Case 1077

Eurorad

Caroli's disease

Published on 04.07.2001

DOI: 10.1594/EURORAD/CASE.1077
ISSN: 1563-4086
Section: Abdominal imaging
Imaging Technique: MR
Imaging Technique: MR
Imaging Technique: MR
Case Type: Clinical Cases
Authors: S. Cakirer
Patient: 19 years, female

Clinical History:

A 19-year-old female patient who had recurrent cramplike right upper quadrant pains more than three years referred with severe abdominal pain, fever and mild jaundice, biochemical analysis revealed high levels of serum alkaline phosphatase, gammaglutamyltransferase, and mildly elevated levels of serum SGOT and SGPT enzymes.
Imaging Findings:

A 19-year-old female patient who had recurrent cramplike right upper quadrant pains more than three years referred with severe abdominal pain, fever and mild jaundice, biochemical analysis revealed high levels of serum alkaline phosphatase, gammaglutamyltransferase, and mildly elevated levels of serum SGOT and SGPT enzymes. MRI study of the abdomen with SE T1, FSE T2 weighted sequences on three planes, and MRCP (MR cholangiopancreatography) study by maximum intensity projection reconstruction of heavily FSE T2 weighted coronal images (TR/TE: 18762/240 msec) were performed on a 1.5 T MR scanner. MR pictures revealed multiple hepatic cystic dilatations communicating with the biliary tree, bile sludge causing fluid-fluid levels and bile calculi. Extrahepatic bile tracts and main biliarl duct were not much dilated as intrahepatic bile tracts.
Discussion:

Caroli's disease, also called communicating cavernous ectasia of intrahepatic ducts, is a rare congenital abnormality characterized by multifocal segmental dilatation of the intrahepatic bile ducts. Caroli's disease appears to be autosomal recessively inherited in most cases. Perinatal hepatic arterial occlusion or hypoplasia -aplasia of fibromuscular wall components, causing dilatation of primitive biliary ducts may play role in development of Caroli's disease. It is most commonly seen in childhood and second to third decades of life. Males and females are equally affected. There are two forms of the disease, one associated with congenital hepatic fibrosis (type II) and the other form occurring alone (pure form, type I). Although classically known that type II is much more common than type I form, the recent reports suggest that the pure form may be as common as that with congenital hepatic fibrosis. Pure form is characterized by segmental, saccular, communicating intrahepatic bile duct ectasia involving a single segment or lobe of the liver or the whole liver. Bile duct dilatation is less prominent in type II. Clinical findings of type I disease are recurrent cramplike abdominal pains, fever, jaundice; clinical findings of type II disease are related to portal hypertension developing secondary to hepatic fibrosis. Medullary sponge kidney (in 80 %), infantile polycystic kidney disease, renal tubular ectasia, and rarely choledochal cyst are the conditions associated with Caroli's disease. Multiple cystic structures that converge toward the porta hepatis as either localized or diffusely scattered cysts communicating with bile ducts is the essential diagnostic feature of Caroli's disease in medical imaging studies (ultrasonography, computed tomography, MRI). Other imaging findings can be listed as segmental saccular or beaded appearance of intrahepatic bile ducts extending toward the periphery of the liver; fibrovascular bundles containing a portal vein radicle and a small branch of hepatic artery completely surrounded by dilated intrahepatic

bile ducts (central dot sign on computed tomography); bridge formation across dilated lumina; frequent ectasia of extrahepatic ducts and common bile duct; sludge or calculi in dilated ducts. Magnetic resonance cholangiopancreatography (MRCP) is a noninvasive technique alternative to invasive imaging techniques (endoscopic retrograde cholangiopancreatography and percutaneous transhepatic cholangiography) to evaluate biliary tree. The MRCP images are obtained from three-dimensional maximum intensity projection reconstructions of heavily T2-weighted fast spin-echo sequences. The maximum intensity projection reconstruction allows rotation of the images in the coronal plane with demonstration of continuity beetwen hepatic cystic dilatations and the biliary tree in Caroli's disease as the most important diagnostic finding. Differential diagnosis of Caroli's disease is not difficult from polycystic liver disease, multiple hepatic abscesses, and other more unusual malformations by the characteristic imaging findings of the disease. Complications of the disease are bile stasis with recurrent cholangitis, biliary calculi, hepatic abscess, septicemia, and increased risk of cholangiocarcinoma in approximately 7 % of the cases. The treatment depends on the clinical features and the location of the biliary abnormality. When the disease is localized to one hepatic lobe, lobectomy relieves symptoms and appears to remove the risk of malignancy. In diffuse Caroli's disease, treatment options include conservative or endoscopic therapy, internal biliary bypass procedures and liver transplantation in carefully selected cases.
Differential Diagnosis List: Caroli's disease

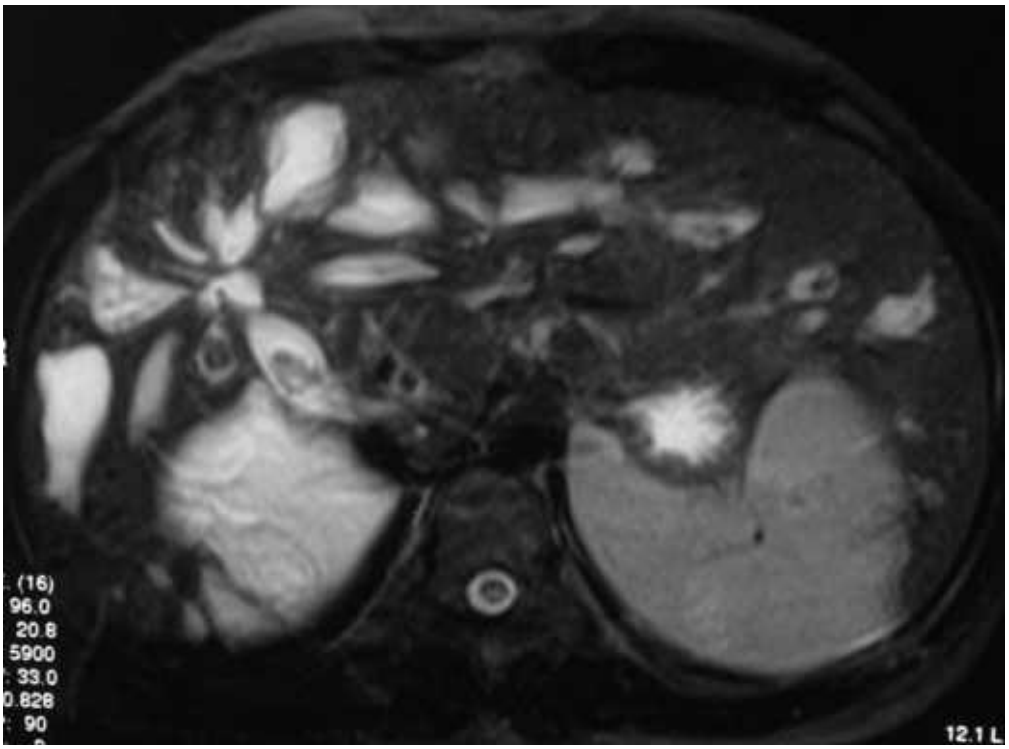
Final Diagnosis: Caroli's disease

References:

Pavone P, Laghi A, Catalano C, Passariello R. Caroli's disease: evaluation with MR cholangiography. AJR Am J Roentgenol 1996 Jan;166(1):216-7. (PMID: 8571889)
Jung G, Benz-Bohm G, Kugel H, Keller KM, Querfeld U. MR cholangiography in children with autosomal recessive polycystic kidney disease. Pediatr Radiol 1999 Jun;29(6):463-6. (PMID: 10369907)
Zangger P, Grossholz M, Mentha G, Lemoine R, Graf JD, Terrier F. MRI findings in Caroli's disease and intrahepatic pigmented calculi. Abdom Imaging 1995 Jul-Aug;20(4):361-4. (PMID: 7549744)
Taylor AC, Palmer KR. Caroli's disease. Eur J Gastroenterol Hepatol 1998 Feb;10(2):105-8. (PMID: 9581983)
Pavone P, Laghi A, Catalano C, Materia A, Basso N, Passariello R. Caroli's disease: evaluation with MR cholangiopancreatography (MRCP). Abdom Imaging 1996 Mar-Apr;21(2):117-9. (PMID: 8661756)

Figure 1

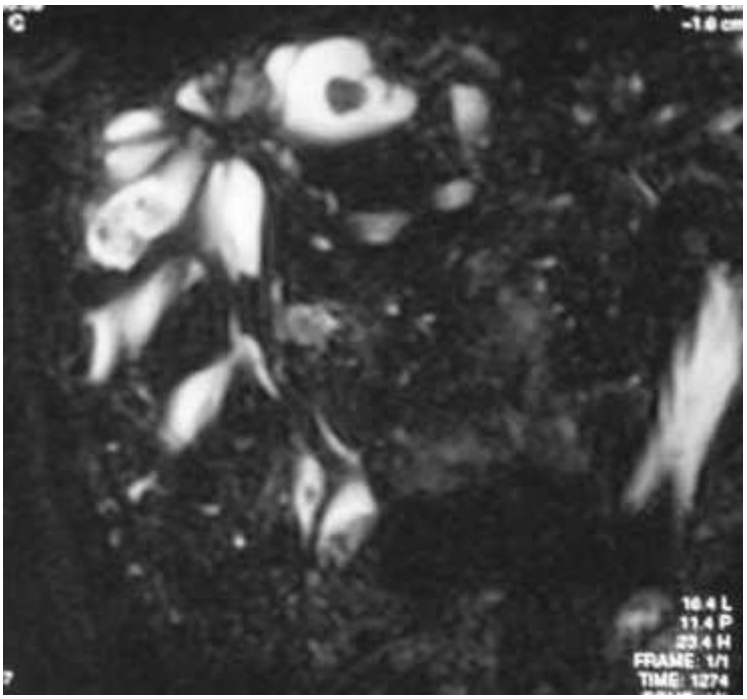
a



Description: Axial FSE T2-weighted MR image shows diffuse cystic dilatations of the intrahepatic bile ducts, some of which contain hypointense calculi, and some fluid-fluid levels due to bile sludge.**Origin:**

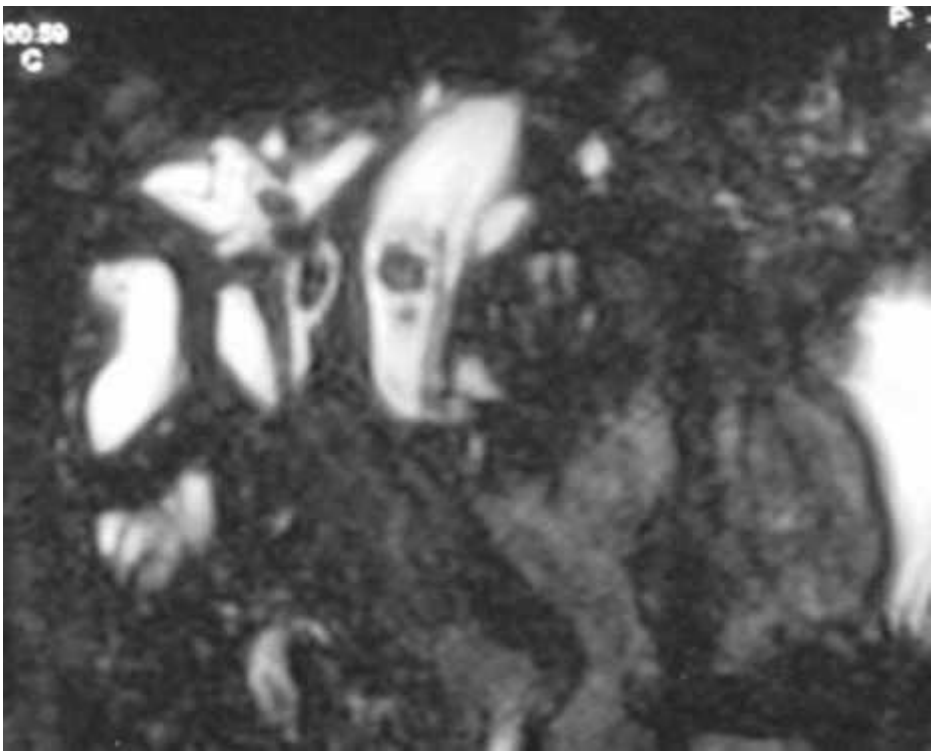
Figure 2

a



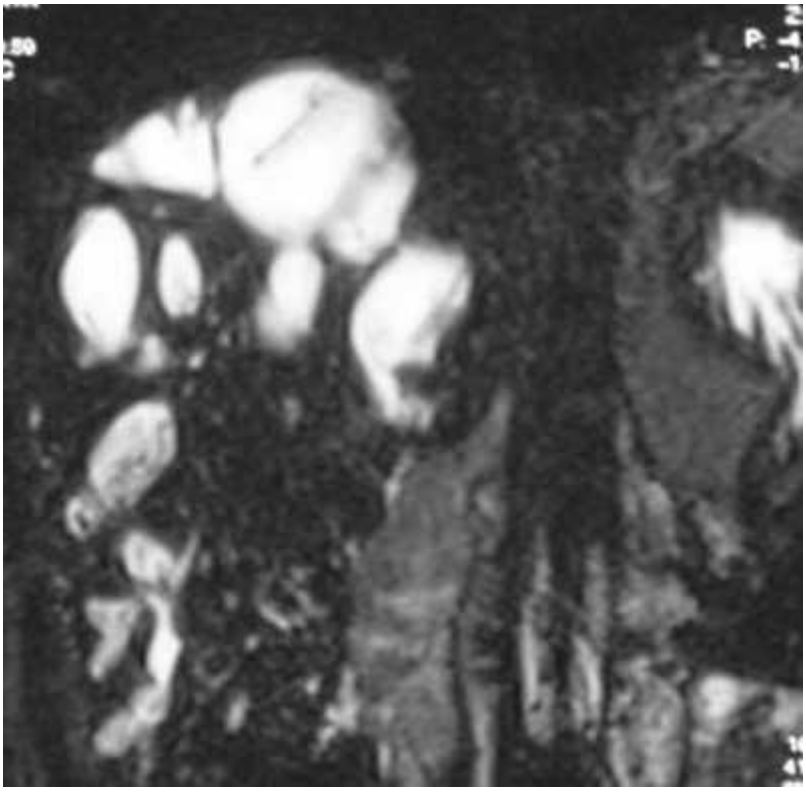
Description: Coronal heavily FSE T2-weighted MR image shows diffuse cystic dilatations of the intrahepatic bile ducts, some containing hypointense calculi, and some containing fluid-fluid levels due to bile sludge. **Origin:**

b



Description: Coronal heavily FSE T2-weighted MR image shows diffuse cystic dilatations of the intrahepatic bile ducts, some containing hypointense calculi, and some containing fluid-fluid levels due to bile sludge. **Origin:**

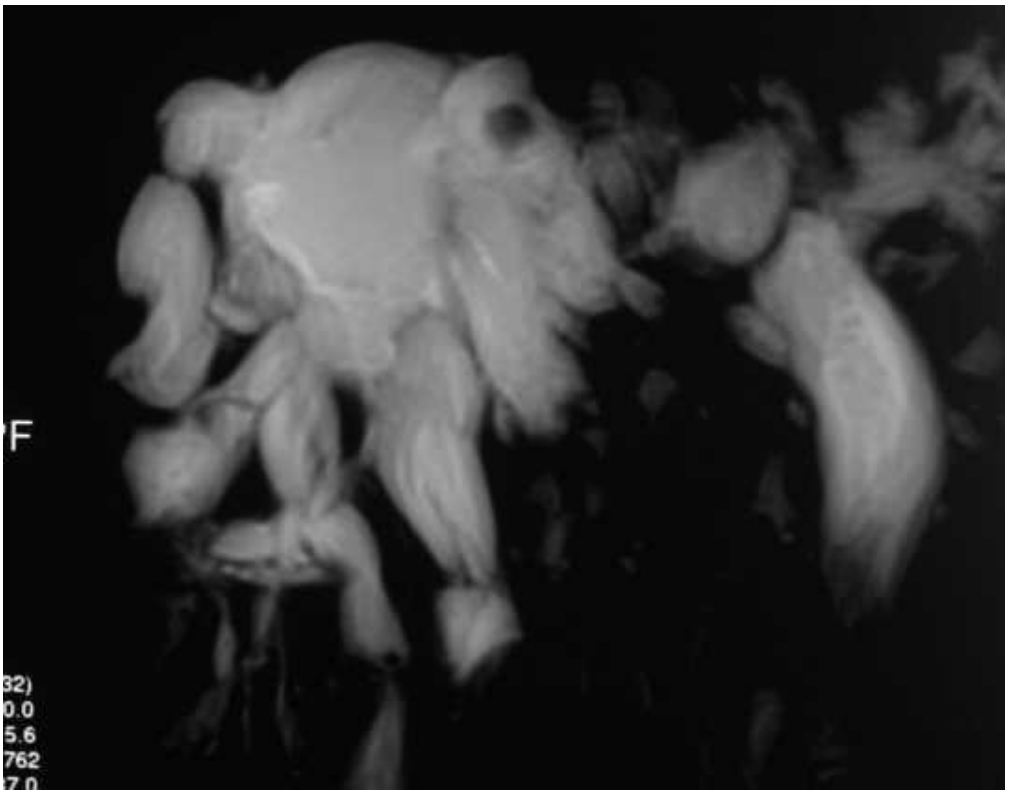
c



Description: Coronal heavily FSE T2-weighted MR image shows diffuse cystic dilatations of the intrahepatic bile ducts and gall bladder, some containing hypointense calculi, and some containing fluid-fluid levels due to bile sludge. **Origin:**

Figure 3

a



Description: Coronal MRCP image demonstrates continuity between cystic dilated biliary tracts, and calculi as hypointense filling defects. **Origin:**