M. Afifi El-Sayed
T.H. Juma

Department of Surgery, Al-Amiri
Hospital, Kuwait

Primary Cystic Duct Carcinoma

Case Report and Literature Review

Key Words

Cystic duct carcinoma
Extrahepatic bile ducts
Empyema of the gall bladder
Tumours
Mirrizzi's syndrome

Abstract

Primary cystic duct carcinoma is rare. To date only 27 cases have been reported in the English literature, and 17 cases in the Japanese literature. This paper reports another case of primary cystic duct carcinoma. A 70-year-old male presented with signs and symptoms of acute calculous cholecystitis and dilated extrahepatic bile ducts. An ERCP failed technically. An ultrasound-guided aspiration was performed to drain an obstructed gall bladder and an adjacent pyogenic liver abscess. A cholecystogram showed an obstructed cystic duct. On table a tumour was suspected at the junction of the cystic duct and common hepatic duct (CHD). En masse excision of the gall bladder, cystic duct, tumour, part of the CHD and the supraduodenal part of the common bile duct was performed with a Roux-en-Y hepatico-jejunostomy. Histopathologically the tumour fulfills all of Farrar's criteria for primary cystic duct carcinoma. Our case is of interest since it is the 2nd case to present with Mirrizzi-like syndrome.

Introduction

Primary cystic duct carcinoma is a rare disease. Establishing a pre-operative diagnosis of this disease is very difficult because of the narrow lumen of the cystic duct [1]. This case is

the 2nd case of primary cystic duct carcinoma in our centre. The diagnosis was suspected on table and confirmed later by the histopathological report. This case fulfills all diagnostic conditions mentioned in Farrar's [2] criteria which were published in 1951 (table 1).

KARGER

Fax + 41 61 306 12 34
E-Mail karger@karger.ch
www.karger.com

© 1998 S. Karger AG, Basel
1011-7571/98/0072-0147\$15.00/0

This article is also accessible online at:
<http://BioMedNet.com/karger>

Dr. M. Afifi El-Sayed
PO Box 31956
Sulaibikhat 90809 (Kuwait)

Fig. 1. Ultrasound: Hepatic biliary duct dilation.

Table 1. Farrar's [2] criteria for the diagnosis of primary cystic duct carcinoma

- 1 Tumour restricted to cystic duct
- 2 No evidence of neoplasm involving gall bladder, hepatic or common bile duct
- 3 Histological examination confirming the presence of cancer cells

Fig. 2. Cholecystogram: Complete obstruction of the cystic duct.

Case Report

The patient, a 70-year-old man, was admitted to the hospital with complaints of abdominal pain, fever (38°C) and rigors of 1-week duration. Clinically he was not jaundiced, and an abdominal examination revealed a tender palpable mass in the right upper abdomen. Laboratory investigations showed the following: WBC $14.6 \times 10^9/l$, and normal liver enzymes except for alkaline phosphatase which showed a count of 230 IU/l (normal range 120 IU/l). An abdominal ultrasound showed a distended gall bladder full of sludge with a focal lesion (abscess) in the liver adjacent to the gall bladder bed. The extrahepatic biliary ducts were dilated to 15 mm (fig. 1). The following day an attempt at ERCP failed because of peri-ampullary diverticulum. Calculous obstruction of the common bile duct (CBD) and gall bladder with secondary liver abscess was diagnosed. The patient was managed con-

servatively with intravenous antibiotics (cefotaxime and metronidazole). On the 3rd day of admission both the liver abscess and the gall bladder were percutaneously drained under ultrasound guidance. The result was purulent fluid and both sites grew *Escherichia coli*. The patient's condition improved 72 h later. A cholecystogram via the cholecystostomy tube showed complete obstruction of the cystic duct (fig. 2).

Surgery was done on the 7th day of admission. At laparotomy the common hepatic duct (CHD) was dilated and the tumour was felt at the junction of the cystic duct and CHD radicle. En masse excision of the gall bladder, cystic duct, tumour, part of the CHD and

the supraduodenal part of the CBD was performed, together with all adjacent lymphatics. Malignancy was suspected and was confirmed by frozen sections from the tumour. Biliary drainage was achieved by Roux-en-Y hepatico-jejunostomy. The patient had an uneventful recovery and was discharged from the hospital 8 days after surgery. Repeat ultrasound of the liver showed complete resolution of the liver abscess. The final histopathology showed a poorly differentiated adenocarcinoma of the cystic duct. There was no breach of the peritoneal surface and there was a perineural and not a vascular invasion. There were no lymph node metastases in the excised node, and the gall bladder showed xanthogranulomatous cholecystitis with no signs of malignancy or calculi in it.

Our patient was followed up in the surgical outpatient clinic. He underwent a Hida scan 2 weeks after discharge which demonstrated a patent hepatobiliary system. Two months later our patient was admitted to the CCU with massive myocardial infarction and unfortunately we lost him.

Discussion and Conclusion

The incidence of primary cystic duct carcinoma in autopsy studies is 0.03–0.05% [3]. It constitutes 2.6–12.6% of the extrahepatic biliary malignancy [4]. The criteria for diagnosis of primary cystic duct carcinoma were put forward by Farrar [2] in 1951. Only 27 cases

have been reported so far in the English literature [5] and 17 cases in the Japanese literature [6]. Presentation of primary cystic duct carcinoma is reminiscent of calculous gall bladder disease. Most reported cases were diagnosed either peroperatively when it is suspected on table as in our present case, or postoperatively which is confirmed by the histopathologic report [1, 7]. The first ever reported case of pre-operative diagnosis of primary cystic duct carcinoma was in 1990 from our centre [8], which serves a population of 150,000. The present case is of interest as it is the 2nd in the literature to present with signs and symptoms of compression of the CBD, i.e. Mirrizzi-like syndrome.

The average survival of patients after the excision of primary cystic duct carcinoma is reported to be 27.2 months [6], which is better than that for gall bladder carcinoma (5.8 months) [9] and extrahepatic bile duct carcinoma (3.2–11.4 months) [10]. We believe, therefore, that our patient would have had a favourable prognosis, if he had survived the myocardial infarction, because of the absence of vascular or lymphatic metastases after the en bloc excision of the tumour.

References

1 Kimura H, Takamura H, Arakawa H, Maeda K, Kanno M, Uogishi M, Sodani H, Kanno S: A case of primary cystic duct resected at the relatively early stage. *JJ Gastroenterol Surg* 1992;25:2535–2539.

2 Farrar DAT: Carcinoma of the cystic duct. *Br J Surg* 1951;39:183–185.

3 Philips SJ, Estrin J: Primary adenocarcinoma in a cystic duct stump. *Arch Surg* 1996;98:225–227.

4 Brown DB, Strong R, Gordon J, Hendry E: Primary carcinoma of the extra hepatic duct. *Br J Surg* 1961; 49:22–28.

5 Leonard G, Gary LF, Joseph JS: Case report, cystic duct carcinoma. *N J* 1991;88:337–339.

6 Yamaguchi K, Nishahara K, Tsuneyoshi M: Carcinoma of the cystic duct. *J Surg Oncol* 1991;48:282–286.

7 Radin DR, Chandrasoma P, Ralls PW: Carcinoma of the cystic duct. *Gastrointest Radiol* 1990;15:49–52.

8 Baraka A, Mokhtar NY, Madda JP, Amirrad M, Asfar S: Primary carcinoma of the cystic duct causing obstructive jaundice. *J R Soc Med* 1990;83:746–747.

9 Pallette EW, Harrington RW, Pallette EL: Carcinoma of the extra hepatic biliary system. *Am Surg* 1963; 29:719–723.

10 Van Heerden JA, Judd ES, Dockerty MB: Carcinoma of the extra hepatic bile ducts: A clinicopathological study. *Am J Surg* 1967;113:49–56.