

CAROLI DISEASE: CLINICAL CASE PECULIARITY

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ABSTRACT

Objective: Caroli disease is a rare disorder characterized by the saccular segmental dilatations of the intrahepatic bile ducts communicating with the principal biliary tract, without concomitant histological anomalies in the liver parenchyma. The disease has few symptoms and sometimes, it can remain silent. The clinical expression occurs with the development of the complications related to the bile stasis. The sonography examinations consist of the dilatation of the bile ducts and of the characteristic protrusion, which join the duct walls to the intraluminal portal branches. If a diagnostic doubt lasts after the diagnostic sonography, the use of the hepatobiliary nuclear stratigraphy or of the X-ray computed tomography or of the nuclear magnetic resonance can give important discriminatory information. The last diagnosis can be confirmed by the intraoperative cholangiogram procedure.

Methods: A woman was admitted to our Hospital because of left flank pain. She reported widespread abdominal pain in the left flank associated with nausea and vomiting in the ER. The diagnostic sonography showed that the volume of the liver was higher than normal with the obvious presence of multiple cystic formations of several dimension (3.5 cm max). The principal biliary duct, detected in its proximal portion, showed an increased caliber (1.4 cm). The intrahepatic biliary ducts were dilated. Both kidneys showed several cystic formations, which altered the dimensions and the ecostructure of both kidneys *in toto* in a polycystic kidney disease. There is an additional biliary lithiasis of about 6 mm of the left ureter's intramural tract. Therefore, the patient was hospitalized with diagnosis of left-sided colitis on a polycystic kidney with a renal failure and dilatation of the bile ducts TBD and with abnormal blood texts. The woman was submitted to the MRI of the abdominal cholangiography. **Results:** During the observation we have witnessed the gradual improvement of the symptomatology, so the patient was discharged with a diagnosis of renal colic, lithiasis of the urinary tract in a polycystic kidney, chronic renal failure and multicystic dilatation of the intrahepatic bile ducts (Caroli disease) with medical therapy. **Conclusions:** Caroli disease, that is part of this group of disorders, has the reputation of being a rare illness, above all in the pure form. This is a real rarity, but it is overestimated by the benign clinical course of the first 5-20 years of life, sometimes longer and, even if in few cases, asymptomatic, insomuch as it is often fortuitously suspected or diagnosed after a liver biopsy or after an ultrasonography survey or a tomography investigation due to another cause. The diagnostic approach of the Caroli disease is based on various anamnestic, clinical, biological and radiological data, which together allow making the correct diagnosis. Therapeutic behaviour recommends in the first instance some cycles of antibiotic therapy to hinder the rare cholangitis episodes and, if it is possible, extracorporeal lithotripsy, and then to pass to the more radical one of the hepatic section when the symptomatology becomes ingravescant and only one lobe is interested

KEYWORDS: Caroli disease, Multicystic intrahepatic bile duct dilatation, Cholangiography.

CASE REPORT

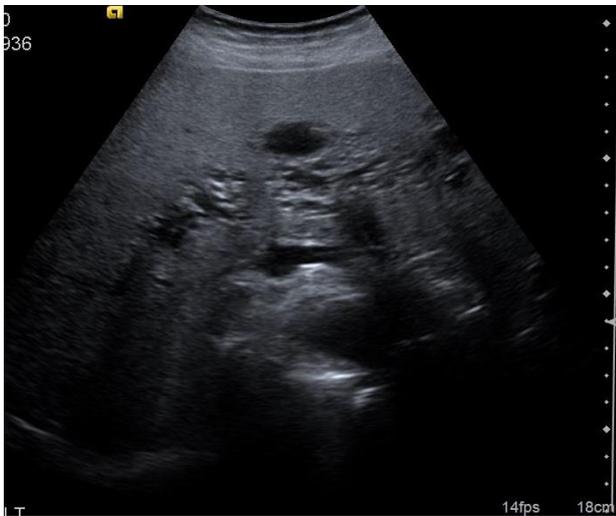
A woman was admitted to our Hospital because of left flank pain. She reported widespread abdominal pain in the left flank associated with nausea and vomiting in the ER. The abdomen was slightly distended, the tympanism was modestly accentuated at the abdominal percussion, and the liver was at the upper border, at the IV intercostal space, on the right midclavicular line with a palpable and sharp lower edge at the right costal arch.

The diagnostic sonography showed that the volume of the liver was higher than normal with the obvious presence of multiple cystic formations of several dimension (3.5 cm max). The principal biliary duct, detected in its proximal portion, showed an increased caliber (1.4 cm). The intrahepatic biliary ducts were dilated. Both kidneys showed several cystic formations, which altered the dimensions and the ecostructure of both kidneys *in toto* in a polycystic kidney disease. There is an additional biliary lithiasis of about 6 mm of the left

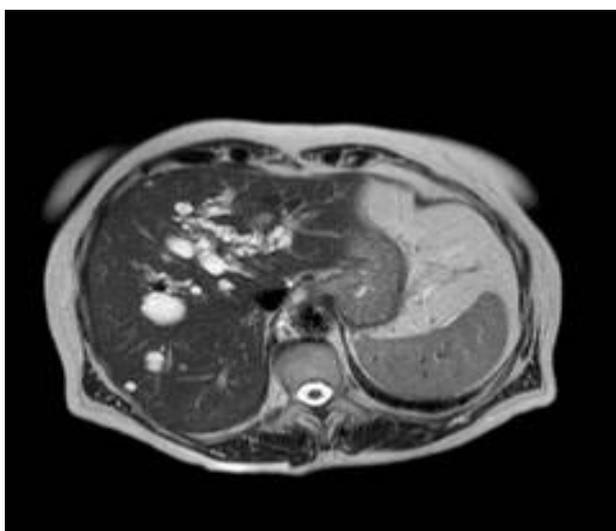
ureter's intramural tract. Therefore, the patient was hospitalized with diagnosis of left-sided colitis on a polycystic kidney with a renal failure and dilatation of the bile ducts TBD.

Abnormal blood texts: creatinine 2.9 mg/dl, Procalcitonin 0.13 ng/ml, NT PRO-BNP 2086 pg/ml, GGT 51 UI/L, lipase 71 UI/L, Alkaline Phosphatase 140 UI/L, LDL Cholesterol Calc 118 g/dl, HDL Cholesterol 41 mg/dl. The woman was submitted to the MRI of the abdominal cholangiography with the following medical report: a multicystic dilatation of the intrahepatic bile ducts is provided, but without endoluminal filling defects due to lithiasis. The principal biliary duct is normal and does not reveal endoluminal filling defects, too. The medical case suggests the Caroli disease. It is associated to a bilateral polycystic kidney disease. The gallbladder is well distended, with regular walls, and alithiasic. The spleen and pancreas are normal, and the last one does not show Wirsung dilatation.

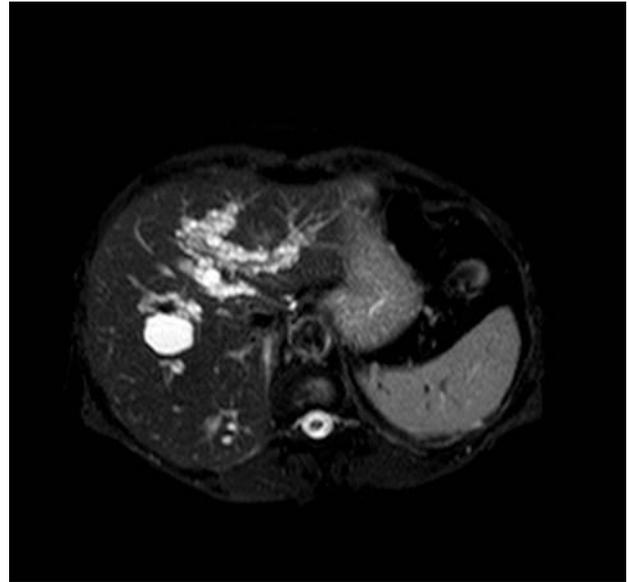
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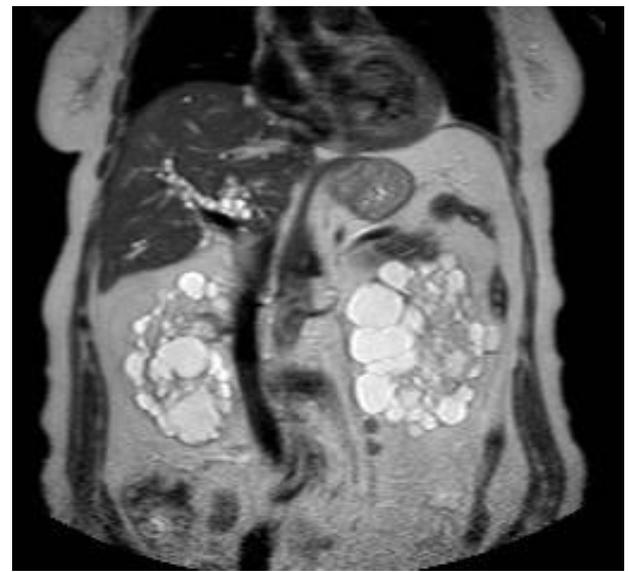
1(a)



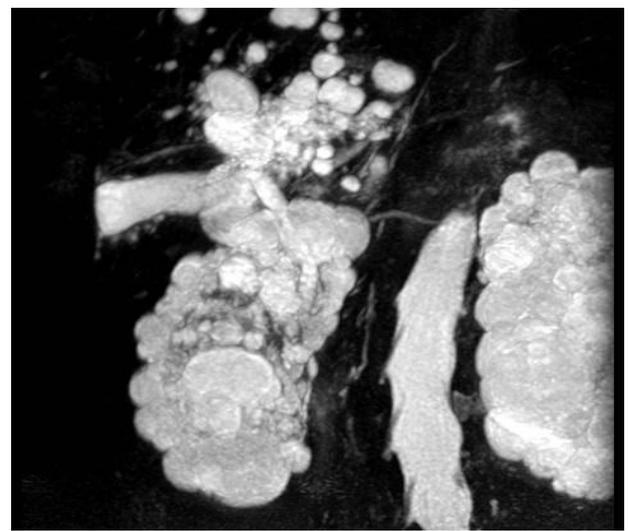
1(b)



1(c)



1(d)



1(e)

Fig.1 (a) Show a sonography examinations of the liver with multiple cystic formations with initial dilatation of the bile ducts at the confluence without demonstration of intrahepatic stone; The MR performed at the same patient with T2 sequences on axial and coronal plane with and without fat suppression (b,c,d) confirmed a multicystic dilatation of the intrahepatic bile ducts without endoluminal filling defects due to lithiasis. The principal biliary duct is normal and does not reveal endoluminal filling defects, too. It is associated a bilateral polycystic kidney malformation that you can also see with the cholangiography reformatted image (e)

During the observation we have witnessed the gradual improvement of the symptomatology, so the patient was discharged with a diagnosis of renal colic, lithiasis of the urinary tract in a polycystic kidney, chronic renal failure and multicystic dilatation of the intrahepatic bile ducts (Caroli disease) with the following therapy: omeprazole, Triatec^[1] 2.5 mg 1 pill, Congescor 1.25 mg 1 pill, Rasilez HCT 1 pill, Deursil^[2] 450 mg 1 pill, Moduretic 1 pill, Totalip 20 mg, hydration and gastroenterological and nephrology follow-ups.

REFERENCES

1. Trade name for Ramipril.
2. Trade name for the Ursodeoxycholic acid.