Case study

CAROLI DISEASE REVEALED BY ACUTE PANCREATITIS AND ANGIOCHOLITIS: CASE REPORT

Abstract

Caroli's disease is a cavernous ectasia of the intrahepatic bile ducts, characterized by the formation of intraductal lithiasis exposing to a high risk of infection. Two types have been described for caroli disease: Simple And the diffuse complex form. The treatment of the patient with caroli disease depends on the type of pathology. Diffuse involvement can use conservative treatment including endoscopic treatment with internal biliary bypass, and Liver transplantation in patients with diffuse caroli disease may be optimal

INTRODUCTION

Caroli's disease is defined as cavernous ectasia or saccular dilation 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 dilation 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 disease: Simple, localized form (often in the left hepatic lobe). And the diffuse complex form known as caroli syndrome (congenital hepatic fibrosis, polycystic kidney disease) (3)

Case presentation

We report the case of a 27 year old patient, 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.
On the biological assessment; the lipaemia was at 1136ui / l, CRP at 139.7mg/l, ASAT at 309ui / l, ALAT at 246ui / l, BT at 89.6 mg / l, BC at 43.2mg / l, GGT at 978 ui / l, PAL at 683 ui / l, Hb at 12g / dl, WBC at 22860 / mm³, Plt at 347000 / mm³, and TP was at 56%.

In the imagery; Abdominal scan showed a stage B pancreatitis of balthazar with multisegmentary cystic dilation 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: dilation 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 cm^3 with thin wall full of micro and macro-lithiasis.

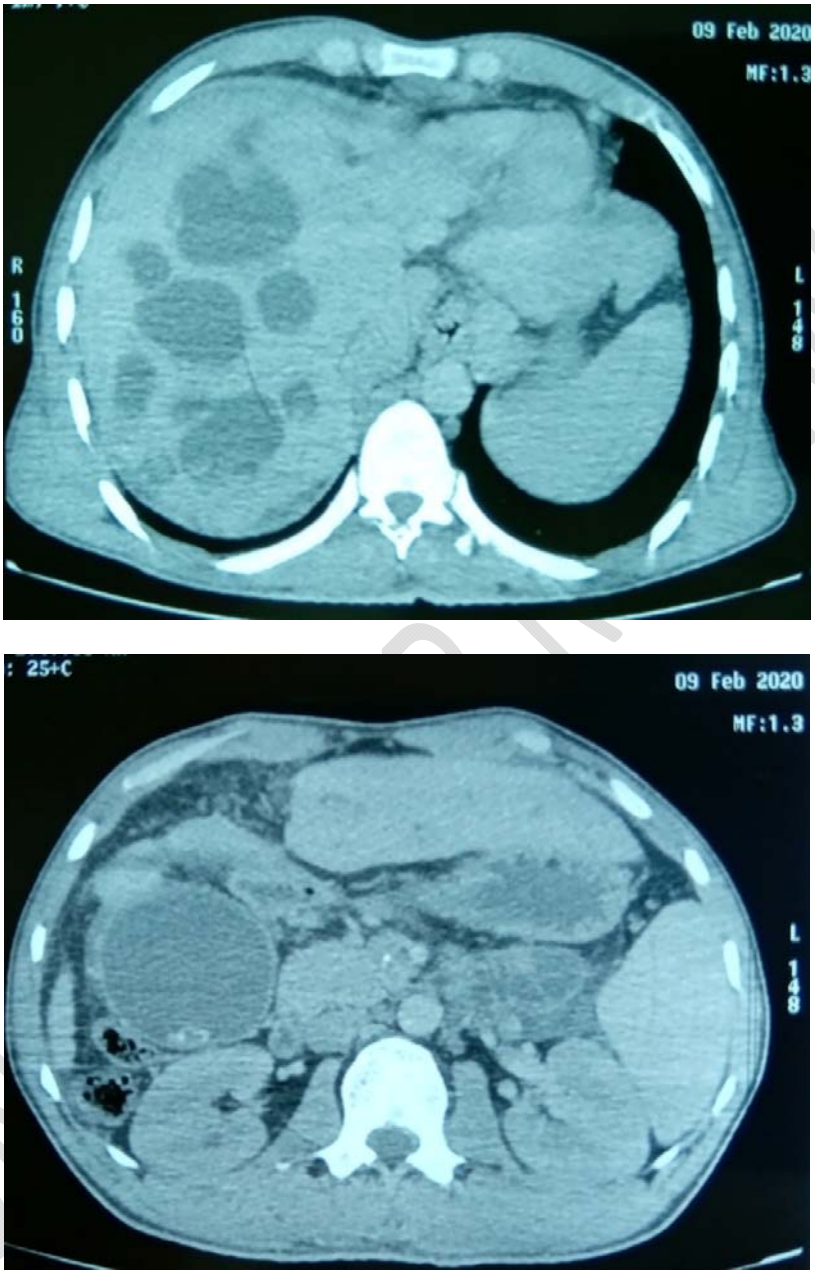

Fig. 1. MRI images

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 sieges of lithiasis. Main bile duct measuring 17mm in diameter with lithiasic content. Gallbladder measuring 6.28cm in anteroposterior diameter, with multiple lithiasis. Pancreatitis stage B from balthazar.

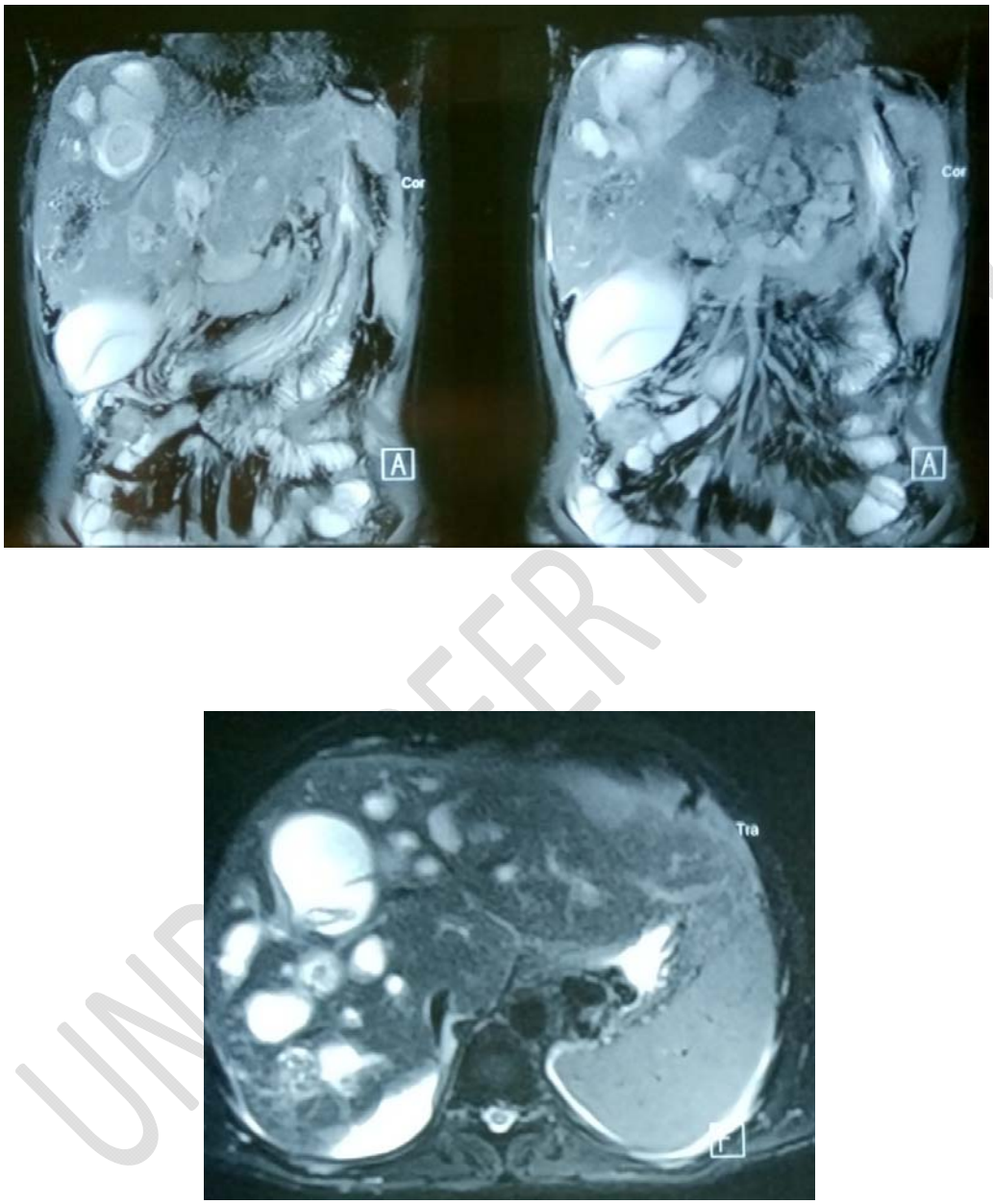

Fig. 2. MRI showing post-operative consequences

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 simples , 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, 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 dilation 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 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).

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 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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