

Caroli's disease: report of one case

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Abstract

Caroli disease is a condition characterized by cystic dilatation of the intrahepatic biliary tree that usually affects the entire liver parenchyma, although extrahepatic biliary involvement is rare. The following is the case of a male patient who was admitted due to cholestatic jaundice, low weight, and pruritus for 45 days. Magnetic resonance cholangiopancreatography reported multiple cystic dilatations in the intrahepatic bile ducts.

Keywords

Caroli disease; Intrahepatic bile ducts; Cholestasis; Jaundice.

INTRODUCTION

Caroli's disease (CD) was first described in 1958 by Jacques Caroli (1). Also known as communicating cavernous ectasia of the intrahepatic ducts, this is a congenital disease characterized by the dilatation of the bile ducts in the liver (2, 3). It is an autosomal recessive disorder, in which ultrasound reveals multiple cystic spaces throughout the liver parenchyma (4). The extrahepatic bile duct is usually not affected.

Its prevalence is estimated at 1 case per 1 000 000 inhabitants, and the most affected population are women with a ratio of 2:1 (5). If there is an association between

this pathology and liver fibrosis, such association is called Caroli's Syndrome (6). In that sense, CS is a bile duct cyst type V according to the Todani classification.

The most common clinical manifestations related to CD is recurrent cholestatic jaundice, associated or not with abdominal pain in the right upper quadrant, fever, pruritus, and sometimes recurrent cholangitis or multiple liver abscesses (5, 7, 8). The greater predisposition to stone formation in the intrahepatic bile duct seems to be related to the length of the illness, as well as to the degree of intrahepatic cholestasis (9, 10).

The present is the case report of a 42-year-old male patient, who was admitted to the emergency room due to

cholestatic jaundice, fever, and low weight. Imaging studies allowed establishing the diagnosis.

CASE REPORT

A 42-year-old male patient was referred to the emergency room from the Pisci prison (Chiclayo). The man reported a history of jaundice and choluria for approximately 45 days before consultation. Other symptoms, such as hyporexia, loss of 5 kg of weight, and general malaise were found. The day he was admitted to the emergency room, he also reported feeling moderate abdominal pain in the upper right quadrant.

He also reported a history of cholera at the age of 16, which required hospitalization for about 10 days at the Hospital de Sullana in Piura. The patient also had a history of drug (marijuana), tobacco and alcohol use. However, he denied having comorbidities associated with these habits.

The patient also reported abdominal pain in the upper right quadrant, which appeared sporadically since he was 15, but he denied having changes in skin color or suffering from jaundice in mucous membranes.

On physical examination upon admission, the man had blood pressure of 100/50 mm Hg, heart rate of 112 bpm, and temperature of 36.4 °C. A yellow color was evident in the skin and mucous membranes, which was compatible with the long-lasting jaundice he experienced and with some excoriations caused by scratching.

Pain in the abdomen on palpation was also evident in the upper right quadrant, but no hepatomegaly was observed since liver dullness was 11 cm. The patient was awake, oriented in time, space, and person, with no signs of focal neurologic deficits. The rest of the test was normal.

On the other hand, the liver function test reported hyperbilirubinemia in a predominantly conjugated form (bilirubin: 18.42 mg/dL; direct bilirubin: 13.41 mg/dL; elevated transaminases: glutamic oxaloacetic transaminase [GOT]: 1460 IU; glutamic pyruvic transaminase [GPT]: 1280 UI; and alkaline phosphatase: 453.8 UI/L).

Proteins were not affected, nor was prothrombin time. Tests for human immunodeficiency virus (HIV), hepatitis B and C were negative. The abdominal ultrasound suggested the presence of multiple simple liver cysts and biliary sludge, without determining the existence of lithiasis. The markers for carbohydrate antigen 19-9 (Ca19-9), carcinoembryonic antigen (CEA) and alpha-fetoprotein (AFP) were in the normal range.

During his hospital stay, a contrast-enhanced magnetic resonance cholangiography was performed, finding multiple saccular dilatations in the intrahepatic bile ducts: CD (type V according to the Todani classification) (**Figure 1**)

The work plan included the performance of an endoscopy of the upper digestive tract and biopsies of the stomach

and duodenum. Based on such tests, evidence of moderate erythematous gastritis was found. Finally, the results of biopsy taken confirmed the endoscopic diagnosis.

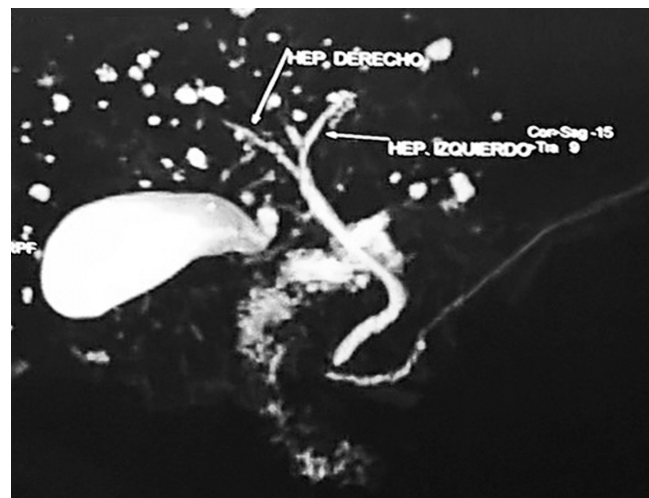


Figure 1. Multiple saccular dilatations involving intrahepatic bile ducts throughout the liver parenchyma. The extrahepatic bile duct is preserved, with no evidence of dilatation. Bile sludge is observed in the gallbladder, without stones.

DISCUSSION

The diagnosis of CD is mainly based on imaging findings, which show distal dilatations dependent on intrahepatic bile ducts. Cystic dilatation of the bile duct is commonly observed in ultrasound and computed tomography (CT) (11).

Also common is the imaging of adjacent intrahepatic stones, which occurs due to the stasis generated. However, recognizing the symptoms associated with liver problems is relevant.

CD is one of the fibrocystic ductal plate diseases, which also include autosomal recessive polycystic kidney disease, congenital hepatic fibrosis, autosomal dominant polycystic kidney disease, biliary hamartoma, and mesenchymal hamartoma.

In that context, ductal plate represents the angle of the intrahepatic biliary system (**Table 1**). Remodeling of the ductal plate, in mature intrahepatic biliary dilatation, follows a complex series of precisely timed events. In the case of CD, incomplete ductal plate remodeling involves the larger bile ducts (interlobular and more central). In CD, the smaller peripheral intrahepatic ducts are affected, but they remodel during embryonic life and manifest with coexisting liver fibrosis. A pattern of segmental inflammation and stenosis then occurs, alternating with saccular and fusiform dilatation of the affected ducts (5, 7, 9).

The differential diagnosis includes severe biliary dilatation generated by any of the other causes of biliary obstruction.

Table 1. Fibrocystic ductal plate diseases

Disease	Liver involvement
Congenital hepatic fibrosis	Progressive fibrosis of the portal vein with portal hypertension; association with Caroli disease
Autosomal recessive polycystic kidney disease	Chronic liver failure
Autosomal dominant polycystic kidney disease	Non-communicating bile duct cysts
Autosomal dominant polycystic kidney disease	Cysts derived from biliary microhamartomas and periductal glands
Caroli disease	Cystic dilatation of segmental intrahepatic bile ducts
Bile duct cyst	Intra- and extrahepatic bile duct involvement
Biliary hamartoma	Dilated ducts embedded in the fibrous stroma

Adapted based on reference 11.

tion, polycystic liver disease (in which the cysts have no communication with each other's intrahepatic bile ducts), and congenital hepatic fibrosis.

Other noninvasive imaging tests, such as 99m Tc-HIDA scan, may help reach the diagnosis (12), but it has been speculated that CD, as well as polycystic disease and congenital hepatic fibrosis, are all parts of the same spectrum (10, 11). Therefore, a cholangioresonance or a computerized axial tomography (CAT) scan may be needed to achieve a specific diagnosis.

Treatment should be individualized and should consider the management of associated complications, such as recurrent cholangitis, liver abscesses, stone formation, and the development of cholangiocarcinoma (12, 13). In some cases, cholestatic jaundice is favored by ursodeoxycholic acid (7, 9, 11). Furthermore, sometimes the lobar duct is affected,

in which scenario surgical resection is appropriate, but in diffuse cases, transplantation may be considered curative.

Thus, the clinical case presented here met the findings compatible with CD. From a clinical perspective, treatment of symptoms and regular assessment were established given the risk of intraluminal obstruction or infectious processes such as acute cholangitis. The patient was discharged after being administered ursodeoxycholic acid at a dose of 500 mg every 8 h for the 10 days of his hospital stay, during which time bilirubin values decreased notably, and a prescription of ursodeoxycholic acid for outpatient management.

In summary, patients with CD must be closely monitored to avoid complications, especially infections, which could increase morbidity and mortality or the need for high-cost treatments —such as liver transplantation— or therapies that worsen the patient's quality of life, such as liver lobectomy.

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