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Very Rare Association Between Caroli's Disease and Polycystic Kidney Disease

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Abstract

Caroli's disease is a rare disease. The diagnosis of Caroli's disease is often late responsible for several complications. The presence of asymmetric dilation of the bile ducts and evidence of Dot Sign on ultrasound should suggest the diagnosis. Biliary MRI is essential for determining the topography of dilation, communication with the bile ducts and intrahepatic lithiasis. Therapeutic management is problematic, ranging from clinical monitoring to liver transplantation. The association with polycystic kidney disease has only rarely been described in the literature. In this regard, we present the case of a 58-year-old woman, presenting with chronic constipation revealing Caroli's disease associated with polycystic kidney disease, having consulted with the gastroenterology department at the Arrazi hospital of the Mohammed VI University Hospital of Marrakech.

Keywords: Caroli disease; Polycystic kidney disease; Constipation

Introduction

Caroli's disease is defined by congenital cystic dilation of the intrahepatic bile ducts, it is a rare entity, whose clinical and radiological data have been reported in the literature, but on short series of patients. The diagnosis of this disease must be early in order to watch for the occurrence of complications. Polycystic kidney disease is a common inherited disease characterized by the development of renal cysts associated with extra-renal manifestations. The combination of these two pathologies is rare and has only been described in the literature in a few rare cases. Through this work, we propose to report a case of Caroli's disease associated with polycystic kidney disease discovered fortuitously, in a 58-year-old woman.

Observation

We present the observation of a 58-year-old patient, her personal history was limited to cholecystectomy for gallstones 32 years ago. The patient consulted for chronic constipation without other digestive or extra-digestive manifestations, evolving for 6 months in a context of preservation of the general condition. The somatic examine ion of the abdomen was unremarkable. The biological workup was without hepatic abnormality, renal function was impaired with a creatinine level of 17.32 mg/L and urea at 0.43 g/L with a glomerular filtration rate of 39 ml/min/1.73m^2. A colon CT scan was performed on the patient as part of the etiological assessment of her constipation, objectifying kidneys increased in size, measuring 15.6* 8.5 cm on the right and 15.3* 8.2 cm on the left, with lobulated contours, seat of multiple shapes and of variable size, some of which are the site of fine and regular parietal calcifications, associated with a marked dilation of the intra and extra hepatic bile ducts at the level of the common hepatic duct measuring 27.6 mm, with no obvious visible obstacle (Figure 1).

The imaging was supplemented by a biliary MRI which revealed a liver increased in size, measuring 18 cm, with lobulated contours in places, site of multiple cystic formations, scattered throughout the hepatic parenchyma, sub capsular and intra parenchymal, of variable size and shape, in T1 hypo-signal, in T2 hyper-signal and diffusion, not enhanced after injection of the contrast product, without detectable septum or tissue bud. The largest of these cysts sits at the level of segment IV and measures 21 mm in diameter. Some of these cysts seem to communicate with segmental dilated bile ducts with individualization in places of portal veins

arranged in a bridge or enhanced within the cystic formations (Dot Sign). A main bile duct of normal caliber measuring 9 mm in diameter. A normal-gauge, permeable trunk with no signs of portal hypertension. Kidneys increased in size measuring 16×9 cm on the right and 13×9 cm in lobulated contours on the left, sites of multiple cystic formations scattered throughout the renal parenchyma which is laminated into a T1 and T2 hypo-signal and enhanced over time arterial and portal (Figure 2).

Renal cystic forms have a T1 hypo-signal, T2 hyper-signal not enhanced after injection of the contrast product, without a septum or detectable tissue bud. These arguments led to the conclusion that Caroli's disease was associated with polycystic kidney disease. Management of the disease consisted of prevention of lithiasis by abundant hydration > 2L/day with blood pressure monitoring by six-monthly monitoring of blood pressure, renal function and liver function tests (Figure 3).

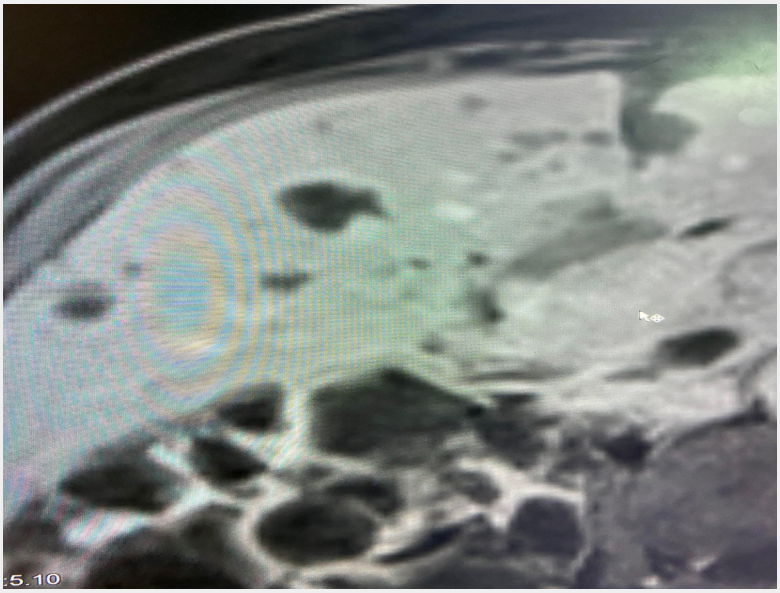

Figure 1: Caroli's disease.

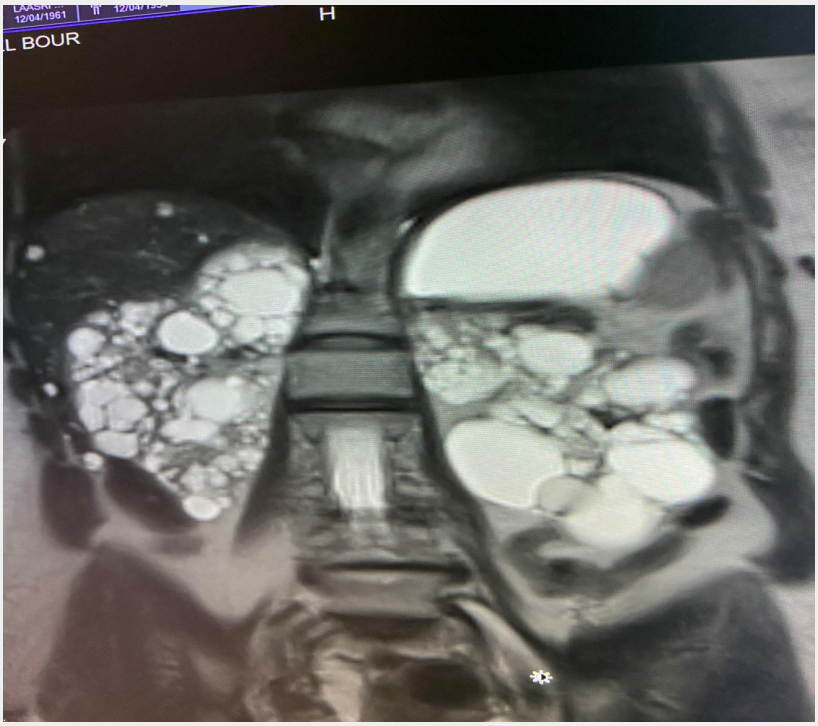

Figure 2: Congenital cystic dilation of the intrahepatic bile ducts.

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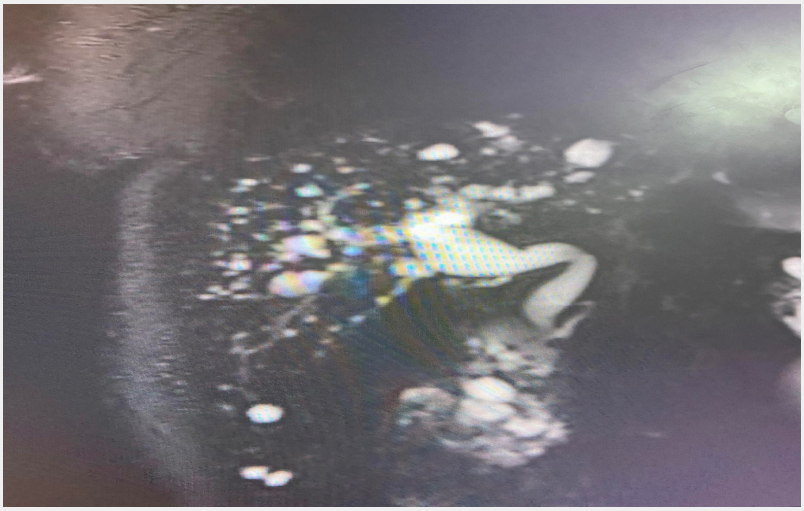

Figure 3: Caroli's disease associated with polycystic kidney disease.

Discussion

Caroli's disease is defined by non-obstructive segmental dilation of the intrahepatic bile ducts. This dilation can be diffuse or localized due to an anomaly of remodeling of the ductal plaque during embryogenesis, it can exist in two forms: the pure form characterized by attacks of cholangitis and the mixed form which associates d 'other hepato-biliary lesions in Caroli's disease. We speak of Caroli syndrome when the diffuse form is associated with congenital hepatic fibrosis complicated by cirrhosis or portal hypertension. This syndrome may be associated with polycystic kidney disease and Cacchi-Ricci disease [1,2]. This congenital anomaly affects both sexes equally and is generally not discovered until adulthood and which testifies to a clinical latency explained by the absence of specific signs of the disease, thus making the discovery of the disease. disease at the stage of complications such as intrahepatic lithiasis or acute and repetitive cholangitis often reported in the literature, secondary to retention of infected bile in ductal ectasia [2,3]. This retention of bile can turn into an abscess [4-9], or even degeneration into cholangiocarcinoma which remains rare but the risk is 100 times higher than that of the general population and which can reveal the disease [10]. Liver function tests show in most cases an increase in the level of direct bilirubin and an increase in alkaline phosphatase, while the level of transaminases can be preserved at the beginning. The diagnosis of Caroli's disease can be made based on data from a hepatobiliary ultrasound which shows cystic saccular or spindle-shaped dilation without any detectable underlying obstruction. The communication between the intrahepatic cystic dilations and the rest of the biliary tree is easily demonstrated on ultrasound. It also makes it possible to search for intrahepatic lithiasis with high sensitivity, the inconstant «Dot Sign» which is the most characteristic, described as a small point in the center of the dilation which corresponds to a vessel [11]. When the diagnosis is strongly suspected on ultrasound, second line CT also highlights: cystic dilation of the central periportal VBIHs as well as cystic dilations of the common bile duct, intrahepatic lithiasis: less sensitive than ultrasound. Bili-MRI is a non-invasive exam and currently the gold standard exam supplanting ERCP. It has the advantage of avoiding direct opacifications of the bile ducts, which are often serious sources of iatrogenic cholangitis, and of not requiring the use of an intravenous contrast agent. Biliary MRI visualizes the entire biliary tree and gives a better biliary mapping, highlights cystic dilations of the proximal VBIHs as well as cyst communications with the bile ducts, Dot Sign, and intrahepatic stones [12]. The pathological study allows confirmation of the diagnosis, the search for an associated lesion, such as congenital hepatic fibrosis, and the detection of neoplasia.

The treatment of Caroli's disease is problematic. However, in localized forms the therapeutic option is represented by liver resection, which is only possible in 3% of patients [5,7,13-16]. Hepatectomy allows the eradication of the lithogenic focus which represents a source of septic complications and also helps protect the patient from malignant degeneration. The therapeutic problems are mainly related to the diffuse forms. The most performed procedure is internal bypass, however endoscopic treatment should be discussed before its indication [17]. It is an endoscopic sphincterotomy with sweeping of the main bile duct. This treatment requires iterative endoscopy, stent replacement, and the prescription of long-term deoxycholic or urso-deoxycholic acid [5]. Liver transplantation is the treatment of choice in diffuse forms [18].

Polycystic kidney disease is a genetic disease characterized by the development of cysts in the kidneys, which gradually destroys the renal parenchyma, leading to impaired kidney function and impaired glomerular filtration rate. Other organs such as the brain, liver, pancreas and arteries can also be affected. Polycystic kidney disease remains silent for many years. The development of intra-

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renal cysts will be responsible for renal complications such as, intracystic hemorrhages, lower back pain correlated with cystic mass, urinary tract infections, high blood pressure and lithiasis. The management is based on the prevention of complications of the disease and the protection of renal function by symptomatic treatment and treatment of renal and extra-renal complications as well as the reduction of cardiovascular risks by controlling blood pressure and menstruation. hygieno-dietetics, etc. [19]. In end stage renal failure, the treatment offered is hemodialysis or kidney transplantation. No treatment to slow the progression of the disease was available until recently.

Conclusion

Caroli's disease is a rare entity, often discovered late and responsible for several complications. The absence of clinical manifestations and association with gallstones can misdiagnose. Polycystic kidney disease is a genetic disease, defined by the development of cysts in the kidney, which can be responsible for kidney failure.

References

1. Brancatelli G, Federle MP, Vilgrain V, Vullierme MP, Marin D, et al. (2005) Fibropolycystic liver disease: CT and MR imaging findings. Radiographics 25(3): 659-670.
2. Mabrut JY, Partensky C, Jaeck D, Oussoultzoglou E, Baulieux J, et al. (2007) Congenital intrahepatic bile duct dilatation is a potentially curable disease: long-term results of a multi-institutional study. Ann Surg 246(2): 236-245.
3. Guntz Ph, Coppo B, Lorimier G, Cronier P, Guntz M (2005) La maladie de Caroli unilobaire. Aspects anatomopathologiques. Démarche diagnostique et thérapeutique. A propos de 3 observations personnelles.
4. Versaci A, Macrí A, Scuderi G, Certo A, Bottari A, et al. (2005) Caroli's disease: literature review and characteristics of a clinical case. G Chir 26(5): 195-200.
5. Mercadier M, Chigot JP, Clot JP, Langlois P, Lansiaux P (1984) Caroli's disease. World J Surg 8(1): 22-29.
6. J Silvera, MP Vullierme, S Precetti, F Degos, D Valla, et al. (2009) Maladie et syndrome de caroli : etude muticentrique clinique et de l'imagerie de 39 patients. Journal de Radiologie 90(10): 1426.
7. Suchy FJ. (2006) Caroli's disease. Up to date Version 14.1.
8. Nagasue N. (1984) Successful treatment of caroli's disease by hepatic resection. Report of six cases. Ann Surg 200(6): 718-723.
9. Bockhorn M, Malagó M, Lang H, Nadalin S, Paul A, et al. (2006) The role of surgery in Caroli's disease. J Am Coll Surg 202(6): 928-932.
10. Gillet M, Favre S, Fontolliet C, Halkic N, Mantion G, et al. (1999) Monolobar Caroli's disease. Apropos of 12 cases. Chirurgie 124(1): 13-18.
11. Benhamou JP, Erlinger S. (1987) Kystes non parasitaires du foie, fibrose hépatique congénitale. Syndrome de Caroli. Dans Maladies du foie et des voies biliaires. Flammarion Médécine-sciences 3ème edition 85-88.
12. C Chammakhi jemli, N Achour, S Ben dahia (2006) Apport de l'imagerie dans le diagnostic de la maladie de Caroli: a propos de 20 cases.
13. Kasper HU, Stippel DL, Töx U, Drebber U, Dienes HP (2006) Primary cholangiocarcinoma in a case of caroli's disease: case report and literature review. Pathologe 27: 300-304.
14. Schrumpf E, Bergan A, Blomhoff JP, Hoel PS, Kolmannskog F (1981) Partial hepatectomy in Caroli's disease. Scand J Gastroenterol 16(5): 581-585.
15. Ramond MJ, Huguet C, Danan G, Rueff B, Benhamou JP. (1984) Partial hepatectomy in the treatment of Caroli's disease: Report of a case and review of the literature. Dig Dis Sci 29(4): 367-370.
16. Watts DR, Lorenzo GA, Beal JM. (1974) Congenital dilatation of intrahepatic biliary ducts. Arch Surg 108(4): 592-598.
17. Marin C, Robles R, Pastor P, Parilla P (2008) Liver resection in the treatment of intrahepatic lithiasis. Immediate and long term results in a single-center series. Rev Esp Enferm Dig 100(4): 225-229.
18. Habib S, Shakil O, Couto OF, Demetris AJ, Fung JJ, et al. (2006) Caroli's disease and orthotopic liver transplantation. Liver Transpl 12(3): 416-421.
19. Jacques Buxeraud, Sébastien Faure, Nicolas Picard (2018) Polykystose rénale, pour une bonne dispensation du tolvaptan. Actualités Pharmaceutiques 57(578): 49-53.

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