Scientific Research Publishing

Open Journal of Internal Medicine, 2026, 16(3), 378-383
https://www.scirp.org/journal/ojim
ISSN Online: 2162-5980
ISSN Print: 2162-5972

Delay in Diagnosis of Caroli's Disease: About a First Case in Internal Medicine at Donka University Hospital

Mamadou Diakhaby, Mohamed Cissoko, Kalil Nouny Sidibé, Mohamed Lamine Conté, Thierno Amadou Wann, Mamadou Lamine Yaya Bah, Lanciné Kourouma, Aboubacar Dioubaté, Sâa Joseph Téliano, Mohamed Adama Oularé, Amara Magassouba, Kanté Mamadou Aliou II, Diallo Mamadou Tafsir, Idrissa Diallo, Abraham Geopogui, Oumar Camara, Abdourahmane Diallo, Elhadj Salmana Diallo, Amadou Baillo Barry, Fatoumata Bah, Djibril Sylla, Amadou Kaké

Internal Medicine Department, Donka National Hospital, Conakry, Guinea

Email: Mamsdiakhaby@gmail.com, Mohamed.cissoko25nov@gmail.com, Kalilsidibe3@gmail.com, docteurconte@yahoo.fr, thwann@yahoo.fr, mlambah1@yahoo.fr, Lasskourouma99@gmail.com, Samksell2013@gmail.com, Josephteliano74@gmail.com, mohadani10@gmail.com, magassoubaamara125@gmail.com, Alioukante1993@gmail.com, diallomamadoutafsir378@gmail.com, idrissanegue@gmail.com, abrahamg.ag38@gmail.com, camaraoumar.oc69@gmail.com, Pelloun1989@gmail.com, salmanadiallo2014@gmail.com, amadoubailob707@gmail.com, Bahfatimatou11@gmail.com, docdjibril@gmail.com, amadoukake@gmail.com

How to cite this paper: Diakhaby, M., Cissoko, M., Sidibé, K.N., Conté, M.L., Wann, T.A., Bah, M.L.Y., Kourouma, L., Dioubaté, A., Téliano, S.J., Oularé, M.A., Magassouba, A., Aliou II, K.M., Tafsir, D.M., Diallo, I., Geopogui, A., Camara, O., Diallo, A., Diallo, E.S., Barry, A.B., Bah, F., Sylla, D. and Kaké, A. (2026) Delay in Diagnosis of Caroli's Disease: About a First Case in Internal Medicine at Donka University Hospital. *Open Journal of Internal Medicine*, **16**, 378-383.
https://doi.org/10.4236/ojim.2026.163029

Received: July 26, 2026
Accepted: August 28, 2026
Published: August 31, 2026

Abstract

Caroli disease is a rare congenital condition characterized by multifocal segmental dilatation of the intrahepatic bile ducts, described in 1958 by Jacques Caroli due to a genetic mutation. It is a cause of chronic cholestasis and intrahepatic lithiasis in children and young adults. We report the case of Caroli's disease diagnosed in a 36-year-old patient. The onset of symptoms was gradual and began three months prior (January 10, 2023), marked by the onset of jaundice, persistent fever, early postprandial vomiting without any apparent triggering factors, associated with mild epigastric pain without radiation that subsided when vomiting stopped, and prostration. She consulted a health facility in Nongo on January 15, 2023, where a diagnosis of severe malaria was made based on the jaundice, fever, and vomiting. She was treated with artesunate 60 mg, sodium chloride 0.9%, and vogalene 10 mg. After a period of remission, on January 19, 2023, she experienced a fixed, moderate pain in her right hypochondrium without radiation, and non-bloody, watery diarrhea. On January 19, 2023, she was using self-medication based on tramadol 50 mg and amoxicillin 1000mg. Given the persistence of the aforementioned symptoms after an unsuccessful course of treatment since January 10, 2023, she was referred to Internal Medicine on April 20, 2023, hence her hospitalization, admitted for

watery diarrhea without mucus or blood without tenesmus or straining more than 4 times at a time, early postprandial food vomiting, jaundice, fixed pain in the right hypochondrium of moderate intensity without triggering or relieving factors, fever, permanent dizziness, not diabetic, nor hypertensive, with a history of pulmonary TB on microscopy (−) in 2016 treated and declared cured. The patient's general condition was preserved, with normal skin and conjunctiva. Scleral jaundice was noted, but there was no venous circulation, ascites, lower limb edema, or splenomegaly. Physical examination revealed tenderness in the right hypochondrium and epigastrium without a palpable mass, and a normal cardiorespiratory function. Laboratory tests showed cholestasis (Total Bilirubin: 2.3N and Conjugated Bilirubin: 2.3N, Alkaline Phosphatase: 1.4N, GGT: 1.8N), cytolysis (ALT: 3.4N and AST: 4.4N), and viral markers: HBsAg (−), total Anti-HBc (−), Anti-HCV (−). Abdominal and pelvic ultrasound revealed heterogeneous hepatomegaly due to the presence of a biliary cyst in the right lobe and segmental dilation of the intrahepatic bile ducts. She received 0.9% saline, 500 mg metronidazole, 1 g ceftriaxone, 80 mg spasfon, 1000 mg paracetamol, and 200 mg Ursolvan. Her condition improved significantly, particularly after the administration of ursodeoxycholic acid.

Keywords

Caroli's Disease, Young Woman, Internal Medicine, Donka University Hospital

1. Introduction

Caroli disease is a rare congenital disorder characterized by multifocal segmental dilation of the intrahepatic bile ducts, first described in 1958 by Jacques Caroli. It is generally attributed to a genetic mutation, although an intrauterine malformation of the bile ducts causing inflammation is also discussed, as well as a recessive or dominant genetic alteration [1]-[3]. Caroli syndrome is defined as its association with congenital hepatic fibrosis; it is a cause of chronic cholestasis and intrahepatic gallstones in children and young adults [2] [4]. Monolobar Caroli disease may be present at birth or remain asymptomatic until the age of 45. The left lobe is most often affected, and it is monolobar in 20% of cases (92% left and 8% right). The prevalence is estimated at 1/1,000,000 cases. It represents less than 1% of congenital cystic dilatations of the bile ducts [1] [5]. We report the case of Caroli's disease diagnosed in a 36-year-old patient in the Internal Medicine department at Donka University Hospital.

2. Observation

This is a 36-year-old female entrepreneur who consulted on April 20, 2023 for watery, non-mucusy and non-bloody diarrhea without tenesmus or straining more than 4 times/day, early postprandial food vomiting without triggering factors, fixed pain in the right hypochondrium of moderate intensity without trig-

gering or relieving factors, fever, permanent dizziness, evolving for 3 months (January 10, 2023) with a history of pulmonary TB on microscopy (−) in 2016 treated and declared cured, no history of jaundice, scarification, or known liver disease. The patient had no known diabetes or hypertension, and her lifestyle did not include alcoholism or smoking. The clinical examination revealed the following parameters: Blood Pressure: 100/63 mmHg, Heart Rate: 127 Beats/min, SpO2: 97%, T = 38.2˚C, Respiratory Rate: 20 cycles/min, Fasting Blood Glucose: 1.04 g/l, Weight: 89 kg, Height: 1.80 m, BMI: 27 Kg/m^2.

The patient's general condition was good, with a normal-colored conjunctiva. Physical examination revealed tenderness in the right hypochondrium without a palpable mass, hepatomegaly, or splenomegaly, and no collateral venous circulation or normal cardiorespiratory function. The rest of the examination was unremarkable. A diagnosis of amebiasis with intestinal and hepatic involvement (liver abscess) was made. She was treated with Flagyl 500 mg, Paracetamol 1 g, and 0.9% saline for 7 days of hospitalization. Without improvement, jaundice, dark urine, and normal-appearing stools developed, with no pruritus. Clinically, scleral jaundice, tenderness in the right hypochondrium, and painful hepatomegaly were noted, with a smooth surface, soft consistency, and regular lower borders. Initially, we considered cholangitis (given jaundice, fever, and pain in the right hypochondrium), obstructive cholangitis, and primary sclerosing cholangitis in the presence of incomplete clinical cholestasis syndrome (jaundice, dark urine). An ultrasound was performed, revealing heterogeneous hepatomegaly due to the presence of a biliary cyst in the right lobe and segmental dilation of the intrahepatic bile ducts. Diagnoses of cholangitis, obstructive cholangitis, and primary sclerosing cholangitis were ruled out based on ultrasound findings that suggested segmental dilation of the intrahepatic bile ducts, which were not specific to the suspected diagnoses. The diagnosis of Caroli's disease was made based on the cholestasis syndrome and the segmental dilation of the bile ducts, suggestive of this condition.

Laboratory tests revealed cholestasis (Total Bilirubin: 2.3N and Conjugated Bilirubin: 2.3N, Alkaline Phosphatase: 1.4N, Gamma-glutamyl transferase: 1.8N) and cytolysis (Alanine Aminotransferase: 3.4N and Aspartate Aminotransferase: 4.4N). Viral markers: HBsAg (−), Total Anti-HBc (−), Anti-HCV (−). Hemoglobin: 9.7 g/dL, MCV: 88.1 fL, MCH: 31.2%, MCHC: 35.5 g/dL, Leukocytes: 23 × 10^9/L (90.7% PMN, 5.2% Lymphocytes), Platelets: 75 G/L, Creat: 74 micromol/L, K^+: 3.96 mmol/L, Na^+: 143 mmol/L, Ca^+: 2.37 mmol/L, Mg^{2+}: 0.72 mmol/L. Blood Group: B−, BU negative, HIV serology negative.

The diagnosis of Caroli's disease was made based on clinical arguments (right hypochondrium pain, fever, jaundice, vomiting, diarrhea), confirmed by abdominal ultrasound while awaiting the result of the abdominal scan.

She received 0.9% saline solution, metronidazole 500 mg, ceftriaxone 1 g, Spasfon 80 mg, paracetamol 1000 mg, and Ursolvan (ursodeoxycholic acid) 200 mg for 10 days. Her condition improved significantly, with a marked improvement in her clinical state.

3. Discussion

Caroli's disease is a rare entity (<1/1,000,000 in the general population) whose clinical and imaging data are studied in the literature based on small patient series (**Figure 1**). The clinical manifestations are not specific to Caroli's disease, leading to diagnostic errors [1] [5]. Clinical symptoms are variable, but right upper quadrant pain, fever, jaundice, pruritus, vomiting, and diarrhea are frequently observed [3]. These signs were found in our patient, which corroborates the data in the literature.

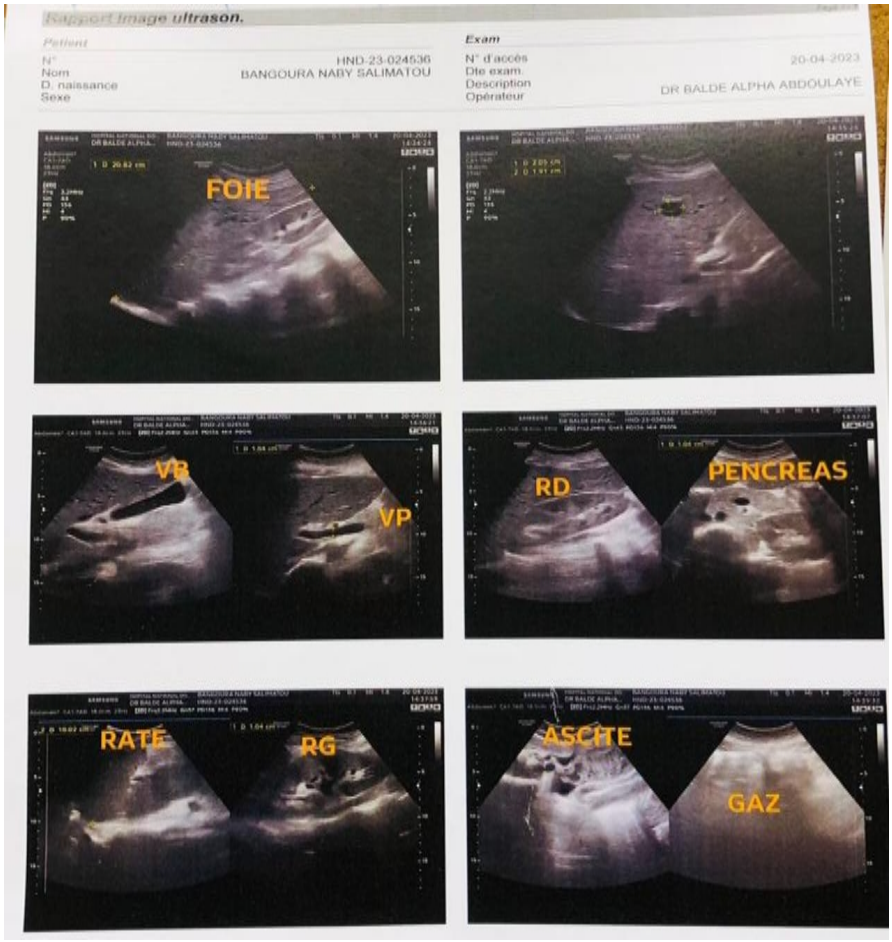

Figure 1. Abdominal-pelvic ultrasound consistent with Caroli's disease.

Sometimes, the condition is discovered incidentally during an abdominal ultrasound or intraoperative cholangiography during cholecystectomy for gallstones; ultrasound and computed tomography allow for diagnosis [1]. In our case, we used an abdominopelvic ultrasound, which allowed us to identify this pathology in the absence of any known history of liver disease. Our limitations included the inability to perform an abdominopelvic CT scan, which would have revealed communication between the cystic spaces and the biliary tree, as well as liver biopsy and cytoculture, which were unavailable in the area. As the literature states, apart from abdominal CT scans, abdominal ultrasound can confirm the diagnosis.

The usual medical treatment involves antibiotics and bile solvents (ursodeoxycholic acid), as well as bile duct drainage techniques. Biliary drainage via endoscopic, radiological, or surgical methods is often associated with high morbidity and mortality due to infectious complications and a high recurrence rate [1] [3]. The risk of developing intrahepatic cholangiocarcinoma is 100-fold increased in patients with Caroli's disease/Caroli's syndrome; moreover, it is very difficult to detect malignant transformation within dilated bile ducts [1] [3]. She received 0.9% saline, metronidazole 500 mg, ceftriaxone 1 g, Spasfon 80 mg, paracetamol 1000 mg, and Ursolvan (ursodeoxycholic acid) 200 mg.

The evolution was favorable, with a marked improvement in his clinical condition before his discharge; the cholestasis and cytolysis levels had returned to normal, and all clinical signs had regressed, namely jaundice. The planned imaging follow-up was an abdominal scan.

4. Conclusion

Caroli's disease is a congenital, non-obstructive dilation of the intrahepatic bile ducts. Diagnostic delays are due to the absence of specific clinical signs. Diagnosis is made by abdominal ultrasound or CT scan, which reveals asymmetrical dilation of the bile ducts.

Author Contributions

Summary, Introduction, Methods, Results, Conclusion, Review: Dr Mamadou Diakhaby, Dr Mohamed Cissoko, and Dr Kalil Nouny Sidibé; First Proofreading: Dr Mohamed Lamine Conté, Dr Wann Thierno Amadou, Dr Bah Mamadou Lamine Yaya, Dr Aboubacar Dioubaté, Dr Lanciné Kourouma, Dr Sâa joseph Téliano, Dr Mohamed Adama Oularé, Dr Amara Magassouba, Dr Kanté Mamadou Aliou II, Dr Diallo Mamadou Tafsir, Dre Idrissa Diallo, Dr Abraham Geopogui, Dr Oumar Camara, Dr Abdourahmane Diallo, Dr Elhadj Salmana Diallo, Dr Amadou Baillo Barry, and Dr Fatoumata Bah. Second proofreading with advice: Pr Djibril Sylla and Pr Amadou Kaké. All authors have read and agreed to the published version of the manuscript.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] El Alami, Y., Benamr, S., Mouslik, R., Massrouri, R., Essadel, E., M'daghri, J., *et al.* (2010) Place de la chirurgie dans la maladie de Caroli monolobaire. *Journal Africain d'Hépato-Gastroentérologie*, **4**, 152-155. https://doi.org/10.1007/s12157-010-0173-y

[2] Bouzidi, A., Lezrek, M., Kabiri, H., Zraza, I., Boumdin, H., Rimani, M., Azennag, M., *et al.* (1997) Maladie de caroli à propos d'un cas localisé au lobe gauche avec revue de la littérature. *Médecine du Maghreb*, **65**, 40-42. https://www.santetropicale.com/Resume/6505.pdf

[3] Fahrner, R., Dennler, S.G.C., Dondorf, F., Ardelt, M., Rauchfuss, F. and Settmacher,

U. (2019) Résection et transplantation hépatique pour maladie de Caroli et syndrome de Caroli. *Journal de Chirurgie Viscérale*, **156**, 107-112. https://doi.org/10.1016/j.jchirv.2018.01.004

[4] Gillet, M., Favre, S., Fontolliet, C., Halkic, N., Mantion, G. and Heyd, B. (1999) Maladie de Caroli monolobaire. À propos de 12 cas. *Chirurgie*, **124**, 13-19. https://doi.org/10.1016/s0001-4001(99)80037-3

[5] Kettabi, F.Z., Benzzoubeir, N., Errabih, I., Ouazzani, L. and Ouazzani, H. (2017) Maladie et syndrome de Caroli: Deux entités rares (à propos de 5 cas). *Journal Marocain des Sciences Médicales*, 2**1**, 20-25. https://doi.org/10.48401/IMIST.PRSM/jmsm-v21i1.7630