An unusual case of monolobar Caroli's disease

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AJ GOMES, RJ BAILEY. An unusual case of monolobar Caroli's disease. *Can J Gastroenterol* 1994;8(3):185-188. A 27-year-old male with recurrent upper abdominal pain was found to have a suspicious mass in the right hepatic lobe. Right hepatectomy was performed. Pathological examination and further radiological evaluation proved this to be a focal form of Caroli's disease.

Key Words: Caroli's disease, Cholangiocarcinoma, Cholangitis, Intrahepatic duct dilation, Monolobar

Cas inhabituel de maladie de Caroli monolobulaire

RÉSUMÉ : Un homme de 27 ans souffrant de douleurs abdominales hautes récurrentes présente une masse douteuse au lobe hépatique droit. L'hépatectomie droite a été effectuée. L'examen pathologique et d'autres épreuves radiologiques confirment qu'il s'agit d'une forme localisée de la maladie de Caroli.

CAROLI'S DISEASE IS A RARE biliary anomaly that usually involves the liver diffusely. In some cases, however, it may be focal and present as a suspicious mass within the liver. An unusual case of pure Caroli's disease with monolobar localization to the right hepatic lobe – diagnosis was made following right hepatic lobectomy – is described, followed by a review of the relevant literature.

CASE PRESENTATION

A 27-year-old Caucasian male presented with a nine-year history of intermittent epigastric pain not associated with fever or jaundice. Physical examination was normal. Liver function tests were within normal limits. An ultrasound of the liver suggested a mass in the right lobe of the liver. Computed tomography (CT) scan of the abdomen revealed a 13x11x11 cm mass in the

posterior segment of the right lobe (Figure 1). Multiple internal areas of attenuation were noted, suggesting either dilated biliary radicals or focal areas of necrosis. Normal vascular structures were displaced anteriorly and medially. The kidneys and spleen were normal. It was felt that the mass was a primary hepatic neoplasm, either hepatoma or fibrolamellar carcinoma. A hepatic angiogram failed to yield additional information.

A laparotomy and right hepatic lobectomy were performed. Findings included a palpable mass in the superior portion of the right lobe, but the liver surface appeared grossly unremarkable.

Pathological examination revealed an area of thickening within the right lobe. Incision into this area revealed multiple dilated channels grouped in an area approximately 8 cm in diameter (Figure 2). Microscopically, the portal tracts were enlarged, with dilated cystic bile ducts.

These ducts were lined by a single layer of columnar mucinous cells with some papillary infoldings (Figure 3). Some ducts contained large biliary concretions. The remainder of the parenchyma showed mild steatosis only. These findings were compatible with a diagnosis of Caroli's disease, focal type,

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Figure 1) Augmented computed tomography scan of the upper abdomen showing a large inhomogeneous mass within the posterior segment of the right lobe of the liver displacing normal vascular structures

Figure 2) Cut section (gross) of the resected right hepatic lobe showing cystically dilated bile ducts

involving the right lobe of the liver. The patient made an uneventful recovery, and was discharged home on the 11th postoperative day. An endoscopic retrograde cholangiogram (ERCP) six months later revealed a normal intra- and extrahepatic biliary system, with no evidence of remaining segmental duct dilation (Figure 4). The patient has been asymptomatic since surgery.

DISCUSSION

Caroli's disease (1,2) is a rare condition characterized by dilation of the segmental intrahepatic bile ducts. It is congenital, and appears to be inherited in an autosomal recessive fashion (2-4).

The etiology of this condition is unknown, but has variously been attributed to embryological hepatic vascular occlusion (5), failure of resorption of

the primitive biliary ducts (6) and inequality in the rate of growth between embryological liver connective tissue and biliary epithelium (7).

Caroli's disease appears to be just part of a spectrum of developmental abnormalities of the biliary tree (8,9). This spectrum includes choledochal cysts, which accompany intrahepatic duct dilation in 22 to 53% of cases (9,10), and congenital hepatic fibrosis in 35% of cases (9). Nearly one-third of patients with Caroli's disease have both (9).

Two forms of Caroli's disease have been described (2). The first – the simple or 'pure' form – is not associated with congenital hepatic fibrosis and is uncommon (13%) (9). The second form is 'periportal fibrosis associated type', which is linked to congenital hepatic fibrosis, cirrhosis and portal hypertension (2).

The cystic biliary radicals most commonly involve the entire liver. Boyle et al (11) found monolobar involvement in 34 of 180 known cases of Caroli's disease and noted that monolobar involvement may be more common in the pure form. The right lobe is rarely affected in monolobar disease (8%).

Renal anomalies, most commonly medullary sponge kidney and dilation of collecting ducts, are common, and in the pure form of the disease are present in 60 to 80% of cases (11).

Our patient appears to have had the less common pure form of Caroli's disease, with rare monolobar localization to the right hepatic lobe. In addition, there was no evidence of other biliary tract or renal anomalies.

In the pure form of Caroli's disease, biliary stasis leads to intrahepatic calculi and infection. In 80% of cases, symptoms begin before age 30 years (10). Most commonly, recurrent fever, abdominal pain and, occasionally, mild jaundice are presenting features. Physical examination may reveal tenderness of hepatomegaly. Laboratory investigations are generally within normal limits, apart from a leukocytosis during attacks (2). Uncommonly, patients with associated hepatic fibrosis may present with complications of portal hypertension. Migration of intrahepa-

tic stones may result in obstructive jaundice or pancreatitis (11,12). Recurrent cholangitis characterizes the course of disease, and patients eventually die as a result of septic complications (2).

This malformation has only relatively recently been recognized, and missed diagnosis is common. Even when the diagnosis is made, the delay from onset of symptoms is between four and 12 years (11,13). Often the diagnosis is made after multiple abdominal explorations, often with cholecystectomy and common bile duct exploration (10). With the advent of ultrasound and CT, however, the diagnosis is now more commonly made noninvasively (14). The monolobar form of Caroli's disease is diagnosed preoperatively in 27% of patients, most commonly by ERCP or CT scanning (11). In our case, the diagnosis was not entertained preoperatively.

No effective, definitive medical therapy exists for Caroli's disease (11,14). Chenodeoxycholic acid has been recommended for patients with documented stones in Caroli's disease, and has been used successfully for dissolution following incomplete liver resection (15,16).

Traditionally, surgery has consisted of bilioenteric anastomoses to improve drainage and aid clearance of common duct calculi (10). This, however, often results in incomplete decompression because the ectatic intrahepatic ducts often have stenotic segments (10,13). External drainage has proven to be inadequate, and other surgical procedures are often necessary (10). If Caroli's disease is localized to a segment or lobe, hepatic resection is the procedure of choice; it invariably results in complete resolution of symptoms (11,14,17).

Caroli's disease is complicated by cholangiocarcinoma in the affected ducts in up to 7% of cases (12,13). Patients with cholangiocarcinoma tend to be older (mean 48 years of age) than those presenting with cholangitis (12). Epithelial dysplasia has been noted in resected cysts, supporting the premalignant nature of the lesion (18). In addition, malignant degeneration has been

Figure 3) Low power photomicrograph of a dilated bile duct with papillary infoldings and biliary concretions within the lumen

Figure 4) Postoperative endoscopic retrograde cholangiogram confirming the absence of dilated biliary ducts in the remaining liver

shown to occur in cystic areas despite adequate drainage (12,19). This further reinforces the importance of early complete resection of symptomatic lesions when feasible.

In the presented patient, right lobectomy was performed for a suspicious liver mass. The patient's symp-

toms remitted, and the future risk of cholangiocarcinoma should be reduced markedly.

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