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Left Monolobar Caroli's Disease: A Case Report

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Abstract | **Case Report**

Caroli's disease is a rare condition. It is characterized by multifocal dilatations of intrahepatic segmental bile ducts. This pathology can be complicated by lithiasis and hepatic abscesses. It is most often diffused, but can be localized mainly in the left lobe. We report a case of left monolobar Caroli's disease revealed by a febrile mild jaundice in a patient in his sixties, with an interval of 2 months between the first symptom and the diagnosis, which was suggested by compelling MRI findings and confirmed by the anatomopathological study. The patient underwent a left hepatic lobectomy without apparent complications the following 6 months. Despite the rarity of Caroli's disease, this case illustrates its typical form in the literature.

Keywords: Caroli's disease, Intrahepatic biliary tract dilatation, Cholangitis, Jaundice, case report.

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INTRODUCTION

Caroli's disease is a cystic dilatation of the segmental intrahepatic bile ducts, first described by Caroli and Couinaud in 1958. It is a rare condition, with a prevalence of 1 in 1,000,000 in the general population [1]. It represents the second most common congenital malformation of the biliary tract after biliary atresia [2].

The disease usually occurs in its isolated form, known as the "pure" form, but it may also be associated with other hepatobiliary or renal malformations. The most frequently associated malformations are congenital hepatic fibrosis and polycystic kidney disease. This association justifies the use of the term "Caroli's syndrome" [1].

The disease may affect the liver diffusely in 80% of cases or be localized to a lobe or segment in 20% of cases, of which 92% are in the left lobe [3].

We present a case of Caroli's disease confined to the left hepatic lobe.

CASE REPORT

A man in his sixties reported experiencing hepatic colic, accompanied by intermittent prolonged fever beginning two months prior to consultation. However, the symptom that prompted him to seek medical attention was conjunctival jaundice.

The patient had insulin-dependent diabetes and a surgical history that included bilateral inguinal hernia repair, appendectomy, and prostatectomy.

Upon consultation, the patient was apyretic, hemodynamically stable, and classified as WHO performance status 0, with mucosal jaundice, dark urine, and pale stools. Abdominal examination revealed tenderness in the right hypochondrium, while the rest of the physical exam was unremarkable.

Biologically, a CRP level of 41 mg/L was noted, indicating inflammation, along with an isolated cholestasis profile: alkaline phosphatase (ALP) at 1.6 times the normal level and gamma-glutamyl transferase (GGT) at 4.2 times the normal, with moderately elevated bilirubin but no hepatic cytolysis. Serologic tests for viral hepatitis and autoimmune markers were negative.

An initial outpatient ultrasound revealed a hepatic mass in the left lobe. A subsequent abdominal-pelvic CT scan showed a heterogeneous left hepatic lobe due to a large pseudo-nodular area occupying segments

II and III of the liver, containing very hypodense, cyst-like zones with some calcifications (Figure 1).

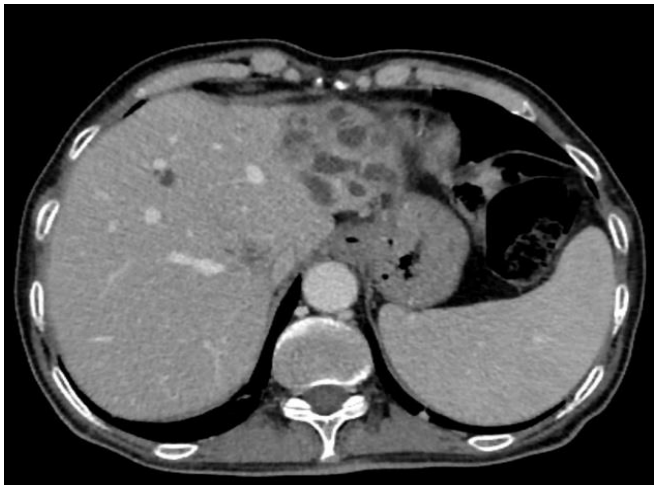

Figure 1: Abdominal CT scan: dilatation of the intrahepatic bile ducts in the left hepatic lobe

A hepatic MRI, performed for its superior sensitivity in detecting and characterizing hepatobiliary lesions, revealed a tight stricture of the proximal left hepatic duct near the main biliary confluence, harbouring a 16 mm macro-calculus, associated with cystic dilatation of the intrahepatic bile ducts in segments II and III, along with multiple bile stasis stones (Figures 2 and 3).

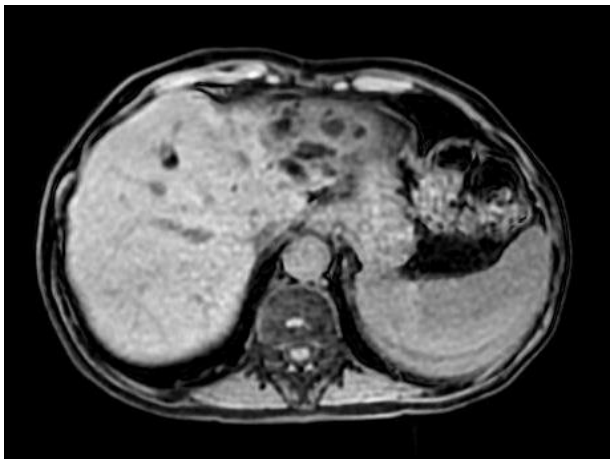

Figure 2: Abdominal MRI: cystic dilatations of the bile ducts in hepatic segments II and III

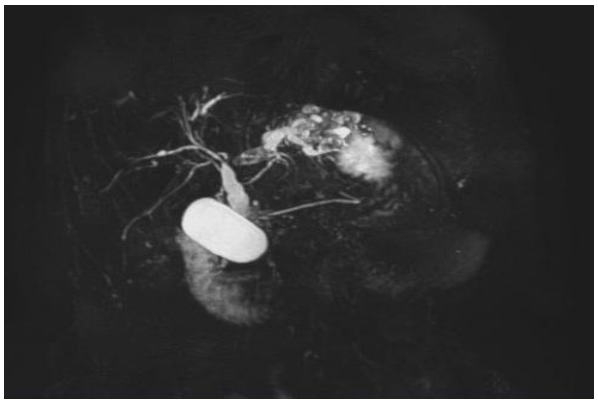

Figure 3: MR cholangiography: cystic dilatations of the left intrahepatic bile ducts with multiple stones

An abdominal CT angiography revealed:

- Signs of severe diffuse cholangitis in the left lobe, with cystic dilatations and micro-abscesses at the site of the left lateral sectoral duct stricture.

- Significant atrophy of the left hepatic lobe with compensatory hypertrophy of the right lobe.
- Perfusion abnormalities in the left hepatic parenchyma, secondary to a chronic organized thrombosis of the left portal vein branch.
- A hepatic arterial anatomical variant: an accessory left hepatic artery arising from the left gastric artery, supplying the left hepatic lobe (Figure 4).

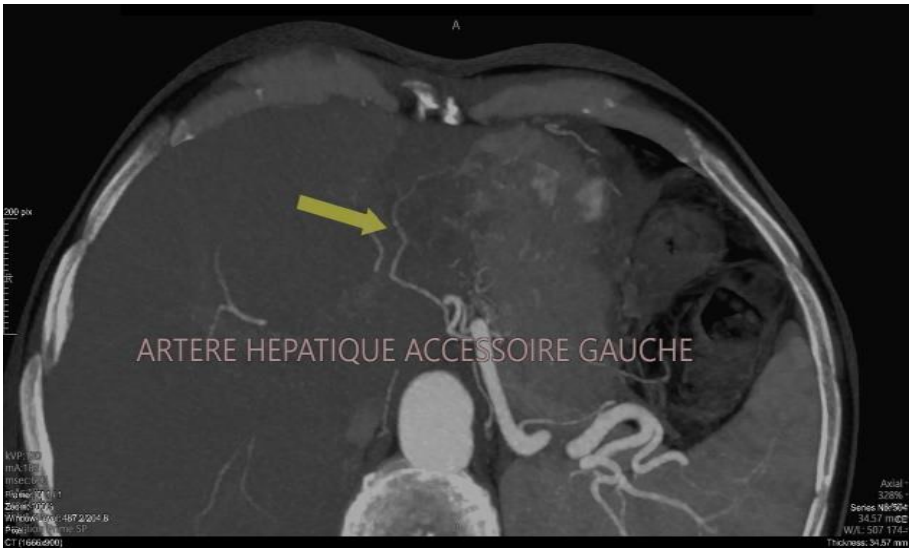

Figure 4: Abdominal CT angiography: accessory left hepatic artery originating from the left gastric artery

- **Signs of portal hypertension**.

A liver biopsy showed no significant abnormalities except for glycogen overload, likely related to poorly controlled diabetes, and no evidence of malignant cells.

Based on the exploratory findings, a diagnosis of Caroli's disease in its localized left monolobar form was suggested; due to the presence of multiple intrahepatic cystic dilatations with stones and abscessed collections in the left lobe, along with recurrent cholangitis syndrome. A clear indication for surgery was identified, and a left hepatic lobectomy was performed via a conventional approach.

Histopathological examination of the resected specimen revealed ductular hyperplasia and multiple cystic dilatations of the intrahepatic bile ducts, upholding the diagnosis of Caroli's disease, with no signs of malignancy.

The postoperative course was uneventful, and the patient showed favourable clinical and biological progress one year after the surgery**.**

DISCUSSION

Caroli's disease is a rare condition characterized by multifocal segmental dilatation of the intrahepatic bile ducts [4]. It corresponds to type V of the Todani classification (Figure 5) [5].

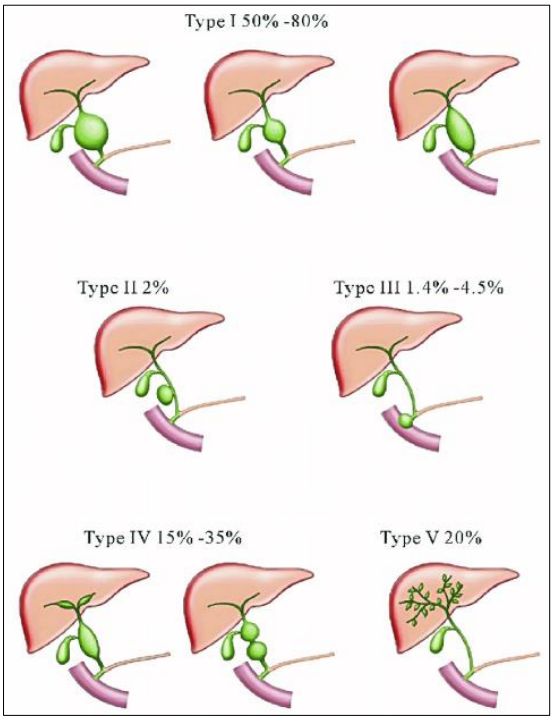

Figure 5 : Todani *et al.*, Classification [5]

Two forms are recognized: a diffuse form, which is the most common, and a localized form, involving a single lobe in only 20% of cases, of which 92% affect the left lobe—as was the case with our patient.

The most widely accepted hypothesis involves an embryological defect in ductal remodelling, leading to discontinuous and irregular dilatations along the biliary

tree. If this anomaly occurs early in embryonic development, it affects the right or left hepatic ducts or segmental ducts, resulting in pure Caroli's disease. When it appears later, it involves subsegmental ducts and leads to congenital hepatic fibrosis, thus forming Caroli's syndrome [1].

Despite being congenital, the disease is often diagnosed in adulthood, due to the long clinical latency period and the lack of specific symptoms [6]. In reality, it often remains asymptomatic for years, but manifests before the age 30 in 80% of cases [7]. It affects both men and women equally [8]. In our case, the patient was a man in his sixties at diagnosis.

Caroli's disease frequently presents with recurrent fever, jaundice, and pain in the right hypochondrium [8]. Clinical features are dominated by recurrent cholangitis, seen in 64% of patients. The disease often evolves with intra- and extrahepatic bile stones, which may lead to hepatic abscess, cholangitis, and, in the long term, cholangiocarcinoma. Our case exemplifies the typical clinical profile described in the literature., with recurrent fever, conjunctival jaundice, and recurrent hepatic colic, indicating repeated episodes of cholangitis.

Cholangiocarcinoma is a serious but rare complication and may even be the initial presentation [6]. The incidence of malignancy ranges from 2.5% to 16% [3], representing a 100 times higher risk than in the general population. A hepatic tumor can also be seen with the disease in 7% to 24% of cases [4]. The presence of a choledocholithiasis without gallbladder stones should raise suspicion for Caroli's disease [1]. Sometimes, the disease is discovered incidentally during abdominal ultrasound or intraoperative cholangiography during cholecystectomy for gallstones [4].

Liver function tests often reveal elevated alkaline phosphatase (ALP) and direct bilirubin. Transaminase levels may initially be normal but can rise with progressive hepatic fibrosis, particularly in Caroli's syndrome [6]. In accordance with the literature, our patient exhibited biological cholestasis (elevated ALP and bilirubin) without hepatic insufficiency.

Ultrasound, followed by CT and MRI, helps guide the diagnosis. CT imaging shows cystic fluid-filled structures and can confirm their communication with bile ducts. MRI more clearly identifies cystic bile duct dilatations and differentiates them from vascular structures. These cysts often contain stones, leading to hepatobiliary complications [4].

Imaging also helps rule out hepatic polycystic disease, which lacks communication with bile ducts. This differential diagnosis can also be excluded using Tc-99m scintigraphy [9].

Fibrovascular bundles—containing small branches of the portal vein and a hepatic artery branch connected to the saccule—appear on imaging as contrast-enhancing dotted signs, known as the "Dot Sign" (Figure 6), considered pathognomonic of Caroli's disease [3]. This sign is defined as a small portal vein branch surrounded by dilated bile ducts [8]. In our patient, imaging findings were consistent with the literature, but the Dot Sign was absent, highlighting that although this sign is highly characteristic, it is not always present.

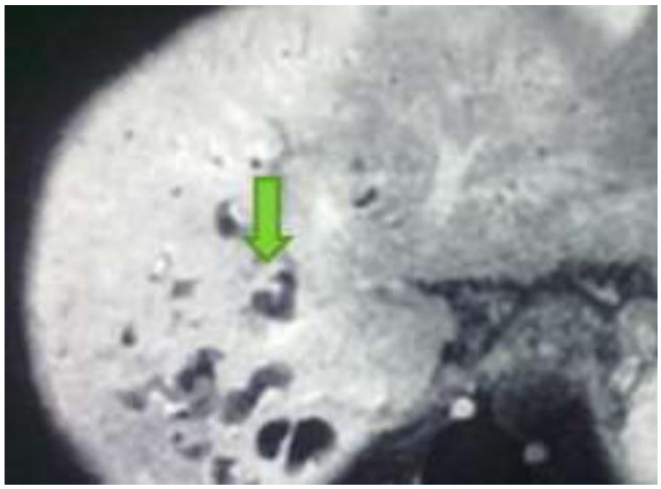

Figure 6: Portal-phase MR cholangiography: DOT SIGN [6]

Histopathological examination serves to confirm the diagnosis, identify associated lesions (especially congenital hepatic fibrosis), and screen for malignancy. Despite the specificity of radiological findings, diagnostic errors are sometimes corrected through histology. Precancerous or cancerous lesions must be thoroughly investigated [9].

A notable finding in our case was that the left liver lobe was atrophic, confirmed by hepatic volumetry (Figure 7). Furthermore, this lobe exhibited perfusion

abnormalities due to a chronic organized thrombosis of the left portal vein branch and received additional vascular supply from an accessory artery arising from the left gastric artery.

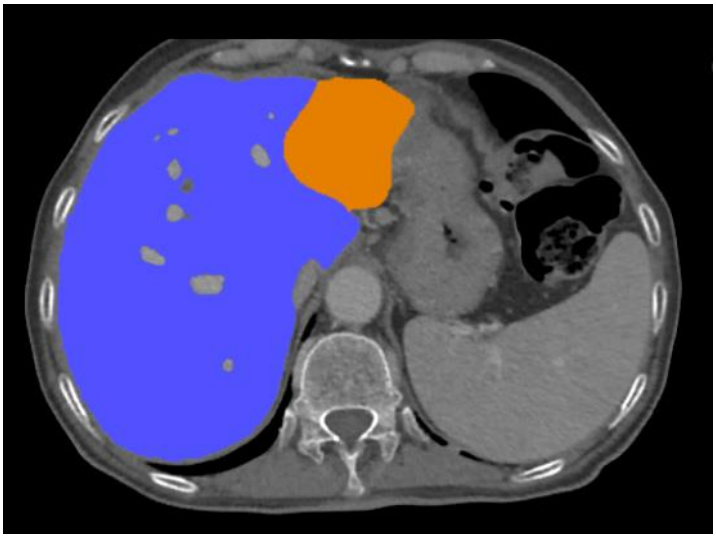

Figure 7: Hepatic volumetry: atrophy of the left hepatic lobe with hypertrophy of the right lobe in our patient

The treatment of monolobar Caroli's disease is complete surgical resection of the affected area. When the disease is confined to the left hepatic lobe, as in our case, left lobectomy is the standard and usually successful treatment [4]. This surgical approach definitively resolves the disease by eliminating the lithogenic focus, thereby preventing septic complications and malignant transformation [6].

In contrast, treatment of diffuse bilobar forms is more challenging and mainly medical, involving antibiotics and sometimes bile solvents [9]. However, if both lobes are affected and complications arise, hepatic lobectomy becomes ineffective, and liver transplantation should be considered [8].

The timing of the transplantation is a real dilemma: It is not advisable to subject a clinically stable patient to the risks associated with liver transplantation; however, delaying intervention until complications arise may result in a missed therapeutic window [6].

When the diffuse form coexists with congenital hepatic fibrosis, potentially leading to cirrhosis or portal hypertension, the diagnosis shifts to Caroli's syndrome [7]. This syndrome may be associated with polycystic kidney disease or Cacchi-Ricci disease. There are no morphological comparison studies between Caroli's disease and Caroli's syndrome in the literature; however, various clinical distinctions between the two entities have been described (Table I) [6].

Table I: Comparative table between Caroli's disease and Caroli's syndrome

Caroli's disease	Caroli's syndrome
No congenital hepatic fibrosis (by definition)	Congenital hepatic fibrosis (by definition)
Typically presents in early adulthood, < 30 years (80%)	Typically revealed in childhood; familial form ++
Clinical features: recurrent cholangitis (64%)	Clinical features: recurrent cholangitis, portal hypertension
Renal abnormalities are rare	Renal abnormalities are common (85%): autosomal polycystic kidney disease

CONCLUSION

Caroli's disease is a rare and challenging condition to diagnose. It most often presents with recurrent episodes of cholangitis. The rarity of the disease and the lack of specific clinical signs frequently lead to delayed diagnosis. Our case illustrates the disease in its typical form.

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