MIRIZZI SYNDROME, A CASE REPORT

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Abstract

Mirizzi syndrome causing obstructive jaundice is rarely seen in Africans. Obstructive jaundice due to gall bladder calculi is even less common in young African men. A case of this condition in an adult male is presented. More common precipitating factors for lithiasis such as haemolytic diseases, sickle cell disease and obesity were not found. Cholecystojejunostomy was performed for suspected periampularly tumour but jaundice recurred within 9 months. Diagnosis was suspected after cholangiography and confirmed at surgery. The biliary ducts were dilated with complete stenosis of the distal CBD. The cystic duct was completely stenosed. No calculi were found in the cystic duct or the Hartman's pouch. Choledochojejunostomy was finally performed and the patient recovered.

Introduction

The classical Mirizzi Syndrome (P. L. Mirizzi 1948) is characterised by a cholecystocholedochal fistula arising from calculi in the cystic duct eroding into the common hepatic duct. The gall bladder eventually collapses due to fibrosis. The terminal bile ducts becomes dilated. A calculus is usually found at the cystic duct or in the Hartman's pouch. This presents an unusual and specific cause of obstruction of the terminal bile ducts. Patients present with obstructive jaundice, and cholangiography shows narrowing of the common bile duct at the portal hepatis, which can have the appearance of a cholangiocarcinoma. The true pathology is eventually identified at surgery. The operation is often difficult because of severe inflammation and fibrosis. For cholecystocholedochal fistula, a cholecystoduodenostomy is a safer operation. In more complex situations a choledochoplasty (using part of the gallbladder wall) or a choledochojejunostomy Rouxen-Y may be required. Preoperative ERCP if obtainable is helpful in deciding the best type of operative approach.

Case Report

Mr. CW, a 39-year-old male, non-diabetic was referred to me with a one-month history of jaundice, generalised body itch and loss of weight. He gave history of heartburn but no nausea or vomiting. He would get constipation followed by episodes of diarrhea. He had lately lost appetite. His stools were pale and urine was deep yellow. Past medical history was non-significant. There was no history of jaundice prior to this illness. He does not smoke but drinks occasionally. He works as a technician in a photocopying business.

On examination, Mr. CW was in good general condition. He weighed 65kg. He was 170 cm tall. He had a generalised papular rash from scratching. He was deeply jaundiced. He was not anaemic. There were no palpable lymphnodes. The abdomen was soft with no ascites. The liver and spleen were not enlarged. The gallbladder was palpable and mildly tender (positive Murphy's sign). The rest of the examination was normal.

Investigation

The following investigations were done.

Blood tests revealed a Hb of 14.3g/dl, WBC 4,200mm^3, ESR 48mm/hr, platelets 399,000. There were no malaria parasites. Random blood sugar on first admission was 6.3mmol/l.

Urinalysis showed some bilirubin but no urobilinogen, proteins, sugar, blood or pus cells.

Liver function tests showed high total bilirubin of 221umol/L with direct bilirubin of 141umol/L. Alkaline phosphatase was raised 935lU/L. The enzymes (SGOT, SGPT, Gamma GT) were also moderately elevated. Hepatitis BsAg, Hepatitis C and HIV tests were negative. The prothrombin time was within normal. The urea, creatinine and electrolytes were within normal range.

Abdominal ultrasound was reported to show a dilated but normal gallbladder, the intrahepatic billiary ducts were normal but the CBD was dilated. CT Scan confirmed a dilated CBD. The pancreatic head was reportedly prominent. There was also a report of vague masses in the para aortic region, possibly lymphnodes. Scrotal ultrasound scan showed normal testis. ERCP was not done. Cholangiography (oral, intravenous or percurtaneous was not done).

A clinical diagnosis of obstructive jaundice possibly due to a periampularly tumour was made. He was scheduled for operation. At operation, the gallbladder was distended, with no calculi. CBD was palpated and no calculi were palpable. The head of pancreas was rather nodular but not enlarged. A biopsy was taken. The ampula of Vater was open and draining clear fluid. There were no calculi.
A billiary diversion was done with a cholecystojejunostomy. Post-operative recovery was uneventful. The jaundice cleared completely within about 20 days. Histology from the pancreatic biopsy showed normal pancreatic tissue with no evidence of malignancy. A follow up CT scans of the abdomen were done three months later, they were normal.

FOLLOW UP

Nine (9) months after the operation, he came back to see me with recurrent jaundice, fever and itch. There were no abdominal masses or organomegaly. Liver function tests showed marked elevation of total and direct direct bilirubin and all the liver enzymes especially alkaline phosphatase and gamma and gamma GT. Repeat spiral CT scans (fig I and II) were reported to show dilated CBD. The pancreatic head was normal.
He was referred to a gastroenterologist for opinion, ERCP and cholangiography. ERCP was not done. A percutaneous transhepatic cholangiography (PTC) (fig. III and IV) were reported to show:
"Choledochojejunostomy, which shows good drainage into the jejunum. There are multiple filling defects in the gallbladder, which are consistent with air bubbles. The common hepatic duct and CBD are dilated. There is a smooth stricture at the distal end of the CBD, consistent with benign stricture".
He was prescribed a cause of antibiotics after which he appeared to improve and jaundice partly cleared. However, jaundice and itch quickly returned. He was also loosing weight fast.

He returned to see me four months later. Meanwhile he had rapaciously chewed sugarcane and drunk a lot of sugarcane juice in an attempt to "cure" as he had been advised by well-wishers. A review of the cholangiograms gave away the diagnosis. He consented for operation.
Pre-operative investigations revealed high blood sugar (25.4mmol/l). The pancreatic enzymes were within normal limits. The LFTS had not changed much from previous figures shown above. The prothrombin time was within the normal range. The blood sugar was easily controlled with insulin.
At re-exploration, the liver, spleen and pancreas were found to be normal. The gall bladder was thickened and contracted. The common hepatic duct and the common bile ducts were grossly distended,

forming a large sac. The cystic duct was thin and tough. Attempts to cannulate it failed. The ballooned sac was opened to review distended CHD, CBD and both the right and left hepatic ducts. There was a lot of sludge, which was cleaned off. The CBD was completely blocked distally (the retroduodenal portion).

Choledochojejunostomy was performed. The remnant of the collapsed gall bladder was removed. Post operatively he recovered without any problem. The jaundice cleared in a few days.

He was back to work in four weeks. I have followed him up for eight months now. He is fine.

Discussion

Lithiasis in general is uncommon in indigenous African men. We see more gall bladder stone disease in immigrant population of Asian and Arabic descent. In populations where lithiasis is common, a calculi in the bile duct may lie dormant for many years and only come to light because of an episode of pain, jaundice, or cholangitis. They may also be discovered by abdominal ultrasonography during routine scanning. Between 8 and 15 per cent of patients with stones in the gallbladder also have stones in the ducts (Julian Britton et al) [1.] Females are more affected than males. The incidence increases with age. Mirizzi syndrome is more common in the elderly, but any patient with cholelithiasis is at risk (Csendes, 1989) [2]. Our patient was 38 years old man at presentation. Three cases aged 49, 58 and 59 years have been reported by Toufeeq Khan et al [3]. Primary stones form within the bile duct. They are usually bilirubinate stones of the soft brown type, and they are associated with biliary stasis due to obstruction, infection and the presence of foreign bodies such as food. In the Orient they may be caused by infection, sometimes associated with parasites within the biliary tract. However, most common duct stones originate in the gallbladder and migrate through the cystic duct into the common bile duct. These secondary stones consist mostly of cholesterol and often grow in size within the duct. We have not established the cause of lithiasis in him.

Mirizzi Syndrome has no consistent or unique clinical features that distinguish it from other more common forms of obstructive jaundice. Symptoms of recurrent cholangitis, jaundice, right upper quadrant pain and abnormal liver function tests may or may not be present (Strugnell, 1995**)** [4.] Patient may present acutely with pancreatitis or cholecystitis. Our patient presented with jaundice. He had no pain or fever initially. These symptoms set in later with increasing derangement of the liver function tests. We suspect he became diabetic because of taking too much sugar (sugar cane).

The most important investigation is ultrasound examination of the liver, the bile duct, the gallbladder and the pancreas. The ultrasonographer need only decide whether or not the bile ducts are dilated. The normal common bile duct should not be greater than 7mm in diameter when measured on ultrasound. Signs of dilatation of the biliary duct system including the CHD, CBD and abrupt narrowing of the CBD are good indicators. Impacted stone may or may not be visualized. There was biliary dilatation especially of the CHD and CBD in our patient. Intrahepatic ducts were reported normal. CT scan has a limited place in the imaging of common duct stones. The ultrasound examination may raise the possibility of a malignant obstruction, and a CT will very reliably detect dilation of the ducts and identify the site and the cause of an obstruction. In this case CT scan actually showed dilated CBD (**figs I &II**). No calculus was seen. The pancreatic head was initially reported to be prominent with vague masses in the para aortic region (nodes?). This error led to a wrong operation being performed.

Cholangiography demonstrates the narrowing of the distal CHD and of the proximal CBD as well as, in some instances, the dilatation of the proximal biliary duct system. Cholangiography gives excellent anatomical visualization. Cholangiography was done late in this case. Percutaneous transhepatic cholangiography (PTC) was reported to show dilated CHD and CBD. There was a smooth stricture at the distal end of the CBD, which was consistent with benign stricture (**figs III & IV**). At this point the diagnosis was suspected.
Liver function tests are usually abnormal. Apart from elevated bilirubin levels, the only other distinguishing abnormality is the high levels of alkaline phosphatase and gamma GT. However, serology for viral hepatitis must be done to rule out concurrent infection. It is also prudent to do pancreatic enzymes. In this case, liver function tests were consistently and progressively abnormal.

An attempt has been made at classification of the Mirizzi syndrome (Gregory B.Snynder) [5] and (Toufeeq Khan)[3]. This classification is more of a division of the same condition in it's various stages, so that type I describes the condition before fistula formation, while type II – IV, a fistula is present but is of different size in relation to the size of the CBD (Gregory B. Snynder) [5]. I did not attempt to classify this case.

Most authors agree that the treatment of choice for Mirizzi syndrome is surgical (Julian Britton et al, 1994; Strugnell, 1995; Pemberton, 1997; Toscano, 1994;) [1,4,6, 7]. However, electrohydraulic lithotripsy

(EHL) was reportedly successful in 38 of 38 patients with known Mirizzi syndrome (Seitz et al, 1998) [8.] The technique used in surgical treatment will depend on the stage of the patient's condition, hence, the attempt to classify Mirizzi syndrome. The stage may however, not be obvious until the procedure has begun. That is why pre-operative cholangiography is indicated to visualize the anatomy and the stage of the disease i.e. presence or absence of fistula. When there is compression without a fistula it is generally treated with cholecystectomy and choledochoduodenostomy. When there is a fistula formation a biliary enteric bypass via Roux-en-Y choledochojejunostomy is the most suitable. Note however, that intrahepatic lithiasis has been reported after cholecystojejunostomy (C. Lazaridis) [9]. This patient initially had cholecystojejunostomy done for biliary bypass. By then the cystic duct was patent and draining, but later blocked as fibrosis progressed. This has been shown to be the progressive outcome in most of these cases (Gregory B. Snynder) [5]. Definitive procedure of Roux-en-Y choledochojejunostomy was carried out after the diagnosis became clear.

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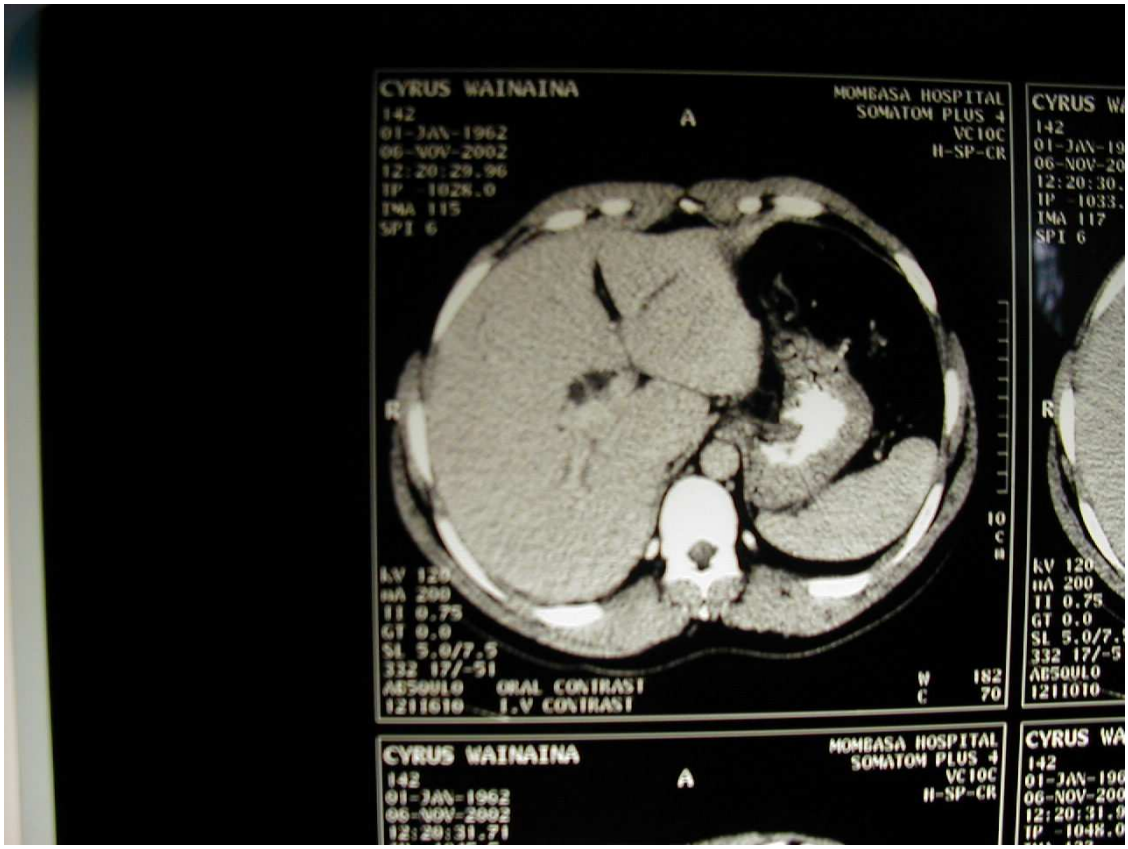

Figure I

This is a CT scan of the abdomen that shows normal liver and pancreas. Please note the dilated CBD.

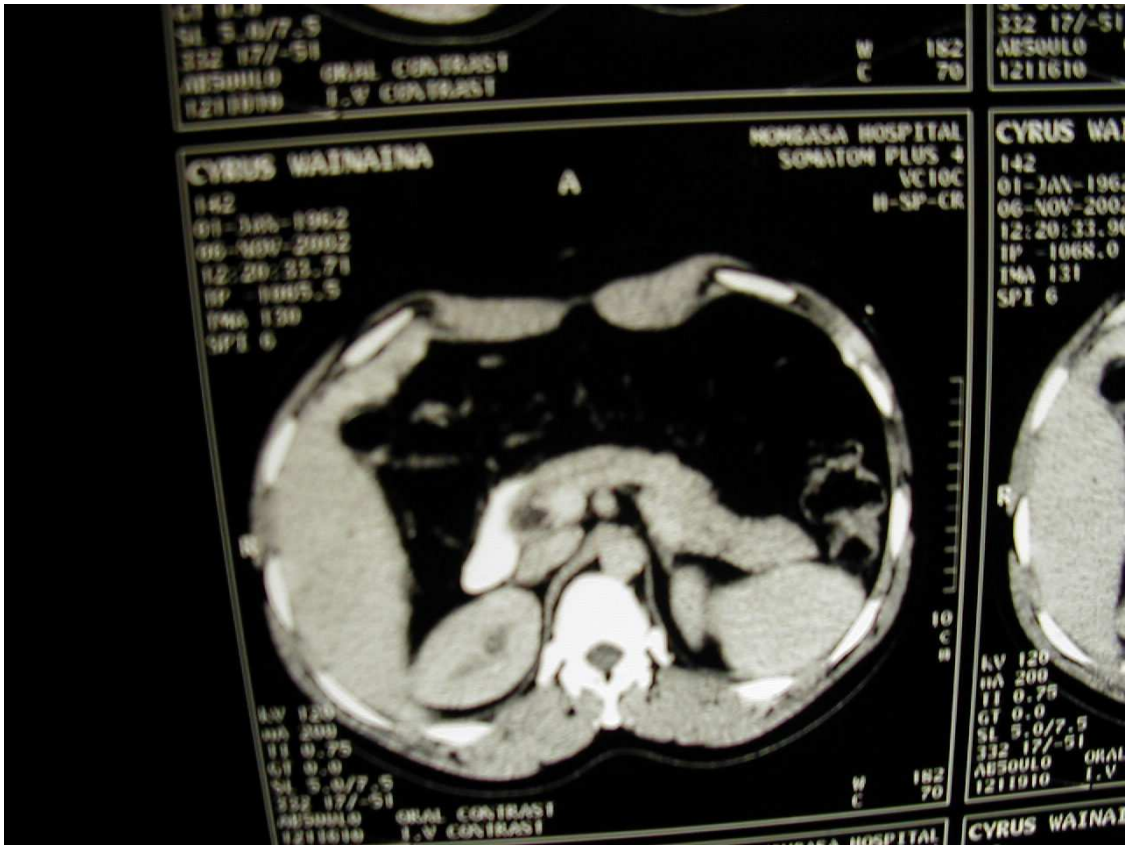

Figure II

This is a CT scan of the abdomen. This section clearly shows you a normal pancreas. Please note again the dilated CBD (in the pancreatic head).

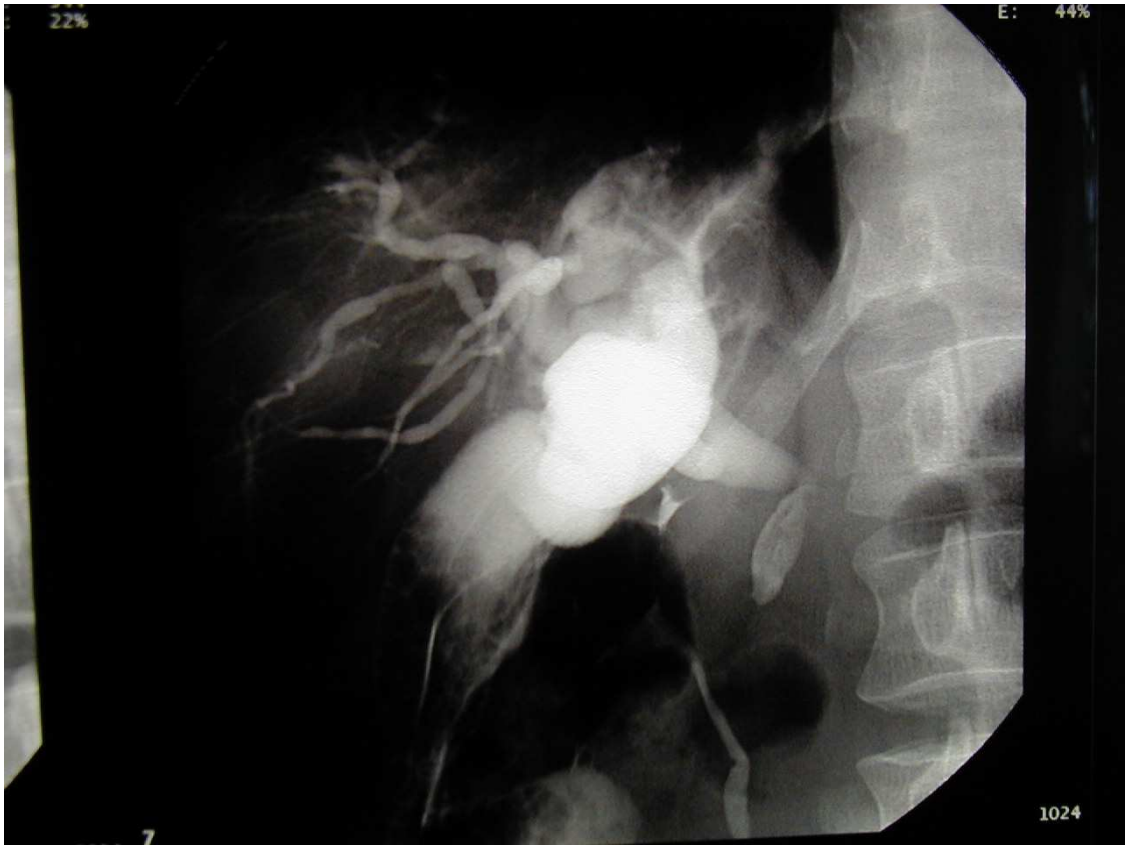

Figure III

This is a percutaneous transhepatic cholangiogram. This film shows dilated intrahepatic, common and hepatic duct. Note the stricture at the distal end of the CBD.

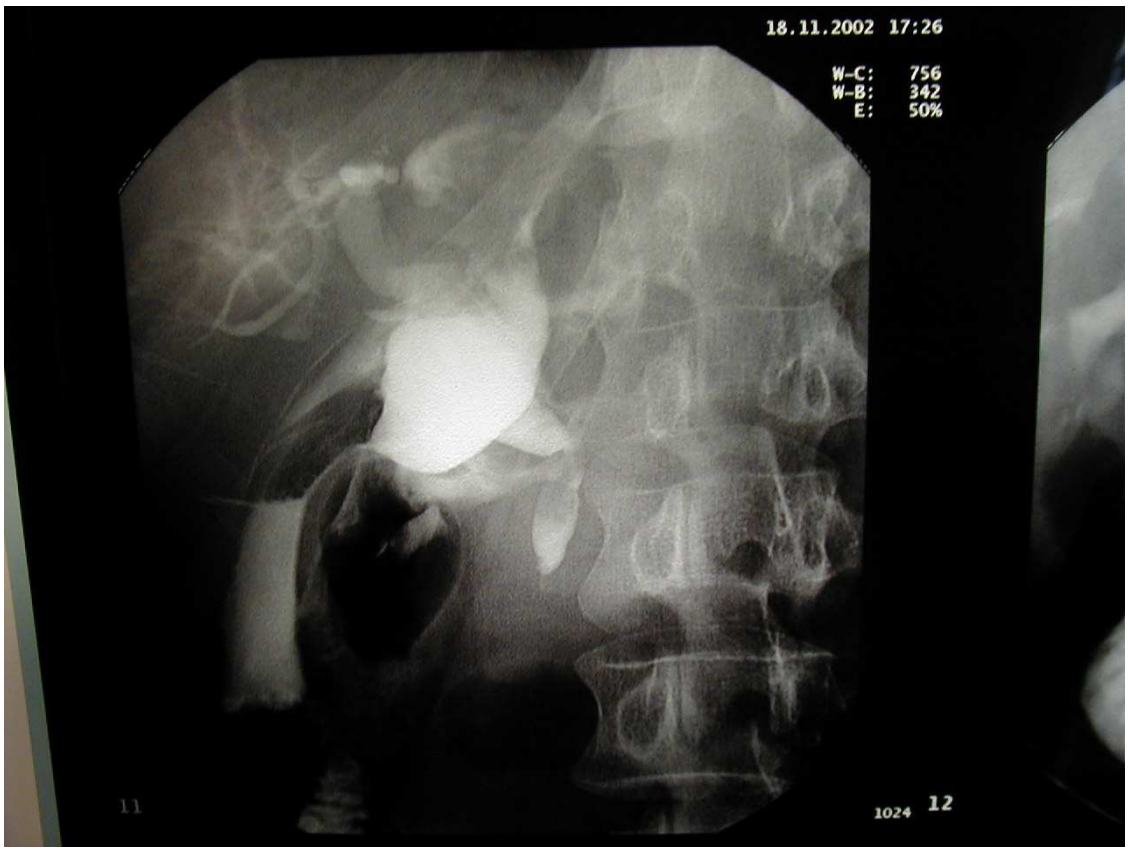

Figure IV

This is another cholangiogram clearly showing a narrowed cystic duct.

Figure Legends

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