
Case Report

Spontaneous Rupture of the Intrahepatic Bile Duct due to Carcinoma of the Ampulla of Vater: A Case Report

Takebumi Usui, Shunichi Shiozawa, Dal Ho Kim, Akira Tsuchiya, Toshio Masuda, Satoshi Inose, Masaki Aizawa, Kazuhiko Yoshimatsu, Takao Katsube, Yoshihiko Naritaka and Kenji Ogawa

Department of Surgery, Tokyo Women's Medical University, Medical Center East

Abstract

We reported the first case of spontaneous rupture of the intrahepatic bile duct due to carcinoma of the ampulla of Vater. The patient was a 67-year-old male and he visited the hospital in a pre-shock state. He was diagnosed as panperitonitis and an emergency operation was performed on that day. A laparotomy revealed ascetic fluid containing bile to be retained in the abdominal cavity. Intraoperative cholangiography showed a complete obstruction of the lower common bile duct and the rupture of the intrahepatic (S3) bile duct. A lateral segmentectomy of the liver was performed, followed by T-tube biliary drainage. A two-stage checkup/operation was planned for treatment of the obstruction of the common bile duct. Endoscopic retrograde cholangiography (ERC) performed postoperatively revealed a mass in the ampulla of Vater causing a marked stricture of the common bile duct in that region. A pylorus-preserving pancreaticoduodenectomy including a regional lymphadenectomy was performed about 5 months after the initial operation. There has been no other reports documenting a two-stage operation for the resection of a malignant tumor after treatment with bile peritonitis. The avoidance of sepsis and multiple organ failure via surgical treatment directed against bile peritonitis at the appropriate time is considered optimal strategy to save the life of a patient with this condition.

Key Words: Bile peritonitis, Spontaneous rupture of intrahepatic bile duct, Carcinoma of ampulla of Vater

(Received February 27, 2009; Accepted March 9, 2009)

Introduction

A spontaneous rupture of the intrahepatic bile duct has rarely been reported in adults. A rupture of the bile duct is complicated by bile peritonitis, resulting in a mortality of as high as 30 to 50%, even in cases undergoing appropriate surgical treatment. In the majority of such cases, this condition is attributable to biliary stones which become impacted in the bile duct and produce a precipitous elevation of pressure in the biliary tract. This can lead to rupture of a fragile portion of the bile duct due to a shock or other causes.

This report describes an extremely rare case of bile duct rupture, which developed in the form of bile peritonitis clinically and upon examination of the biliary duct, was considered etiologically attributable to a carcinoma of the ampulla of Vater.

Case Report

The patient was a 67-year-old male who had experienced epigastralgia for approximately one week prior to the visit to this hospital due to gradually increased abdominal pain. At the time of the first consultation, neither anemia nor jaundice was noted, but the abdomen was generally distended and a marked epigastric tenderness and muscular guarding were present on physical examination. The patient’s vital signs were: blood pressure, 92/56 mmHg; pulse rate, 112 beats/min; and body temperature, 38.5°C. The patient was also in a pre-shock state. The abnormal laboratory data included: hemoglobin, 19.6 g/dL; WBC, 2700/mm³; total bilirubin, 1.6 mg/dL; and serum amylase, 321 IU/L. An abdominal x-ray demonstrated an elevation of right side of the diaphragm but no evidence of free air in the abdominal cavity and revealed a distended pattern of small bowel gas. Abdominal computed tomography (CT) scan disclosed a moderate amount of ascites retained in the perihepatic regions, enlargement of the gallbladder and a slight dilatation of the both intra- and extrhepatic bile ducts (Fig. 1). From these findings, panperitonitis due to a perfora-
tion of the upper gastrointestinal tract was suspected and an emergency operation was performed on that day. A laparotomy revealed approximately 600 mL of ascitic fluid containing bile to be retained in the abdominal cavity and the greater omentum covered the anterior aspect of the stomach and the left hepatic lobe. Detachment of the adherent greater omentum revealed a large amount of turbid bile collected in the omental sac of which, the inner surface was covered with fur. A detailed examination of the stomach and small intestine revealed no obvious perforation. A rupture of the biliary tract due to elevated pressure in the biliary tract was also suspected, on the other hand, as the gallbladder was tensely bulging along with the distended common bile duct. In view of this, an intraoperative cholangiography was performed using 20 mL of meglumine sodiumamidotrizoate (Urografin) injected through the stump of the cystic duct after a cholecystectomy. Further examination showed a complete obstruction of the lower portion of the bile duct although no rupture was identified. Following an injection with 20 mL of the dye test reagent, indigocarmine, into the biliary tract, the leakage of the dye from the surface of lateral segment (Segment III) of the liver was noted, so that the leakage point was identified as the site of rupture of the biliary tract. The liver tissue at that site was fragile secondary to an infection; therefore, closure of the rupture using sutures was judged to be hardly feasible. A lateral segmentectomy of the liver was thus undertaken, followed by a T-tube biliary drainage in the common bile duct. A two-stage checkup/operation was planned for treatment of the obstruction of the lower part of the bile duct and this initial surgical stage was completed.

In the resected lateral segment of the liver, fur was noted adherent to the lateral margin of the liver and there was a perforation of the intrahepatic bile duct (Fig. 2). A histopathological examination disclosed a diffuse infiltration, with neutrophils centered at the site of perforation of the bile duct along with a micro-abscess and a large number of Candida albicans spores (Fig. 3). The patient subsequently developed a concurrent intraperitoneal abscess due to Candida albicans, which necessitated peritoneal drainage over the ensuing period of approximately 5 months. Endoscopic retrograde cholangiography (ERC) performed postoperatively revealed a mass in the ampulla of Vater causing a marked stricture of the common bile duct in that region (Fig. 4). The mass was diagnosed as an adenocarcinoma based on the biopsy results.

Based on the clinical, surgical and histopathological results, the present case was finally diagnosed as an

![Fig. 1 An abdominal CT scan demonstrates a moderate amount of ascitic fluid in the perihepatic spaces (arrows), enlargement of the gallbladder and slightly distended intra- and extrahepatic bile ducts.](image1)

![Fig. 2 The resected specimen of the liver shows fur adherent on the lateral margin and perforations (arrows) of the intrahepatic bile duct.](image2)

![Fig. 3 A histopathological examination shows diffuse neutrophil infiltrates principally in the region of a perforated bile duct and micro abscess and a large number of Candida albicans spores (arrows) are noted.](image3)
obstruction of the biliary tract due to a carcinoma of the ampulla of Vater and a concurrent Candida albicans infection of the biliary tract which led to fragility of the hepatic lateral segment tissue and the consequent rupture of the intrahepatic bile duct, which became complicated by bile peritonitis. A pylorus-preserving pancreatico-duodenectomy including a regional lymphadenectomy was conducted about 5 months after the initial operation. The carcinoma of the ampulla of Vater measured 4.0 x 2.5 cm in diameter and was classified as T3 N1 M0 Stage II B according to the UICC staging criteria. The postoperative course was uneventful and the patient was discharged on the 36th day after the second operation.

Discussion

An obstruction of the biliary tract with a gallstone or tumor usually leads to obstructive jaundice without any associated symptoms while a spontaneous rupture of the bile duct is a rare consequence. Therefore, a rupture of the biliary tract is likely to occur due to a plurality of causes, e.g., biliary tract infection, disturbance of blood flow in the bile duct wall due to intramural thrombosis, pancreatic juice reflux, or malformation of the biliary tract associated with elevated pressure in the biliary tract. The rupture of the gallbladder or extrahepatic bile duct is relatively common, whereas rupture of the intrahepatic bile duct covered by the surrounding parenchymal tissue is a rare manifestation.

A spontaneous rupture of the intrahepatic bile duct was first reported by Smith in 1926 and there have been 40 cases of this disorder reported among the English publications to date, according to an extensive search of the literature. The most frequent cause of this type of rupture is biliary stone impaction and other causes included benign diseases such as Caroli's disease (1 case) and cholangitis (1 case) and malignant disorders such as carcinoma of the bile duct (2 cases), gastric cancer (2 cases), carcinoma of the gallbladder (1 case) and carcinoma of the head of pancreas (1 case). The predominance of a biliary stone as the cause of rupture is thought to be due to the fact that a biliary tract infection is liable to become complicated by cholangitis and once stone impaction occurs, pressure in the biliary tract rises precipitously in contrast to cases of obstruction by a tumor. The present case is the first to be reported in which a spontaneous rupture of the intrahepatic bile duct occurred due to a carcinoma of the ampulla of Vater.

Intrahepatic bile duct ruptures occur in the left lobe of the liver in more than 80% of the reported cases. This may be because the left lobe parenchyma is thinner in comparison to the right lobe, so that the bile duct directly subjacent to the hepatic capsule is susceptible to rupture following a precipitous elevation of pressure in the biliary tract. When a biliary stone is the cause, removal of the stone and biliary drainage should be performed. It should be determined intraoperatively whether the rupture of the liver is to be closed by sutures or if a relevant partial hepatectomy is necessary. In earlier reported cases, the prognosis was grave because of a delayed diagnosis resulting in septic shock or multiple organ failure. With the advances in diagnostic imaging and surgical treatment techniques, the number of survivors from the disease among treated patients has been increasing in recent years. Kang et al reported gratifying results attained by a one-stage operation which included biliary drainage and a hepatectomy in 4 of 9 cases. This procedure may thus be the ideal surgical intervention by which the infective foci can be eliminated. Saving the patient's life is the principal objective of an emergency operation in a patient with a disease with high mortality as in the present case and the surgeon may choose a second-stage treatment especially for patients with the disease attributable to malignancies.

In the present case, an intrahepatic cholangiography did not show any outflow of contrast medium into the duodenum and demonstrated a marked stricture of the bile duct, so that a carcinoma of the lower common bile duct was initially suspected based on the imaging results. Because the site of the rupture remained unclear, however, a dye test reagent was injected into the biliary tract demonstrating leakage of the dye from the lateral segment of liver, thereby allowing identification of the site of the rupture. This method is useful when the results of intrahepatic cholangiography are insufficient to determine the rupture site. A left lateral segmentectomy was performed concomitantly to eliminate the infective foci during the initial operation, but an intraperitoneal abscess developed postoperatively, requiring an interval of about 5 months until the second-stage operation was performed. In cases where malignant tumors account for the condition, as in the present case, it is important to determine the stage of the tumor and decide upon the
appropriate surgical indication without delay, since there is the possibility of tumor cell dissemination due to the rupture of the duct and because the stage of tumor determines the prognosis. There has been no other reports documenting a two-stage operation for the resection of a tumor and the surgeon would need to attempt to surgically eliminate the causative disease as long as there is no obvious factor favoring non-resection.

The therapeutic results have improved thanks to the advances in diagnostic imaging and surgical techniques and yet mortality from bile peritonitis remains high. The avoidance of sepsis and multiple organ failure via surgical treatment directed against bile peritonitis at the appropriate time is considered optimal strategy to save the life of a patient with this condition.

References