Monolobar caroli’s disease: report of three cases

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ABSTRACT

Caroli’s disease is a rare congenital hepatobiliary disease characterized by multifocal segmental dilatation of intrahepatic bile ducts affecting all or parts of the liver. It predisposes to biliary stasis and consequent lithiasis, cholangitis, abscesses, and septicemia. Caroli’s disease is most often diffuse and rarely localized to one lobe of the liver, mainly on the left.

We report three cases of monolobar Caroli’s disease revealed by cholestatic jaundice in 2 cases and recurrent cholangitis in 1 case. All patients were men with an average age of 41 years old. The average duration between first symptoms and diagnosis was 3 months. In all cases the diagnosis was suggested by radiology and confirmed by histology. Caroli’s disease was located on the left liver lobe with the presence of intrahepatic lithiasis in 2 cases. The treatment was consisted to a left hepatectomy in 2 cases and a biliary-digestive anastomosis in 1 case. The evolution was favorable in two cases while the third case was lost.

Key words: Caroli’s disease, Monolobar, magnetic resonance cholangiopancreatography

INTRODUCTION

Caroli’s disease is a rare congenital hepatobiliary disease characterized by multifocal segmental dilatation of intrahepatic bile ducts. It is recognized to occur in two distinct forms: the simple type, and the periportal fibrosis type. Caroli’s disease may diffusely affected the liver or be localized to one lobe or segment. Less than 20% of all reported cases of Caroli’s disease are monolobar type.

Clinical symptoms usually appear during childhood or early adult life and include abdominal pain and jaundice; Caroli’s disease predispose to biliary stasis and intrahepatic stones formation leading to cholangitis, liver abscesses, septicemia, and ultimately to secondary biliary cirrhosis. Moreover, the tendency to cholangiocarcinoma development on these abnormal bile ducts is well documented [1,2]. Endoscopic or percutaneous cholangiography is the traditional method of diagnosis, but Magnetic resonance cholangiopancreatography (MRCP) is emerging as the diagnostic modality of choice. This report describes three cases of monolobar Caroli’s disease confined to the left lobe.

CASES REPORT

Our three patients were men with an average age of 41 years old. The average duration between first symptoms and diagnosis was 3 months. In all cases, the diagnosis was suggested by radiology: MRCP alone in one case and coupled with endoscopic retrograde cholangiopancreatography (ERCP) in two cases. Caroli’s disease was located on the left in all patients (Figure 1) with the presence of intrahepatic lithiasis in 2 cases (Figure 3) and dot sign in one case (Figure2). Surgical resection has been used successfully in 2 patients: left hepatectomy and a biliary digestive anastomosis in 1 case. The evolution was favorable in two cases while the third case was lost.

The clinical characteristics, methods of diagnosis and treatment are summarized in table 1.

Table 1: Summary of case reports

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sax/age</td>
<td>M/35</td>
<td>M/56</td>
<td>M/54</td>
</tr>
<tr>
<td>Clinical</td>
<td>Cholestatic jaundice</td>
<td>Cholestatic jaundice</td>
<td>Recurrent Cholangitis</td>
</tr>
<tr>
<td>Biology</td>
<td>Cholestasis 20 N + Cytolysis 7 N</td>
<td>Cholestasis 5 N + minimal Cytolysis</td>
<td>Cholestasis 7 N</td>
</tr>
<tr>
<td>Ultrasound</td>
<td>Dilatation of left biliary tract</td>
<td>Normal</td>
<td>Dilatation of left biliary tract and common</td>
</tr>
</tbody>
</table>
**DISCUSSION**

Caroli’s disease was first described by Caroli and Couinaud in 1958, it is a rare congenital entity of the fibropolycystic family of liver diseases defined by segmental non-obstructive dilatation of the large intrahepatic bile ducts. Caroli’s disease correspond to type V of congenital bile duct cysts according to the Todani et al classification. The most common form of Caroli’s disease is the diffuse form affecting both lobes of the liver. Monolobar involvement is much more uncommon (20%) and when present involves the left lobe more often than the right. When the diffuse form coexists with a congenital hepatic fibrosis or cirrhosis with portal hypertension, it is called Caroli syndrome. The disease results from an arrest of the normal embryologic remodeling of ducts with resultant destructive inflammation and segmental dilatation. The disease affects about 1 in 1,000,000 people, with more reported cases of Caroli syndrome than of Caroli’s disease. Males and females are equally affected and more than 80% of patients present before 30 years old. We have identified 40 cases only of monolobar caroli’s disease in the literature from 1965 to 2010 outside of our three cases. Clinical characteristics are dominated by recurrent cholangitis in 64% patients and the classical evolution of the disease is intrahepatic stones formation responsible of cholangitis and liver abscesses. Malignancy is a complication of long-term caroli’s disease with an incidence of 2.5 to 16% of cases. US and CT studies may visualize liver cysts, possible intrahepatic lithiasis and provide information on the common bile duct, but differentiation from other liver cysts such as polycystic liver disease is often difficult; dilated bile ducts are anechoic in ultrasonography and hypodense on CT scan, the fibrovascular bundles containing portal vein radicles and a branch of hepatic artery bridging the saccule, appears as a dot sign enhancing with contrast, it’s suggested as a pathognomonic finding in Caroli’s disease. We have found a dot sign in one patient. MRCP is emerging as the modality of choice for diagnosis of Caroli’s disease. This noninvasive technique demonstrated a communication between the cystic malformations of the liver and the biliary system, distinguishing this disease entity from isolated hepatic cysts. MRCP provides also the severity, location and extent of liver involvement. The findings of MRCP and ERCP are similar. At present, MRCP is considered as the first method of choice for the diagnosis of Caroli’s disease. The aim of therapy is to decrease morbidity and mortality associated with recurrent cholangitis, hepatic abscesses and cholangiocarcinoma; lobar or segmental resection may lead to a complete cure of monolobar Caroli’s disease with an acceptable morbidity. In our cases, left hepatectomy relieved symptoms in two of our patients, the remaining patient had biliary digestive diversion with an effective improvement. Histopathological examination of the resected specimen is still the gold standard for diagnosing monolobar Caroli’s disease.

**CONCLUSION**

Caroli’s disease is a rare congenital disease, recurrent cholangitis is the most common suggestive symptom of Caroli’s disease. MRCP is noninvasive examination alternative to ERCP for diagnosis of parenchymal cystic structures communicating with biliary tree and complications such as hepatolithiasis and malignancy demonstration. The hepatic resection should be considered as the treatment of choice for patients with monolobar Caroli’s disease, because this resolves symptoms and averts the development of cholangiocarcinoma.

<table>
<thead>
<tr>
<th>Diagnostic</th>
<th>MRCP</th>
<th>MRCP/ERCP</th>
<th>MRCP/ERCP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Period of diagnosis</td>
<td>6 months</td>
<td>1 month</td>
<td>15 days</td>
</tr>
<tr>
<td>Localization of CD</td>
<td>Left</td>
<td>Left</td>
<td>Left</td>
</tr>
<tr>
<td>intra-hepatic lithiasis</td>
<td>NO</td>
<td>yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Treatment</td>
<td>Left Hepatectomy</td>
<td>Left Hepatectomy</td>
<td>biliary-digestive anastomosis</td>
</tr>
<tr>
<td>Evolution</td>
<td>Favorable</td>
<td>Unavailable information</td>
<td>Favorable</td>
</tr>
</tbody>
</table>

**Figure 1:** MRCP reveals left dilated biliary system, normal right biliary system and no evidence of mass or tumor.

**Figure 2:** MRCP objectiving the « Dot sign » that represents portal radicles (arrow)
REFERENCES