Intrahepatic stones might mimic cholangiocarcinoma on ERCP

Babur Javaid, MRCP, Riaz M. Faizallah, FRCP, PhD

Intrahepatic stones are defined as stones in bile ducts proximal to the junction of the left and right intrahepatic ducts. This condition is rare in western countries where it tends to occur in conjunction with Caroli’s syndrome, biliary strictures, or any ductal abnormality that results in biliary stasis and infection. This is a report of a case of intrahepatic stones with no such underlying abnormalities and which caused misleading cholangiographic appearances.

CASE REPORT

A 70-year-old white man undergoing thrice weekly hemodialysis for chronic renal failure secondary to hypertension presented with a few years’ history of recurrent right upper quadrant abdominal pain and intermittent vomiting for the previous 5 months. He developed jaundice, pale stools, and rigors a few weeks before admission. On examination he was afebrile, icteric, tender over right hypochondrium, and had slight hepatomegaly. There had been gradual deterioration of biochemical parameters of liver function over the previous few months. At admission, these were as follows: bilirubin 79 µmol/L (5-21), alkaline phosphatase 1160 IU/L (35-105), aspartate aminotransferase 34 IU/L (0-40), alanine aminotransferase 73 IU/L (0-40), and gamma-glutamyltransferase 556 IU/L (11-50). International normalized ratio and white blood cell count were normal.

US revealed dilated intrahepatic ducts and common bile duct (CBD) to the level of the cystic duct, possibly with a stone in CBD at this level. The CBD was of normal diameter proximal to this point. Endoscopic retrograde cholangiography (ERC) revealed what appeared to be a smooth narrowing of the bile duct near the hilum with dilatation of left intrahepatic ducts. The right intrahepatic duct system was not opacified. These findings were highly suggestive of a hilar malignant stricture (Fig. 1). A 12-cm 10F plastic biliary stent was inserted (Fig. 2). CT of the upper abdomen was scheduled and treatment continued with intravenous antibiotics. Unfortunately, the patient developed rigors and
high-grade pyrexia after ERC. A diagnosis of biliary sepsis was made and treatment was continued with appropriate antibiotics. A plan for further management including a repeat ERC and replacement of the stent or decompression of the obstructed right biliary system via a percutaneous approach was discussed with the family. They declined further interventions, both investigative and therapeutic, including further dialysis, in view of the patient’s long-term co-morbidity of dialysis-dependent chronic renal failure and the likely diagnosis of cholangiocarcinoma. The patient’s condition deteriorated with biliary sepsis and worsening renal function and he passed away peacefully over the next 48 hours.

At post mortem the right hepatic duct contained 3 large stones (Fig. 3) that caused considerable distortion of the architecture of the duct and compressed the left hepatic duct. The stones were dissected free with some difficulty. No ductal stricture was evident. There were no stones in the left hepatic duct. The proximal end of the stent had slipped distal to the confluence of the cystic duct with the CBD. The gallbladder was normal and did not contain stones. No congenital anatomical anomalies were found.

DISCUSSION

Hepatolithiasis (intrahepatic stones), defined as the presence of stones in the bile ducts proximal to the confluence of right and left hepatic ducts, affects predominantly the left hepatic ducts. This disease is uncommon in western countries and is mainly associated with conditions that cause biliary stasis such as inflammatory, postoperative, or neoplastic strictures, bile duct stones, and cystic abnormalities. The majority of these stones are composed of calcium bilirubinate, which result from precipitation of water insoluble to unconjugated bilirubin. This unconjugated bilirubin is believed to be released by deconjugation of bilirubin glucuronides in bile by the action of beta-glucuronidase producing enteric bacteria (Escherichia coli, Clostridium and Bacteroides species). Bacteria are found in the bile in nearly all patients with intrahepatic stones. Intrahepatic stones are prevalent in east Asia where they are thought to be related to infestation with parasites such as Clonorchis sinensis. The highest prevalence is found...
in Taiwan. The frequency of co-existing cholangiocarcinoma in patients with intrahepatic stones has been reported to be 1.5% to 9.4% in Japan but data on the utility of EUS for the diagnosis of cholecystolithiasis and helping in planning for further management.

On ERC, intrahepatic stones may appear as filling defects in contrast filled bile ducts. However they can easily be missed if bile ducts are not opacified due to obstruction caused by stones or strictures distally. When there is non-opacification of the bile duct, further delineation of the biliary system proximal to the cut-off point can be attempted by passing a guidewire through the stricture followed by a cannula and then injection of a contrast medium. If this strategy fails, percutaneous transhepatic cholangiography (PTC) can be useful. In the case of hilar obstruction, MRC or EUS can help in differentiating stricture from a stone.

In our case stones in the right hepatic duct caused non-opacification of the ductal system proximally. Extrinsic compression of the left hepatic duct gave the appearance of a stricture suggestive of a cholangiocarcinoma. Other conditions, which can cause similar cholangiographic appearances, include a dominant stricture in primary sclerosing cholangitis, extrinsic compression from metastatic liver disease, chylymphadenopathy at the level of the porta hepatitis and postsurgical biliary strictures. This indicates that the ERC appearance of intrahepatic stones ought to be interpreted with caution. Apart from the management strategy adopted by us, more details about the non-filling system could have been obtained via PTC, which would have also provided an opportunity to decompress the obstructed right-sided biliary system. A previous study has shown that it is not necessary to obtain drainage of both liver lobes in patients with malignant hilar strictures. It is believed that biliary sepsis in our patient resulted from inadequate biliary drainage. The stent should have been advanced deep enough to drain the left or right intrahepatic biliary tree rather than being left at the level of bifurcation. The distal end of the stent came to lay against the duodenal wall (Fig. 2). The biliary stent had slipped further down, as demonstrated on post mortem.

To our knowledge, appearances of a left hepatic duct stricture secondary to external compression by stones in the right hepatic duct or vice versa have not been reported. The importance of appropriate drainage of the obstructed biliary system once it has been opacified with contrast material should also be stressed.