Unexpectedly Diagnosed Caroli's Disease on HIDA Scintigraphy in a Patient with Calculous Cholecystitis

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Abstract Caroli's disease, which is a rare condition with congenital dilatation of the intrahepatic bile ducts, is usually diagnosed postoperatively. The clinical suspicion in a patient with gallstones and choledocholithiasis presenting with dilated intrahepatic biliary radicles and jaundice is usually an obstructive etiology. However, scintigraphic evaluation of this entity, as in this case, gives additional information on liver function, biliary drainage and predisposing conditions like Caroli's disease, which could be missed otherwise.

Keywords Caroli's disease · Calculous cholecystitis · Scintigraphy · HIDA scan

Introduction

Management of patients with abdominal discomfort, jaundice, and stones in the gall bladder and bile ducts on ultrasonography can be challenging and need to incorporate clinical, imaging and biochemical derangements to arrive at the correct diagnosis and prognosis.

Caroli's disease, though a relatively rare condition, is being increasingly reported due to advances in imaging procedures. However, the majority of the cases are still not diagnosed at presentation and are usually uncovered postoperatively and with cholangiography. Scintigraphic evaluation of this entity is not widely utilized; especially in preoperative diagnosis. We would like to present a case of incidentally diagnosed Caroli's disease in a patient with clinical suspicion of cholelithiasis and choledocholithiasis as the cause of hepatic derangement and jaundice, thus altering the clinical management completely.

Case Report

A 48-year-old male patient presented with drowsiness, jaundice, and occasional abdominal discomfort. Serum bilirubin was raised and ultrasound of the abdomen demonstrated dilated fusiform intrahepatic biliary radicles (IHBR) and dilated common bile duct (CBD) with multiple calculi. The gall bladder appeared grossly distended with multiple calculi at the neck region and demonstrated thickened wall with inflammatory changes suggestive of cholecystitis. He was referred for a technetium-99m (Tc99m)-labeled hepatic imino-diacetic acid scan (HIDA scan) to assess the level of biliary obstruction and gall bladder function. On scintigraphy, the liver demonstrated well-preserved function, as seen by good uptake of tracer and only mild delay in blood pool clearance. The left lobe of the liver appeared enlarged. The scan suggested multiple focal photopenic defects initially. The filling of these defects on the delayed images, as the hepatic parenchyma cleared and later stasis with pooling of tracer in these regions, strongly suggested Caroli's disease (Fig. 1). There was delayed excretion of tracer into the intestine, characteristic of Caroli's disease due to underlying bile stasis

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Fig. 1 HIDA scan with dynamic 2-min/frame anterior abdominal images, demonstrating non-uniform tracer localization in the liver filling up progressively. The left lobe appears enlarged in size. The blood pool clearance is prompt. However, there is no tracer visualized in the extrahepatic biliary radicles, gall bladder or small intestine up to 30 min [3–5]

and the scintigraphic hallmark of ‘lumpy-bumpy’ appearance of the tracer filled dilated IHBR in the delayed images (Figs. 2, 3). In addition, non-visualization of the gall bladder, even up to 3 h post injection, suggested occluded cystic duct,

which was consistent with calculous cholecystitis on ultrasound (Fig. 4). Magnetic resonance cholangiopancreatography (MRCP) was done later, which confirmed Caroli’s disease.

Fig. 2 Static 1-min image of the liver at 1 h post injection, demonstrating retention of tracer in certain regions, while the rest of the liver parenchyma clears normally. This is consistent with the characteristic ‘lumpy-bumpy’ appearance of the liver in Caroli’s disease [6, 7]

Fig. 3 Static 1-min image of the liver at 3 h post injection, demonstrating retention of tracer in certain regions, while the rest of the liver parenchyma clears normally. This is consistent with the characteristic ‘lumpy-bumpy’ appearance of the liver in Caroli’s disease [6, 7]

Fig. 4 Same static image at 1-h post injection scaled up in intensity to demonstrate minimal, slow intestinal tracer accumulation

Discussion

Caroli's disease is characterized by an inherited dilatation of the intrahepatic bile ducts. Caroli's disease can be of two types, the simple or isolated case where only the bile ducts are affected and are widened by ectasia. The second, more complex etio-pathological variant with portal hypertension and congenital hepatic fibrosis is commonly known as Caroli syndrome [1, 2]. The differences between the causes of the two cases have not yet been discovered, though more cases of Caroli syndrome have been reported than of Caroli's disease. Liver failure and polycystic kidney disease have also been reported in association with Caroli's disease [2]. Caroli's disease is also distinct from other obstructive disorders that cause ductal dilatation or from choledochal cyst derivatives [1, 2].

Imino-diacetic acid (IDA) analogues, which are used in the HIDA scan, follow the same metabolic pathway as bilirubin. On intravenous injection, IDA analogues are rapidly extracted by hepatocytes and then excreted in the

biliary tree and finally drain into the intestines. Based on the above qualities, HIDA scans employ Tc99m-labeled IDA analogues for the assessment of hepatocyte function, demonstration of biliary tract patency and studying gall bladder function. The commonly used IDA analogue is DISIDA (di-isopropyl imino-diacetic acid). Sequential images of the hepatobiliary system are acquired after intravenous injection of radiotracer Tc99m-labeled IDA. The scintigraphic hallmarks of diagnosis for Caroli's disease are delayed hepatic clearance, with tracer pooling in the beaded, dilated IHBR and delayed intestinal visualization.

Scintigraphy in this case gave an unexpected diagnosis of Caroli's disease in addition to acute calculous cholecystitis. HIDA scan also gives a clearer, dynamic picture of biliary uptake and excretion kinetics in patients suspected to have biliary or gall bladder related pathology; thus enabling the clinician to differentiate between obstructive and non-obstructive causes of intrahepatic biliary dilatation with additional findings like degree of hepatocyte damage and gall bladder function helping management decisions.

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