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Visceral Radiology

Caroli Disease: A Case Report with Review of the Radiological Literature

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Abstract **Case Report**

Caroli disease may manifest as right upper quadrant pain, recurrent biliary lithiasis and cholangitis with fever and jaundice. Caroli syndrome presents the above symptoms plus signs of portal hypertension, including hematemesis and melena secondary to hemorrhagic varices.

Keywords: Congenital Hepatic Fibrosis, Portal Hypertension, Esophageal Varices, Chronic Liver Disease.

INTRODUCTION

Fibropolycystic liver disease, non-hereditary congenital disease, rare < 1/ 1,000,000 in the general population. Embryological abnormality in ductal plate development Non-obstructive dilatation of intrahepatic bile ducts Diffuse (75%) or localized (left lobe), bile duct cyst sometimes associated May be associated with congenital hepatic fibrosis: Caroli syndrome.

Pathologically, Caroli disease and Caroli syndrome belong to the spectrum of fibropolycystic liver disease, which results from in utero malformation of the ductal plate. There is a strong association with fibrocystic abnormalities of the kidneys, which share the same genetic defect (PKHD1 gene, chromosomal region 6p21).

We report the case of a patient with congenital liver fibrosis at cirrhosis stage complicated by portal hypertension with VOs that were ligated in 2016.

The last check-up showed persistent grade II VOs but no red signs.

OBSERVATION

The patient was 20 years old and was being treated for congenital hepatic fibrosis characterized by a digestive hemorrhage in 2016 on a rupture of esophageal varices. At the time, he had undergone a liver biopsy that was compatible with congenital hepatic fibrosis.

In terms of GI bleeding, the VOs were collapsed at the September 2018 fibroscopy and he is on standard Beta Blocker prophylaxis.

He has no hepatic encephalopathy, no ascites, no angiocholitis.

On ultrasonography, the portal trunk was thrombosed and arterialized at the level of the hepatic hilum, which is why he was put on Lovenox in preventive doses.

On MRI, he has a chronic hepatopathic liver with signs of portal hypertension and no visible focal lesions. He has diffuse, relatively smooth dilatation of the bile ducts with no evidence of stenosis. Tapered appearance of the proximal segment of the main bile duct, with fine, regular caliber in its distal two-thirds. Retroport adenopathy 12 mm in diameter.

The intrahepatic bile ducts are diffusely dilated without visible obstruction, with an appearance suggestive of dilatation secondary to portal cavernoma without obvious visualization of the portal cavernoma. The right and left portal branches and the portal trunk are small, and the splenic vein is threadlike with small shunts.

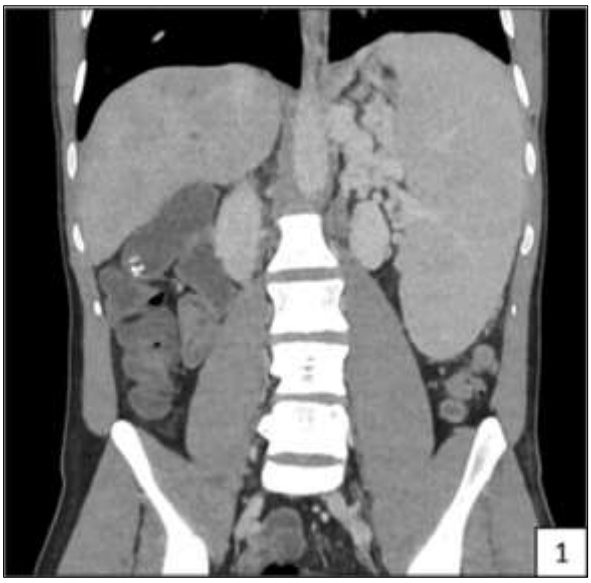

Figure 1: coronal abdominal-pelvis CT at portal time: Homogenous splenomegaly to 20 cm

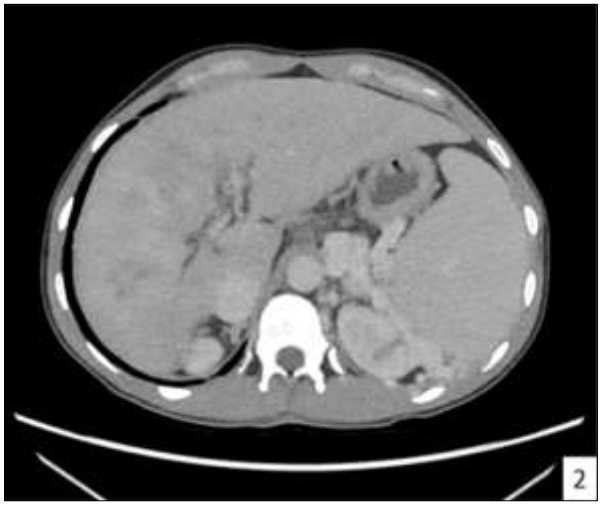

Figure 2: axial abdominal-pelvis CT at portal time: Multiple porto-systemic shunts, notably spleno-renal. Peri-gastric, peri-splenic, peri-esophageal and pelvic varices

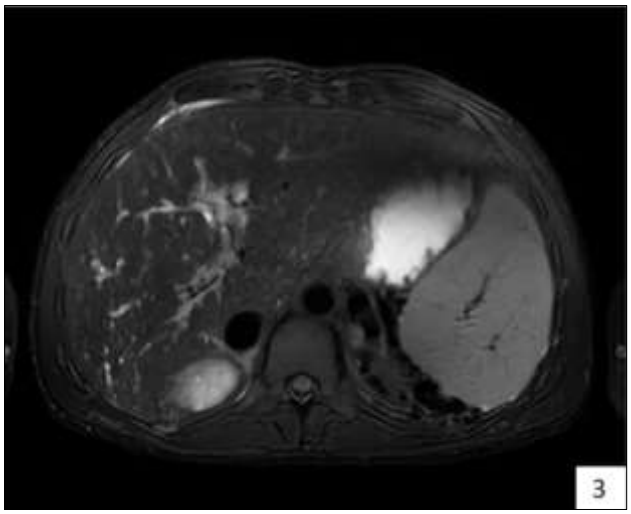

Figure 3: axial T2 SSFSE: Diffuse fusiform and cystic dilatation of the left and right intrahepatic bile ducts

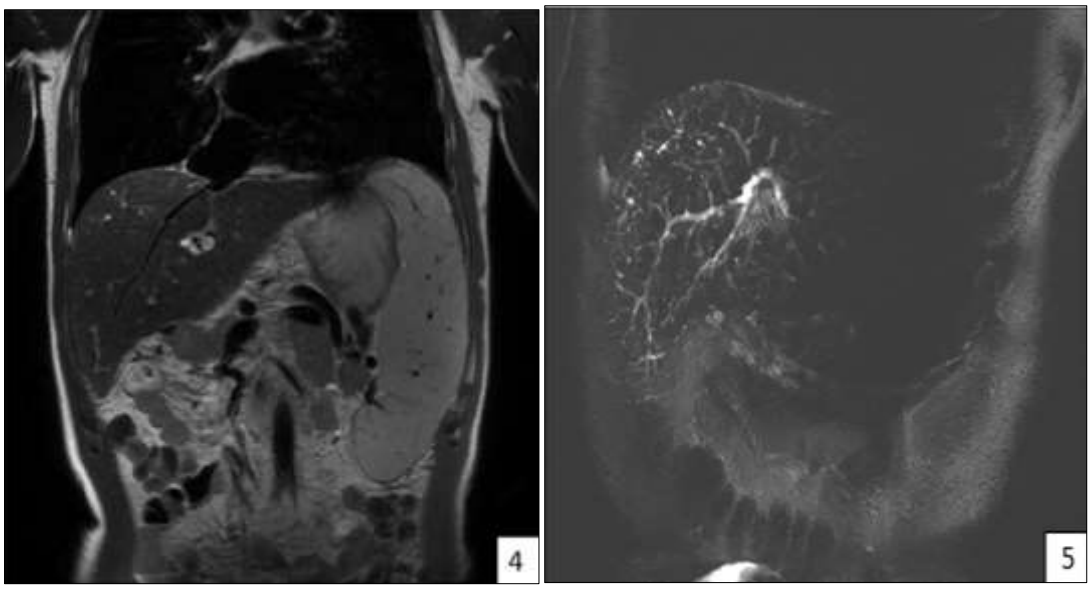

Figure 4-5: coronal T2 SSFSE- coronal obliques BILI: The distal third of the main bile duct is measured at 4.2 mm, with no internal lithiasis

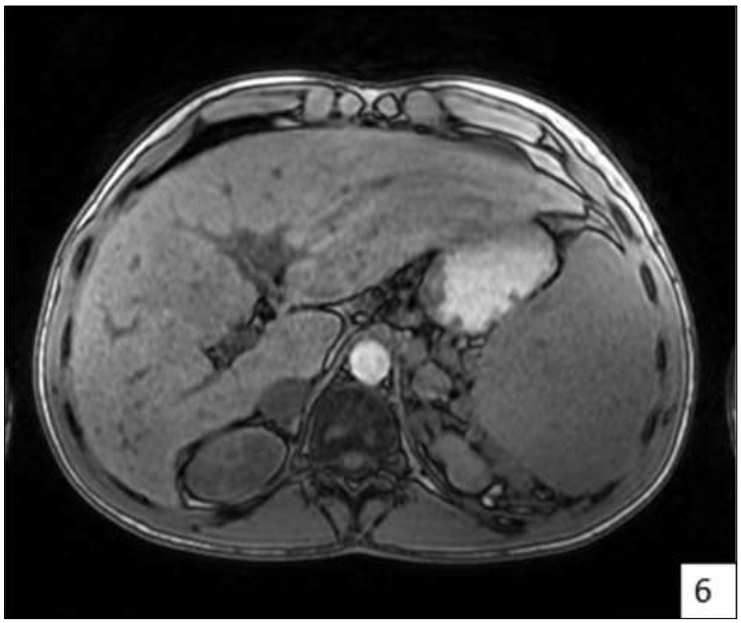

Figure 6: Axial T2 Out phase: Fusiform, cystic dilatation of the periphery of the intrahepatic bile ducts, communicating with the biliary tree

DISCUSSION

The patient is now 27 years old, scheduled for a liver transplant. His latest examinations show:

Liver of normal size, measured at 15 cm long axis on the mid-clavicular line in the coronal, with irregular contours and hepatic dysmorphia (hypertrophy of the left lobe and atrophy of the right liver).

No steatotic overload.
Complete regression of hepatic segment VI subcapsular abscess.

Hepatic dome perfusion disorder. Diffuse fusiform and cystic dilatation of the left and right intrahepatic bile ducts. The distal third of the main bile duct is measured at 5 mm, with no lithiasis.

Normal-sized gallbladder with multilithiasis-enhanced walls. Intrahepatic lithiasis in the posterior sector. Splenomegaly measured at 21 cm in height, associated with numerous porto-systemic shunts.

No suspicious lymph nodes in the volume explored.
No significant abnormality of the pancreas, adrenals or proximal urinary tract.

Presence of bilateral renal cysts, predominantly on the right, related to known pathology. Absence of peritoneal effusion.

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