CLINICAL PRACTICE

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More than simple hepatic cysts

Daniela Tabacelia[1], Mădălina Ilie[1], Alina Constantin[1], Anca Macovei Oprescu[2], Gabriel Constantinescu[1]

Abstract: *Caroli diseaseis a rare congenital disorder that classically causes saccular dilatation of the bile ducts. The complications of Caroli include choledochal cysts with recurrent cholangitis, abscess formation, septicaemia, intrahepatic lithiasis and amyloidosis.We report a rare case of a young female with Caroli disease pointing out the intrahepatic lithiasis as a rare complication of the disease.*

Learning points

- *Caroli disease is an uncommon condition that should be considered in the differential diagnosis of hepatic essential cysts.*
- *Clinically, it is characterized of recurrent episodes of fever and pain.*

The correct and early diagnostic is important because of the different complications and treatment unlike the essential hepatic cysts.

Keywords: *Caroli disease, intrahepatic lithiasis, endoscopic retrograde cholangiopancreatography*.

CASE REPORT

A 28 years old female presented to the emergency department with abdominal pain and fever after an ERCP procedure performed 2 weeks ago. From her disease history we retain an acute pancreatitis that was classified Balthazar B, one year ago, assumed to be caused by biliary main duct lithiasis; at that moment her initial biology showed ASAT 156U/L, ALAT 126U/L, lipase 2,398U/L, bilirubin 32 mmol/l and white cells 14,590/mmc; there was no sign of complication (acute cholangitis) that would have required a surgical treatment in emergency. Her family history was inconclusive for any specific digestive condition.

In our hospital, on physical examination her body temperature was reaching 38^0C, the heart rate 110 bpm, and blood pressure 110/70mmHg; on palpation she presented mild tenderness on epigastric and right quadrant; the usual blood tests showed white cell count with neutrophils raised, hepatic cytolysis and cholestasis until 5 times normal value. Next step was performing an abdominal ultrasound which revealed hepatic cysts (dilation of intrahepatic bile ducts) with liver hyperechoic images with acoustic shadow suggestive of intrahepatic lithiasis (Figures 1 A and B). A CT scan and a MRI were performed and revealed a normal pancreas without Wirsung dilatation, the biliary main duct with a 7 mm diameter without stones inside, and normal gall bladder without signs of cholecystitis but with beaded intrahepatic biliary duct dilatation until 18

[1] Department of Gastroenterology, Clinical Emergency Hospital Bucharest
[2]Department of Gastroenterology, Agrippa Ionescu Hospital

mm alternated with normal caliber ducts with numerous images of stones inside with a 12 mm maxim diameter. The dilated biliary canaliculi wall presented signs of cholangitis. All clinical and paraclinical data were typical for a Caroli disease with intrahepatic lithiasis and secondary cholangitis. The patient received treatment with urso-deoxy cholic acid (UDCA), antibiotics, anti-inflammatory drugs and pain medication followed by the resolution of the inflammatory episode.

Figure 1A

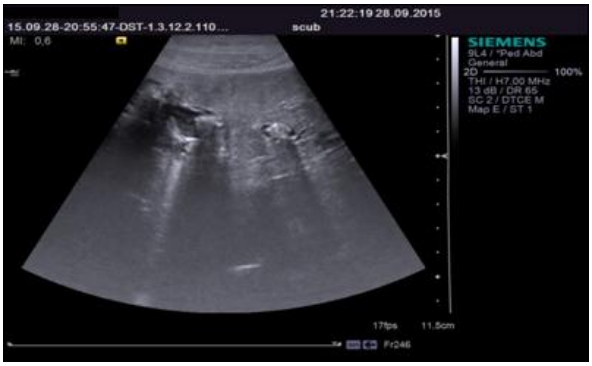
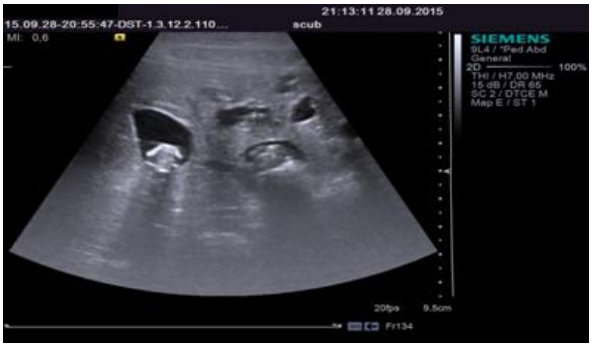


Figure 1B: Abdominal ultrasound revealing hepatic cysts with intrahepatic lithiasis



DISCUSSION

The hallmark of Caroli disease is intrahepatic duct dilatation [1]. Patients with Caroli disease are usually presenting to the hospital, when they develop complications as the result of biliary stasis, which leads to stone formation and infection. The stones are brown pigmented stones, composed of inspissated bile[1].

Caroli disease has two forms, one associated with congenital hepatic fibrosis and a simpler form occurring alone. The former, called Caroli's syndrome is associated with portal hypertension, and it's complications including splenomegaly, hematemesis and melena. Caroli disease is also associated with liver failure and polycystic kidney disease. The cause appears to be genetic; the simple form is an autosomal dominant trait while the complex form is an autosomal recessive trait.[2] Females are more prone to Caroli disease than males.[2] Family history may include kidney and liver disease due to the link between Caroli Disease and ARPKD (Autosomal recessive polycystic kidney disease). The symptoms include fever, intermittent abdominal pain, and hepatomegaly. Occasionally jaundice occurs.[3] Laboratory tests indicate cholestasis and hepatocytolysis associated with an inflammatory syndrome in the acute stages.

Caroli disease usually occurs in the presence of other diseases, such as autosomal recessive polycystic kidney disease, cholangitis, gallstones, biliary abscess, septicemia, liver cirrhosis, renal failure, and cholangiocarcinoma (7% affected).[2] People with Caroli disease are 100 times more at risk for cholangiocarcinoma than the general population.[3] Modern imaging techniques allow the diagnosis to be made more easily and without invasive imaging of the biliary tree.[4] Abdominal ultrasound can detect saccular or fusiform dilation of the bile ducts. Images taken by CT-scan or MRI will show enlarged intrahepatic (in the liver) bile ducts due to ectasia; cholangiography is the best approach to show the enlarged bile ducts as a result of Caroli disease.

The treatment of Caroli's disease depends on the clinical features and the location of the biliary abnormalities. Cholangitis is treated with appropriate antibiotics. In case of intrahepatic cholelithiasis litholytic therapy with urso-deoxy cholic acid (UDCA) is indicated[5]. When the ductal abnormalities are localized to one lobe, lobectomy relieves symptoms and appears to remove the risk of malignancy. In case of diffuse involvements of both lobes of liver, treatment options include conservative management, endoscopic therapy (sphincterotomy for clearance of intra-hepatic stone), internal biliary bypass procedures and in carefully selected cases liver transplantation[5].

References:

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