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Monolobar Caroli's Disease: Report of Three Cases

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Abstract **Case Report**

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. Which predisposes to biliary stasis and consequent lithiasis, cholangitis, abscesses, and septicemia. Most often diffuse; it is much 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. The Caroli's disease was located on the left liver lobe with the presence of intrahepatic lithiasis in 2 cases. Regarding treatment, it consisted of a left hepatectomy in 2 cases and a biliary-digestive diversion in 1 case. The outcome was favorable in two cases while the third case was lost.

Keywords: Monolobar Caroli's left liver biliary-digestive.

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. The disease may diffusely affect 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 expression may include biliary stasis with obstructive jaundice and consequent lithiasis, recurrent cholangitis, abscesses, and septicemia. Cholangiocarcinoma can also develop in these lesions. Several imaging techniques are used to visualize the biliary tract, but endoscopic retrograde cholangiopancreatography is the most accepted modality for accurate diagnosis of Caroli's disease. This report describes three cases of monolobar Caroli's desease confined to the left lobe.

CASE REPORT

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

Clinical characteristics as well as methods of diagnostic and treatment are summarized in table 1.

Table-1: Summary of case reports

	Case 1	Case 2	Case 3
Sexe/Age	M/35	M/56	M/34
clinical caracteristics	Choléstatic jaundice	Cholestatic jaundice	Reccurent Cholangitis
Biology	Cholestasis 20 N Cytolysis 7 N	Cholestasis 5 N + minimal Cytolysis	Cholestasis 7 N
Ultrasound	Dilatation of left biliary tract	Normal	Dilatation of left biliary tract and common bile duct
Diagnostic	MRCP	MRCP/ERCP	MRCP/ERCP
Period of diagnosis	6 moonths	1 month	15 days
Localization of CD	Left	Left	Left
intra-hépatic lithiasis	NO	yes	Yes
Treatment	Left Hépatectomy	Left Hépatectomy	biliary-digestive anastomosis
Evolution	Favorable	Unavailable information	Favorable

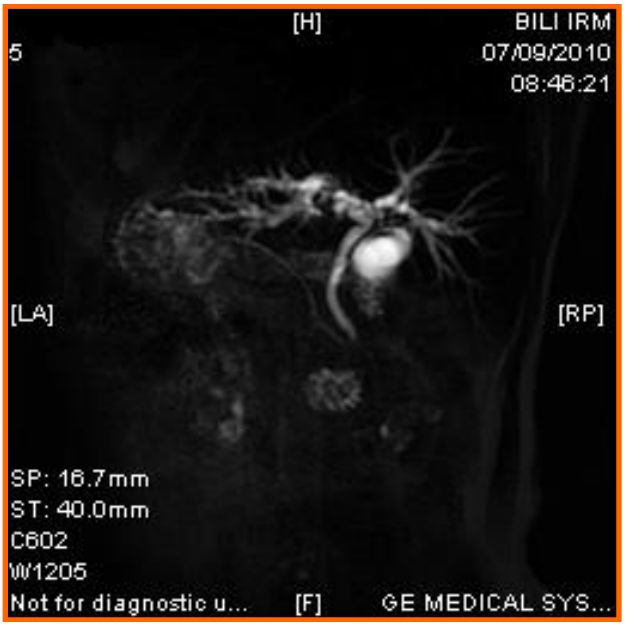

Fig-1: MRCP reveals left dilated biliary system, normal right biliary system and no evidence of mass or tumor

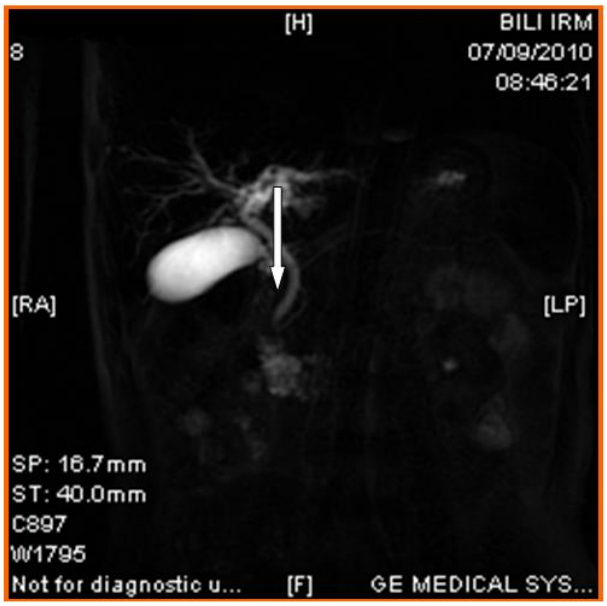

Fig-2: MRCP objectiving the « Dot sign » that represents portal radicles (arrow)

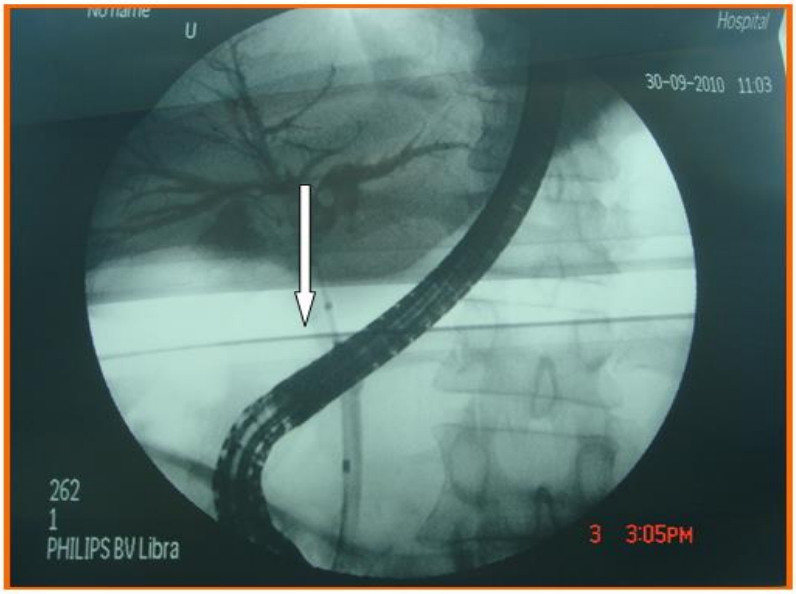

Fig-3: ERCP: dilatation of left hepatic duct + intra-hepatic stones (arrow).

DISCUSSION

Caroli's disease has been described in 1958 by Caroli and Couinaud [1], it is a rare congenital condition, that is defined by segmental nonobstructive dilatation of the large intrahepatic bile ducts in both its forms: diffuse and localized in 20% of cases (left segment 92% of cases); When the diffuse form coexists with a congenital hepatic fibrosis or cirrhosis with hypertension portal, it is called Caroli syndrome.

It results from malformations of the embryonic ductal plate at different levels of the biliary tree [2]. The disease affects about 1 in 1,000,000 people, with more reported cases of Caroli syndrome than of Caroli's disease [3]. Males and females are equally affected and more than 80% of patients present before 30 years of age [4].

We have identified 40 cases only of monolobar caroli's disease over literature from 1965 to 2010 outside of our three cases.

Clinical characteristics are dominated by recurrent cholangitis in 64% patients [2] and the classical evolution of the disease is intrahepatic stones responsible of cholangitis, liver abscesses or even longer-term cholangiocarcinoma with an incidence of 2.5 to 16% of cases [5].

USG and CT studies may visualize liver cysts and 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 [6], the dilated sacculi or cystic spaces 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 [7], we have found a dot sign in one patient.

Magnetic resonance cholangiopancreatography (MRCP) is a noninvasive technique demonstrating that cystic lesions are in continuity with the biliary tree, it provides also the severity, location and extent of liver involvement without any of the risks seen with invasive procedures as endoscopic retrograde cholangiopancreatography (sepsis, bile leak, bleeding); the findings of MRCP and ERCP are similar. This new procedure becomes the first-choice imaging technique for diagnosing Caroli's disease [8].

The aim of therapy is to decrease the morbidity and mortality associated to the 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 [9]. 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 relatively uncommon, recurrent cholangitis is the most common suggestive symptom of Caroli's disease.

Caroli disease is commonly diagnosed after this "central dot sign" is detected on a CT scan or ultrasound; however MRCP is the best, and final, approach to show the enlarged bile ducts as a result of Caroli disease.

The diagnosis of Caroli's disease should be early to avoid complications especially the malignant (cholangiocarcinoma).The logical treatment of monolobar Caroli's disease is hepatic resection.

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