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Thirteen-Year Disease-Free Survival after Surgery for Cystic Duct Carcinoma: A Case Report

Zine Abedine Benchellal[a] Kountélé Gona Soro[a]
Isabelle Orain[b] Hélène Poret[a] Abd Hak Ferhi[a]
Julien Darquies-Chevalley[a]

[a]Department of Digestive Surgery and [b]Department of Pathological Anatomy and Cytology, University Hospital Center of Tours, Tours, France

Key Words
Cystic duct · Carcinoma · Surgery · Extrahepatic bile duct · Disease-free survival

Abstract
Cystic duct carcinoma is known to have a better prognosis compared to its location in other biliary ducts. Only one case with a survival over ten years has been previously published. The authors report a case of survival over 13 years without recurrence or metastasis. Preoperative diagnosis of cystic duct tumor was carried in a 66-year-old male. Under the diagnosis of carcinoma of the cystic duct, the patient underwent en bloc resection of the gallbladder, cystic duct, hepaticocholedochus and lymph node dissection. A Roux-en-Y hepaticojejunostomy was performed. Histological examination revealed a moderately differentiated adenocarcinoma of the cystic duct. Five months later the patient underwent second look surgery for benign obstruction of the hepaticojejunal anastomosis. The patient is still doing well 13 years later without any local recurrence or metastasis. To our knowledge, this is the longest disease-free survival ever published in the literature. This case sustains that better and longer survival is possible with a real chance of potential cure if radical surgery is performed.

Introduction

Carcinoma primarily developing in the cystic duct is uncommon, compared to other locations in the biliary ducts [1]. Carcinomas of the cystic duct (CCD) are less published. They may have better survival than malignancies located in other biliary ducts [2, 3]. To our knowledge no previous study has report a survival over 13 years. We report a 66-year-old male who is still doing well 13 years after surgical resection for CCD.

Benchellal Zine Abedine
Service de Chirurgie Digestive, Hôpital Trousseau
FR–37044 Tours Cedex (France)
Tel. +33 247 474 634, Fax +33 247 474 641, E-Mail z.benchellal@chu-tours.fr

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Case Report

A 66-year-old male was admitted to the digestive surgery service of the teaching hospital of Trousseau for recurrent right upper quadrant pain in January 1995. Abdominal ultrasonography showed cholecystolithiasis. Laparoscopic surgery was performed and peroperative cholangiography made opaque only 1 cm of the cystic duct because of an intraluminal obstruction. At laparotomy a tumor was found in the distal portion of the cystic duct suggesting a CCD. Histological examination of the frozen section revealed a carcinoma. The patient underwent en-bloc resection of the gallbladder, the cystic duct with its tumor, the common bile duct and the choledochus. An extending lymph node dissection in the hepatoduodenal ligament and a Roux-en-Y hepaticojejunal anastomosis were performed. The postoperative course was uneventful. Final histological examination of the resected specimen showed a moderately differentiated excretobiliopancreatic adenocarcinoma of the cystic duct (**fig. 1**) with perineural invasion (**fig. 2**) There was no invasion of the gallbladder or the common bile duct and the choledochus. The tumor was classified as $T_2N_0M_0$. The patient is doing well 13 years after the resection without any sign of recurrence or metastasis.

Discussion

CCD is rare according to the literature [1, 3–5]. CCD fulfilling the definition criteria are likely observed in the early stage. However its incidence would be higher than is believed now if all the advanced stage cases were added. It is proved that inflammation of the biliary duct epithelium potentially leads to malignancy sooner or later, in consideration to the frequently associated carcinomas with pancreaticobiliary maljunction [2]. Indeed, because of its narrow lumen, the cystic duct is more often affected by gallstones during their migration from the gallbladder to the choledochus than other extrahepatic biliary ducts. Therefore this carcinogenesis factor should increase the incidence of carcinoma in the cystic compared to the other biliary ducts.

Clinically, CCD may be completely asymptomatic [1] or be revealed by signs suggesting other pathologies. It sometimes presents with Mirrizi syndrome [3–5] and is diagnosed preoperatively. Peroperative diagnosis is common when performing operation for acute cholecystitis such as in our patient. In consequence, any relapsing cholecystitis, particularly without gallstones, associated with uniform distended gallbladder should suggest the possibility of cystic duct tumor [1, 2]. For us, the impossibility to catheterize the cystic duct for cholangiography or to opacify the hepaticocholedochus, such as in this case, should lead to the search for cystic duct tumor.

On the treatment level, most authors recommend radical surgery [2–4] though, according to the definition, advanced stages of CCD are excluded. These authors propose en bloc resection of gallbladder, cystic duct, common bile duct, choledochus and an extending lymph node dissection of the hepaticoduodenal ligament. Some authors are more aggressive by performing a duodenopancreatectomy [1]. We think this attitude is too aggressive and may not be necessary even if only early-stage CCD are concerned. Biliary duct resection and extending lymph node dissection should be sufficient.

According to the literature [2], among all patients with carcinoma of the extrahepatic trees, those fulfilling the definition criteria of CCD should have better and prolonged survival than those with other locations. This better survival is justified by advanced stage exclusion. In spite of these restricted criteria, the overall published survival to our knowledge varies from 14 months to 125 months [1–4] with a median survival of 20 months. Thirteen years 156 months) after surgery our patient is still doing well without any recurrence or metastasis. To our knowledge this is the second case with a survival over ten years and the first one with a disease-free survival over thirteen years. Beside this

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prolonged survival, this case report may suggest possibly curability when large en bloc resection of the biliary trees with extending lymph node dissection is performed.

Conclusion

The incidence of CCD might be increased if advanced stage CCD were taken into account. Disease-free survival over thirteen years is possible, but large resection and extending lymph node dissection are necessary.

Fig. 1. Invasive moderately differentiated adenocarcinoma of the cystic duct (low magnification).

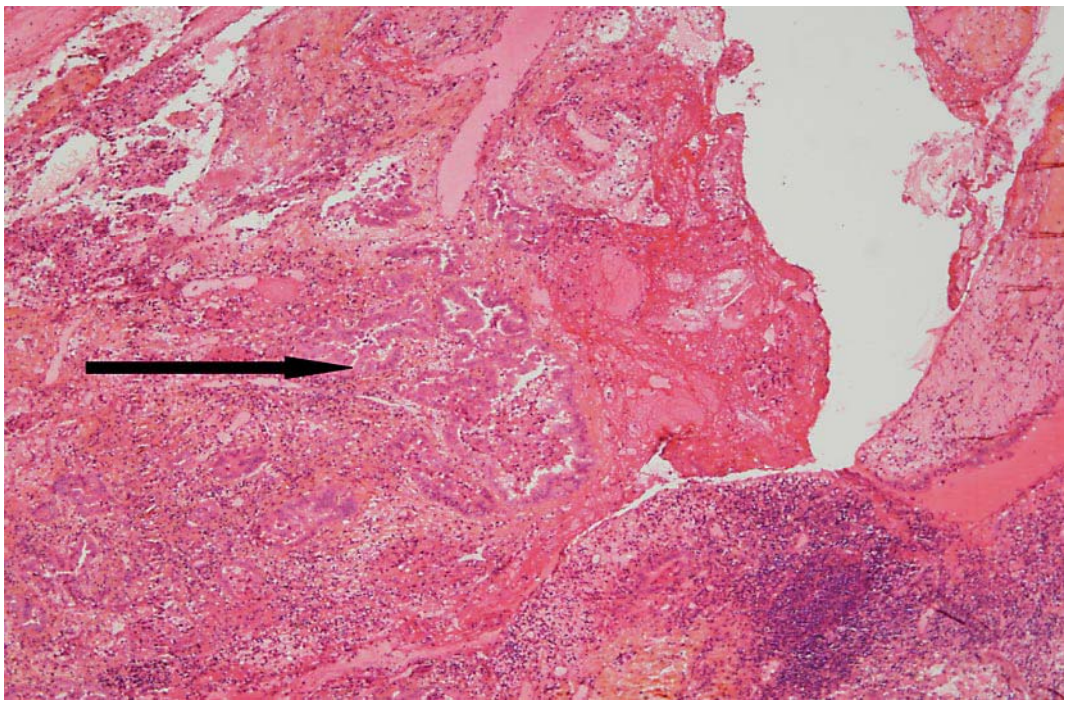

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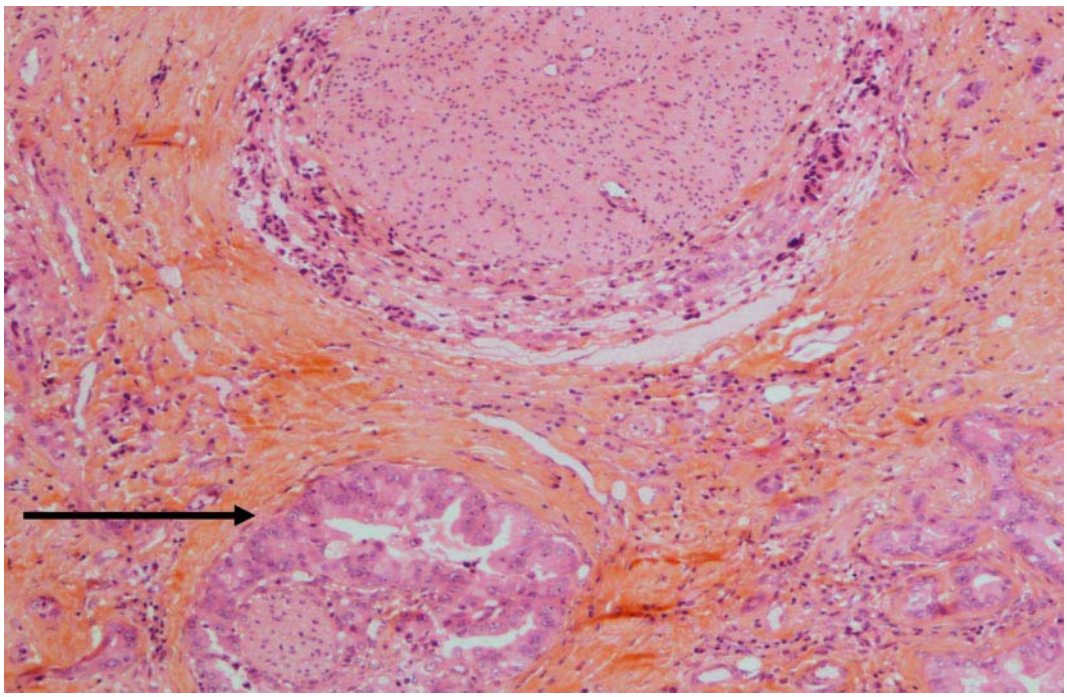
Fig. 2. Branching tubular glands in an abundant fibrous stroma with perineural invasion (higher magnification).



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