Imaging of a Patient with Multiple Intrahepatic Cysts

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Introduction

A 25-year-old male presented with repeated attacks of cholangitis of six months duration. On examination, the patient was febrile and icteric, abdominal examination revealed a 6 cm firm hepatomegaly. The routine blood investigations were normal except for a raised serum bilirubin (4 mg/dL) and alkaline phosphatase (940 IU/mL). The patient underwent an ultrasound followed by an MRCP and contrast-enhanced CT scan of the abdomen, (Fig. 1). Serum alpha fetoprotein was 3 ng/mL and CA 19-9 was 3656 U/mL.

Figure 1: MRCP and contrast-enhanced CT scan of the abdomen.

Question

1. What is the diagnosis?

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Figure 2: MRI showing intraductal calculi.

Figure 3: MRCP showing multiple cystic dilations of bilateral intrahepatic ducts with signal voids seen in left duct suggestive of calculi, (white arrows). This is a typical picture of Caroli’s disease with bilateral intrahepatic cystic dilatations with normal extrahepatic ducts.

Answer

1. Caroli’s disease with hepatolithiasis and cholangiocarcinoma.

Discussion

Caroli’s disease (CD) is a rare autosomal recessive, hereditary disorder of the intrahepatic bile ducts due to a ductal plate malformation. In CD, the large and proximal intrahepatic bile ducts are affected and they correspond to the type V bile duct cyst as incorporated by Todani et al. The patient may present with cholangitis or manifestations of portal hypertension (if associated with congenital hepatic fibrosis). Hepatolithiasis and choledocholithiasis is common. It is associated with both repeated episodes of cholangitis and cholangiocarcinoma. Dysplasia of the biliary epithelium generated by bile stasis and chronic inflammation can develop a malignant transformation to cholangiocellular carcinoma in 5% to 14% of patients with CD. In both of these conditions, CA 19-9 is elevated; however, the levels in cholangiocarcinoma are frequently elevated to >1000 IU/mL and can help distinguish between benign and malignant diseases. Higher levels of these tumor markers have been suggested to have a prognostic significance with a high value correlated to have a higher chance of the lesion being advanced. In this patient, CA 19-9 was 3656 IU/mL and patient was found to have metastatic disease on exploration.

Figure 4: Contrast-enhanced CT scan in portal venous phase showing multiple cystic lesions with calculi in both lobes of the liver. White arrow points towards a cyst in the segment IV packed with stones. A stent is also seen in-situ.

Figure 5: Contrast-enhanced CT scan. White arrow shows bright enhancement of part of wall of the cyst in segment IV on arterial phase with intraluminal mass suggestive of malignant transformation. Cholangiocarcinomas may occasionally show enhancement in the arterial phase.

References


