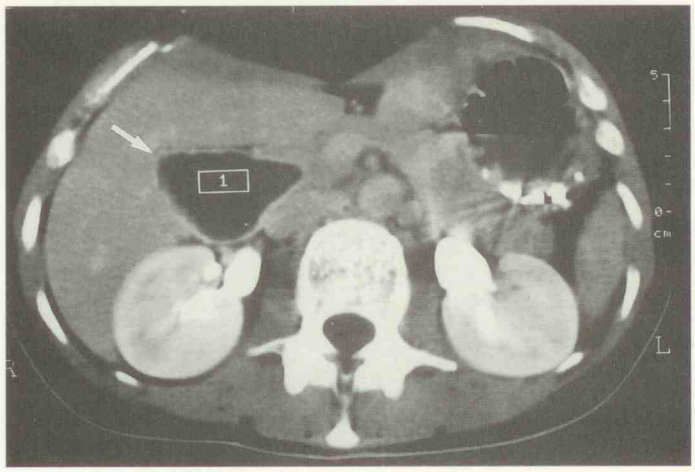

Fig 1: Abdominal computed tomography revealing a huge subhepatic cystic formation measuring 5 x 5 x 5 cm in size (arrow)

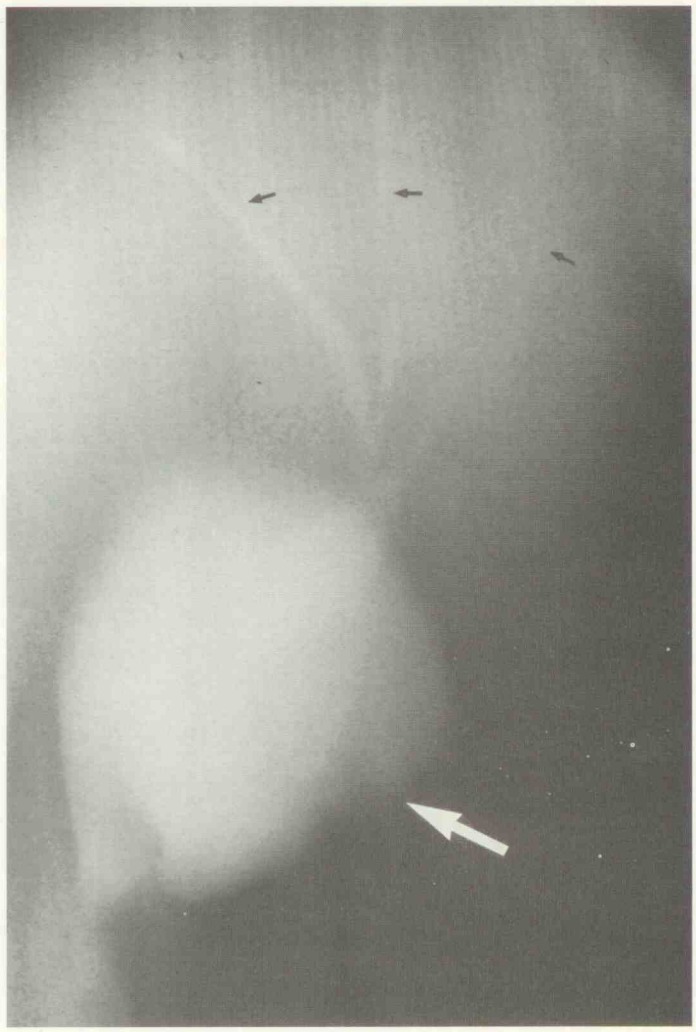

Fig. 2: Intravenous cholangiotomography revealing a choledochal cyst type 1 a according to the *Todani* classification (1, 3) (white arrow). The intrahepatic bile ducts are not dilated (black arrows)