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CAROLI'S DISEASE REVEALED BY ACUTE PANCREATITIS AND ANGIOCHOLITIS: CASE REPORT

ABSTRACT

Background: Caroli's disease (CD) is a rare congenital disorder. The early diagnosis of the disease and differentiation of types I and II are of extreme importance to patient survival. This case report was designed to review, discuss to clarify the clinical characteristics of the disease.

Methods: The demographic and clinical feature, laboratory indicators, imaging findings and pathology results were reviewed.

Results: Caroli's disease can occur at any age specially young age. Magnetic resonance cholangiopancreatography (MRCP) and computed tomography (CT) examinations were most useful in diagnosing CD.

Conclusions: No typical symptoms, signs or laboratory indicators are able to distinguish CD from other conditions. Both MRI and CT were most valuable in diagnosis.

INTRODUCTION

Caroli's disease is defined as cavernous ectasia or saccular dilatation of the intrahepatic bile ducts. it is a rare congenital disorder described for the first time in 1958 (1), autosomal dominant in the form "disease" and autosomal recessive in the form "syndrome". Less than 300 cases have been published. It is characterized by a segmental multifocal dilatation of the intrahepatic bile ducts, involving the entire liver, a lobe, or sometimes a single hepatic segment. it is also characterized by the formation of intraductal lithiasis exposing to a high risk of infection (1-2)

Two types have been described for caroli’s disease: Simple, localized form (often in the left hepatic lobe). And the diffuse complex form known as caroli’s syndrome (congenital hepatic fibrosis, polycystic kidney disease) (3)

MATERIALS AND METHODS

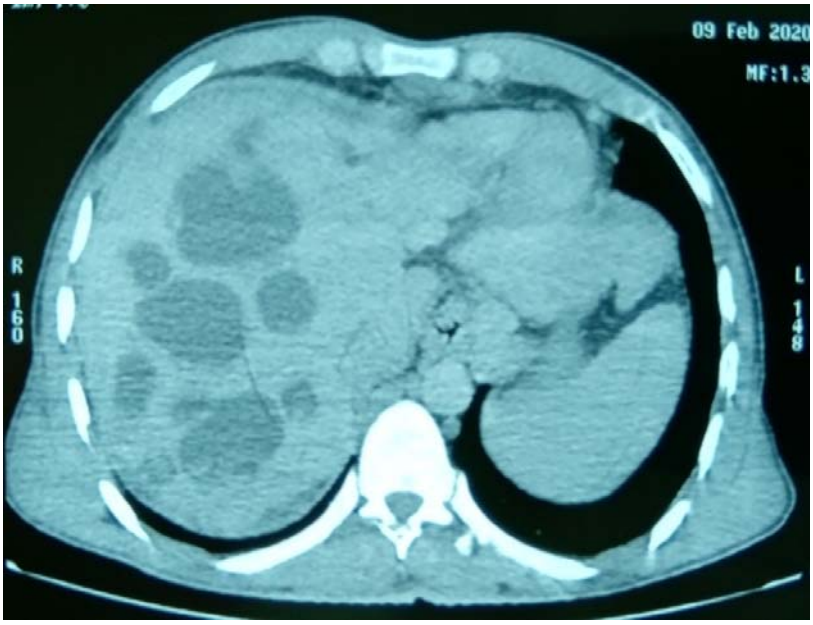
We report the case of a 27 years old man, chronic smoker, presenting for 3 days before his admission a progressive installation of clinical cholestasis syndrome associated with epigastric pain and in the right hypochondrium with vomiting, all developing against a background of fever of 38.4 ° C. in whom the examination found a patient 15/15 conscious, hemodynamically and respiratory stable. T: 38.7 ° C. With generalized mucocutaneous jaundice. The abdominal examination showed an epigastric tenderness, a palpable gallbladder, with whitish stools on the rectal examination.

Laboratory indicators :

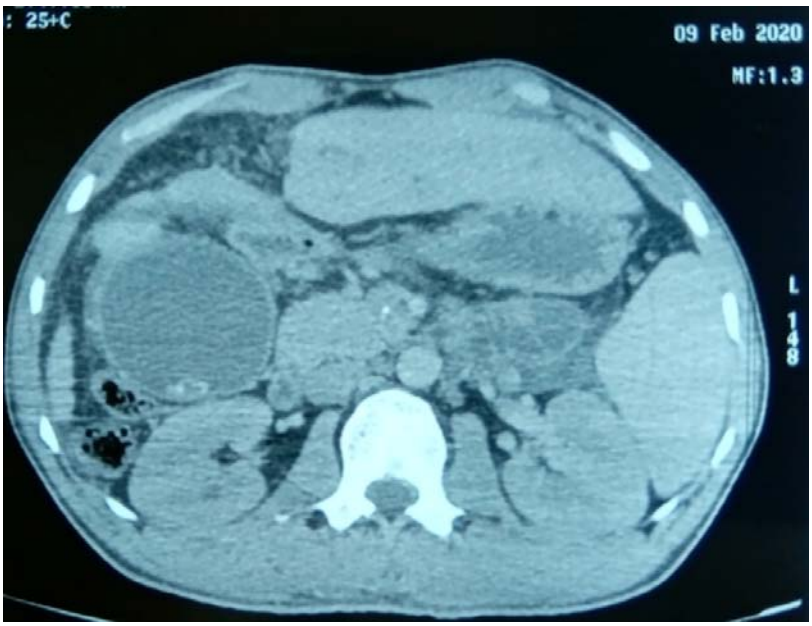
Lipaemia	1136 iu / l (Elevated)
CRP	139.7mg/l (Elevated)
Complete blood counts	
Hemoglobin	12mg/dl
WBC	22860 / mm³
Plt	345000/ mm³
Liver function	
Elevated alanine aminotransferase	246 iu / l
Elevated aspartate aminotransferase	309 iu / l
Elevated total bilirubin	89.6 mg / l
Elevated alkaline phosphatase	683 iu / l
Elevated γ-glutamyl transpeptidase	978 iu / l
Electrolytes	
potassium (K+)	4,1 mmol/l
sodium (Na+)	139 mmol/l
chloride (Cl−)	101mmol/l
Prothrombin Ratio	56%

Table 1 : Laboratory indicators in a 27-year-old man with Caroli's disease

In the imagery; Abdominal scan shows a stage B pancreatitis of balthazar with multisegmentary cystic dilatation of the intrahepatic bile ducts containing multiple lithiasic formations. Main bile duct measuring 13mm in diameter with distended gallbladder containing multiple lithiasic formations. on the Abdominal ultrasound: dilatation of the intrahepatic bile ducts carrying out cystic formations with micro and macro-lithiasis. main bile duct dilation to 15mm in diameter without detectable lithiasis image.
Distended gallbladder measuring 13 * 6 * 7 cm³ with thin wall full of micro and macro-lithiasis.



(a)



(b)

Figure 1 : Computed tomography in a 27-year-old man with Caroli's disease showing secular ectasias and central dotting (a) with distended gallbladder containing multiple lithiasic formations (b)

For the MRI, it showed liver increased in size with regular contours, containing multiple rounded formations well limited in hyposignal T1 and hypersignal T2, some of them are containing lithiasis. Main bile duct measuring 17mm in diameter with lithiasic content. Gallbladder measuring 6.28cm in anteroposterior diameter, with multiple lithiasis. Pancreatitis stage B.

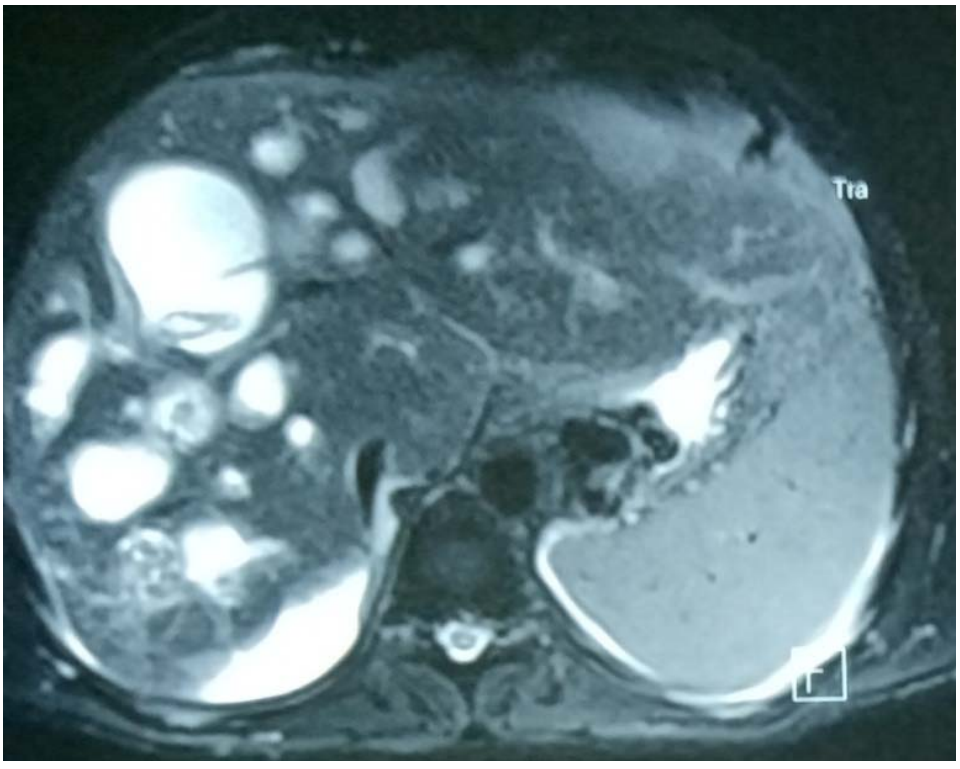


Figure 2 : Magnetic resonance in the same patient shows the connection between the secular ectasias and the normal biliary tract

After cooling the pancreatitis and angiocholitis the patient had benefited from an endoscopic retrograde cholangio-pancreatography with removal of the calculus from the main bile duct and placement of a stent. The post-operative consequences were without peculiarities , characterized by the disappearance of the clinical and biological cholestatic syndrome.

DISCUSSION :

Caroli disease is a congenital disorder of embryonic remodeling of the intrahepatic bile ducts (4). Generally diagnosed in childhood or adolescence, but can be diagnosed later in adulthood (5-6).

Clinically, caroli disease has no specific symptoms and patients can frequently be asymptomatic (7). Intrahepatic ductal ectasia predisposes to the stagnation of bile leading to the formation of lithiasis, and predisposes to infections, in particular angiocholitis, and repeated cholangitis predisposes to calculi of the intrahepatic ducts, which may aggravate bile obstruction and lead to biliary cirrhosis. As well as calculous migration can lead to attacks of acute pancreatitis (8).

Epigastric pain and in the right hypochondrium, fever, and jaundice are the most common symptoms. Repeated episodes of cholangitis can worsen biliary obstruction and lead to biliary cirrhosis (9).

The diagnosis of caroli disease is mostly radiological. On ultrasound, caroli disease appears as anechoic cystic areas, separated by septa, which may contain lithiasis (7). Calculous migration in the main bile duct can result in regular dilatation of the latter. As for the computed tomography, it can show the aspect of the dilated intrahepatic bile ducts and highlight possible complications such as pancreatitis and classify it.

Whereas the MRI has the great sensitivity to study the whole of the biliary tree, with the communications between the various cystic formations and the channels of normal gauge (10).

The principal differential diagnosis of Caroli's disease is caroli's syndrome, the diffuse complex form including congenital hepatic fibrosis and polycystic kidney disease (3).

The treatment of the patient with caroli disease depends on the type of pathology, and the location of biliary abnormalities. The forms located in the right or left lobe of the liver require a hemihepatectomy. Diffuse involvement can use conservative treatment including endoscopic treatment with internal biliary bypass (11). Liver transplantation in patients with diffuse caroli disease may be optimal even if they are asymptomatic (10).

the poor prognosis and the sizeable morbidity and mortality rates after liver transplantation still suggest that the timely recognition of indications for surgical treatment is of major importance. Thus, the early diagnosis of the disease and differentiation between types I and II are of extreme importance to patient survival (7).

CONCLUSION:

Carol's disease is a congenital pathology that can cause very serious angiocholitis, pancreatitis, or hepato-biliary degeneration, and even cancer. The diagnosis of caroli's disease should be made early, and should not be overlooked in patients with cholangitis or pancreatitis. Imaging remains the excellent diagnostic tool, and management must take into account the type and location of the disease.

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