



Caroli's Disease Revealed by Acute Pancreatitis and Angiocholitis: Case Report

**Abdelilah El Bakouri¹, Abderrahmane Lamnaouar^{1*}, Othmane Elyamine¹,
Mounir Bouali¹, Khalid Elhattabi¹, Fatimazahra Bensardi¹
and Abdelaziz Fadil¹**

¹*Emergency Department, University Hospital Center Ibn Rochd, Faculty of Medicine and Pharmacy;
Hassane II University, Casablanca, Morocco.*

Authors' contributions

This work was carried out in collaboration among all authors. Author AEB designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors AL and OE managed the analyses of the study. Author AL managed the literature searches. All authors read and approved the final manuscript.

Article Information

Editor(s):

(1) Dr. Pramod Kumar Sharma, All India Institute of Medical Sciences (AIIMS), India.

Reviewers:

(1) Raluca Simona Costache, Carol Davila University of Medicine, Romania.
(2) Deepika Lather, Lala Lajpat Rai University of Veterinary & Animal Sciences, India.
Complete Peer review History: <http://www.sdiarticle4.com/review-history/61880>

Case Study

Received 15 August 2020
Accepted 19 October 2020
Published 10 November 2020

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.

Keywords: Caroli's disease (CD, MRI, CT scan); chronic smoker.

*Corresponding author: Email: lamnaouar07@gmail.com;

1. 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].

2. 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.

2.1 Laboratory Indicators

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.

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.

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.

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

Lipaemia	1136 iu / l (Elevated)
CRP	139.7mg/l (Elevated)
Complete blood counts	
Hemoglobin	12mg/dl
WBC	22860 / mm ³
Plt	345000/ mm ³
Liver function	
Elevated alanine aminotransferase	
Elevated aspartate aminotransferase	246 iu / l
Elevated total bilirubin	309 iu / 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%

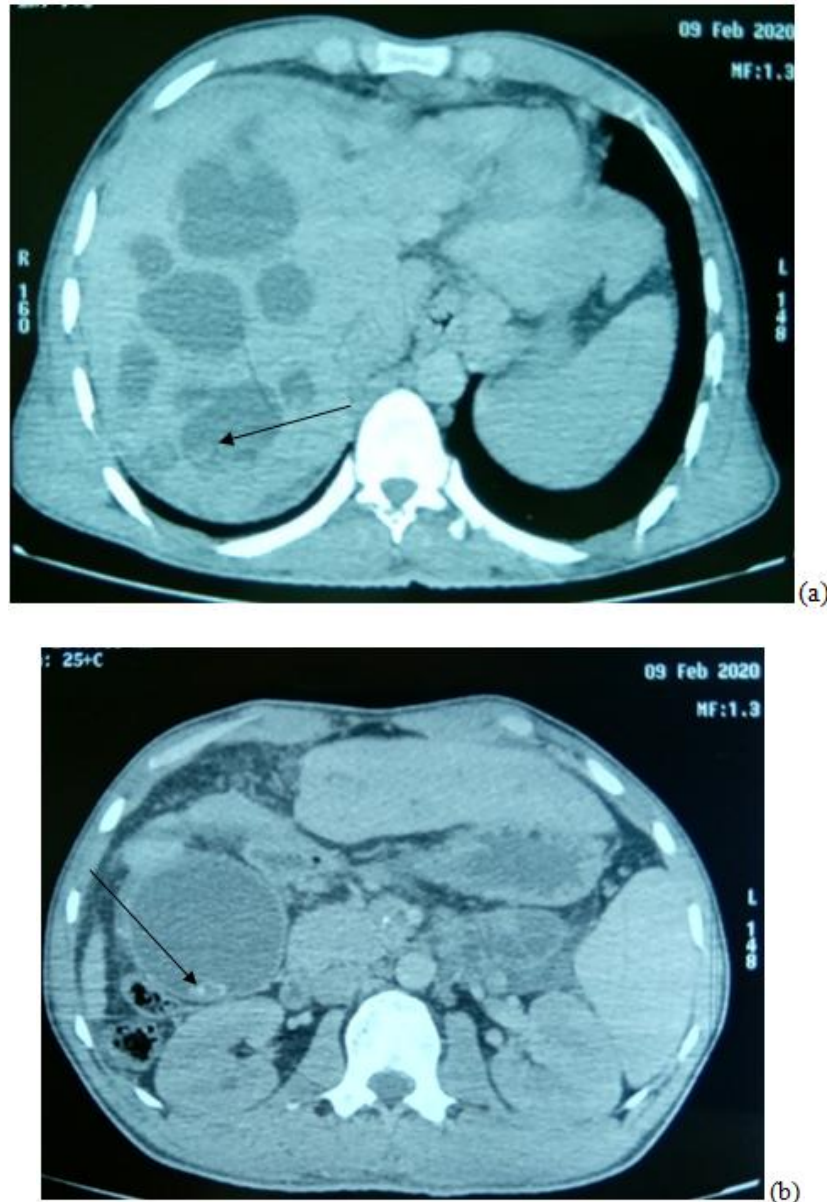


Fig. 1. Computed tomography in a 27-year-old man with Caroli's disease showing secular ectasias and central dotting (a) with distended gallbladder containing multiple lithiasic formations (b)

3. 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].

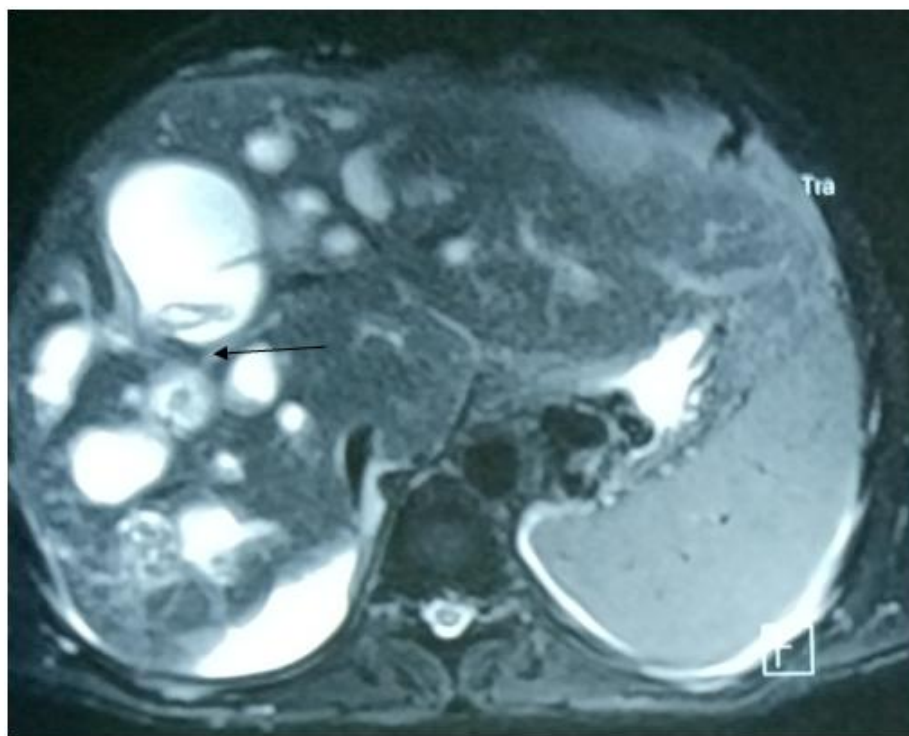


Fig. 2. Magnetic resonance in the same patient shows the connection between the secular ectasias and the normal biliary tract

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].

4. CONCLUSION

Caroli'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.

CONSENT

As per international standard or university standard guideline participant consent has been collected and preserved by the authors.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Caroli J, Soupault R, Kossakowski J, et al. Congenital polycystic dilation of the intrahepatic bile ducts; attempt at classification. La semaine des hopitaux: organe fonde par l'Association d'enseignement medical des hopitaux de Paris. 1958;34(8/2)488.
2. Fulcher AS, Turner MA, Sanyal AJ. Case 38: Caroli disease and renal tubular ectasia. Radiology. 2001;220:720–723.
3. Caroli J. Diseases of the intrahepatic biliary tree. Clin Gastroenterol. 1973;2:147–161.
4. Bettini G, Mandrioli L, Morini M. Bile duct dysplasia and congenital hepatic fibrosis associated with polycystic kidney (Caroli syndrome) in a rat. Vet Pathol. 2003;40:693–694.
5. De Tommaso AM, Santos DS, Hessel G. Caroli's disease: 6 case studies. Acta Gastroenterol Latinoam. 2003;33:47–51.
6. Desmet J. Pathogenesis of ductal plate abnormalities. Mayo Clin Proc. 1998;73:80–89.
7. Wu KL, Changchien CS, Kuo CM, Chuah SK, Chiu YC, Kuo CH. Caroli's disease – a report of two siblings. Eur J Gastroenterol Hepatol. 2002;14:1397–1399.
8. Ozlem Y, Yusuf B. Clinical characteristics of Caroli's disease. World J Gastroenterol. 2007;13:1930–1933.
9. Wang, Zhong-Xia, LI, Yong-Gang, Wang, Rui-Lin, et al. Clinical classification of Caroli's disease: an analysis of 30 patients. HPB. 2015;17(3):278–283.
10. Kassahun WT, Kahn T, Wittekind C, Mossner J, Caca K, Hauss J et al. Caroli's disease: liver resection and liver transplantation. Experience in 33 patients. Surgery. 2005;138:888–898.
11. Desmet VJ. Pathogenesis of ductal plate malformation. JGastroenterol Hepatol. 2004;19(Suppl.):356–360.

© 2020 Bakouri et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/4.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

*The peer review history for this paper can be accessed here:
<http://www.sdiarticle4.com/review-history/61880>*