

SHORT COMMUNICATION

Caroli's Disease—A Rare Case Report

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Abstract

Caroli's disease is a congenital disorder characterized by a segmental and sacular dilatation of intrahepatic biliary ducts. The diagnosis of Caroli's disease depends on demonstrating that the cystic lesions are in continuity with the biliary tree which can be showed by ultrasonography, computerized tomography, endoscopic retrograde cholangiopancreatography, percutaneous transhepatic cholangiography or magnetic resonance cholangiopancreatography. Treatment of Caroli's disease relies on the location of the biliary abnormalities, while the localized forms can be treated with surgery; liver transplantation is the only effective modality for diffuse forms. After a thorough review, here we report an atypical case of Caroli's disease, presented as an hepatic exophytic mass on a 34 year old male. Right hepatectomy with partial extension to left segment IV was done and the patient was asymptomatic after a 3 months follow-up.

Keywords: Caroli's disease, biliary abnormalities, exophytic mass, central dot sign, right hepatectomy.

Introduction

Caroli's disease is a rare clinical entity with only 200 cases described in the literature (Yonem and Bayraktar, 2007). It was first mentioned in 1906 (Vachell and Stevens, 1906), but its morphological and clinical description was given by Caroli in 1958 (Karim, 2004), stating the congenital etiology of the disease. An estimated incidence of 1/1,000,000 inhabitants has been reported without any gender prevalence (Giovanardi, 2003). It mainly inherited in an autosomal recessive manner, although other modes of inheritance (autosomal dominant) were also described (Yoshizawa *et al.*, 1992; Tsuchida *et al.*, 1995). Loss of the distal 3p chromosome and gain of the 8q chromosome is of pathogenic importance (Parada *et al.*, 1999). There is also good evidence that Caroli's disease confers an approximately 7% risk of malignancy (Gupta *et al.*, 2006).

Caroli's disease often coexists with congenital hepatic fibrosis and is then designated as Caroli's syndrome. Both result from malformations of the embryonic ductal plate at different levels of the biliary tree (Sherlock and Dooley, 2002). If the defective remodelling involves the larger intrahepatic ducts where Caroli's disease develops, but when the entire intrahepatic biliary tree is involved, the condition is Caroli's syndrome (Desmet, 2004). Incidence of Caroli's syndrome is more than Caroli's disease (Gupta *et al.*, 2006). In addition, various renal disorders may be seen in conjunction with these hepatic diseases, including autosomal polycystic kidney disease (both dominant and recessive forms), medullary sponge kidney and medullary cystic disease (Parada *et al.*, 1999).

Caroli's disease is also classified by Todani *et al.* (1977) as type V choledochal cyst. Although present from birth, the disease usually remains asymptomatic during the first 20 years and may also remain so throughout life. However, when symptomatic, a significant number of these patients present significant loss in their quality of life and their clinical course frequently worsen due to the repeated episodes of cholangitis with the presence of intrahepatic calculi, intrahepatic abscesses and sepsis (Waechter *et al.*, 2001). Keeping the above facts in mind, here in this study, we report on an atypical case of that disease presented as an hepatic exophytic mass.

Materials and methods

Case report: A 34 year old male patient presented with a history of right upper abdomen pain since 6 months, loss of appetite since 3 months and yellowish discoloration of eye since 1 month. Initially patient had colicky pain in right upper quadrant of abdomen but for last 3 months he had dull aching intermittent pain in the same region. No history of vomiting or haematemesis. Bowel and bladder habits were normal. There was no significant past family or personal history. On general examination, his vital signs showed temperature of 38°C, pulse of 78/min, blood pressure of 130/80 mm of Hg in right arm supine position, respiratory rate of 16/min and mild icterus was present.

Investigations: Routine investigations and imaging studies which includes ultrasonography and computerized tomography scan was carried out. Diagnosis, operative treatment and histopathological examination were also done.

Histopathological examination: Histological procedures aim to provide good quality sections. Tissues was fixed with neutral formalin 10%, embedded in paraffin and then manually sectioned with a microtome to obtain 4-5 µm-thick paraffin sections. Dewaxed sections were then stained with hematoxylin and eosin and then microscopic examination was done.

Results

On systemic examination, the case was noticed to have tender hepatomegaly (4 cm below the right costal margin in mid clavicular line) and no free fluids. On auscultation, normal bowel sounds was heard. The laboratory test showed altered liver function test. In particular, ALT: 92 U/L, AST: 68U/L, alkaline phosphatase: 147U/L, bilirubin total: 1.6 mg/dL, bilirubin direct: 0.4 mg/dL. Kidney function tests were within normal limits. The ultrasound of the abdomen revealed hepatomegaly with dilated IHBD, biliary calculi with complex cystic areas. These findings were also confirmed by CT abdomen which showed marked intrahepatic ductal dilatation involving right lobe of liver with enhancing central fibrovascular bundles (central dot sign) suggestive of developmental biliary ductal malformation likely caroli's disease and multiple intrahepatic biliary calculi with cholelithiasis (Fig. 1). Patient was posted for surgery; intraoperative aspect of the liver is shown in Fig. 2. Right hepatectomy with partial extension to left segment IV is shown in Fig. 3. Intraoperative findings was large cystic space replacing the whole right lobe of liver segment V, VI, VII, VIII and segment IV studded with multiple stone. Common bile duct was normal. Common hepatic duct, right hepatic duct and left hepatic duct was grossly dilated with rotation of hilum. Vascular and bile duct anatomy was distorted. Left lobe of liver hypertrophy and gall bladder calculi was present (Fig. 4). Post-operative period was uneventful. He was discharged in generally good condition with the advice of regular follow-up. Histopathological examination revealed intrahepatic caroli's disease with cholecystitis (Fig. 5). The patient was asymptomatic after a 3 months follow-up.

Discussion

The patient discussed in this report is pure form of Caroli's disease with no evidence of periportal fibrosis and renal cystic disease. Diagnosis of Caroli's disease is based on clinical features and imaging studies. The normal course in Caroli's disease consists of suppurative cholangitis, septicemia with gram-negative organisms and intrahepatic abscesses. The disease must be recognized before these serious complications arise. The hepatobiliary imaging procedures should now allow an early diagnosis and therefore, a better therapeutic approach (Keramidas *et al.*, 1998; Sans *et al.*, 1997). The characteristic findings of the disease are the dilated sacculi or cystic spaces, visible by U/S or CT of the abdomen, only in some patients, particularly in the early stages.

Fig. 1. CT abdomen shows marked intrahepatic ductal dilatation more in the right lobe with central dot sign.

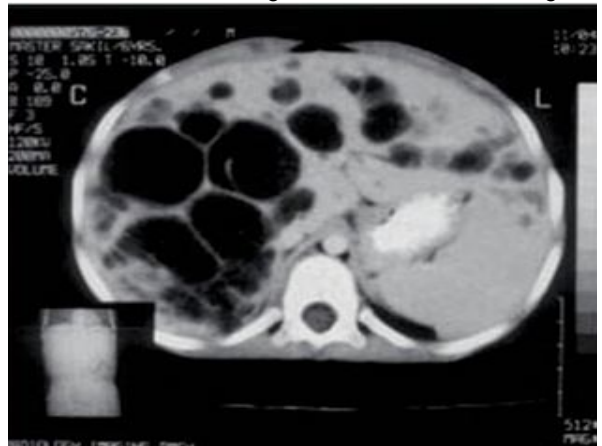


Fig. 2. Intraoperative aspect of the liver.

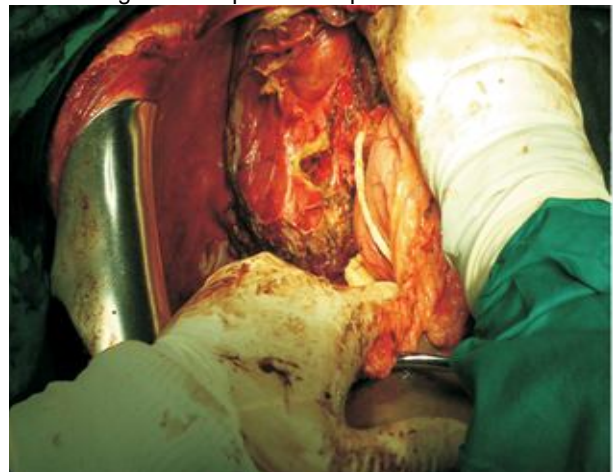


Fig. 3. Specimen photograph (Right hepatectomy with partial extension to left segment IV).



Fig. 4. Gall bladder studded with multiple calculi.

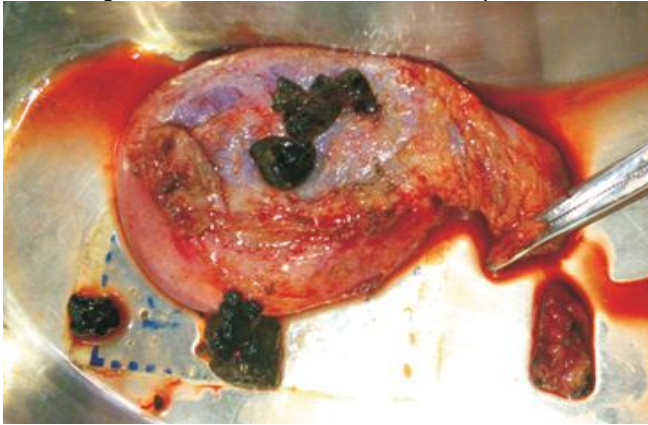
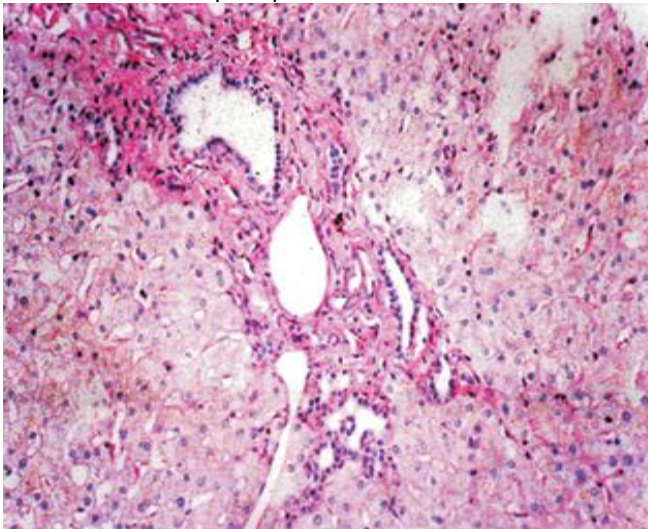


Fig. 5. Histopathological examination shows portal triad widened intrahepatic proliferated and dilated bile ducts.



The findings include saccular or tubular dilatation of intrahepatic bile ducts, intraluminal bulbar protrusions, bridge formation across dilated bile ducts resembling internal septa, within the dilated bile ducts and portal radicles partially or completely surrounded by dilated bile ducts. These portal radicles if seen in axial projection appear as tiny, hyperechogenic structures centrally in the dilated bile duct and have been named; the "central dot" signs (Gupta *et al.*, 2006). Higher density small structures within the dilated ducts represent non-calcified or calcified stones. This central dot sign described on CT scan is suggested as a pathognomonic finding in Caroli's disease (Kaiser *et al.*, 1979; Choi *et al.*, 1990) and it can also be demonstrated on USG (Moorthy *et al.*, 2000). Reviewing the papers on the subject, we were able to find a case of Caroli's disease presented as an exophytic mass of the right lobe. The primary treatment is surgery, for biliary drainage and symptoms relief. Exophytic masses must be totally resected.

Conclusion

Although Caroli's disease is a rare congenital anomaly but its presentation may occur as late as adulthood or middle age. Therefore, Caroli's disease should be considered in differential diagnosis to hepatic exophytic masses.

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