



Laparoscopic Liver Resection in a Case of Asymptomatic Elderly Patient with Caroli Syndrome

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Introduction

Caroli syndrome (CS) is a rare congenital disorder first described in 1958 by Jacques Caroli [1] and it is characterized by dilatation of the intrahepatic biliary tree with malformation of the small bile ducts and hepatic fibrosis. There are two patterns of Caroli disease: focal or simple Caroli disease consists of abnormally dilated bile ducts affecting an isolated portion of liver. The second form is more diffuse, and when associated with congenital hepatic fibrosis, is referred to as Caroli syndrome [2]. It is generally inherited as an autosomal recessive trait and could be associated with other diseases inherited in this manner, e.g., autosomal polycystic kidney disease (ARPKD). It usually affects the left lobe of the liver even though any part could be affected.

The exact prevalence or incidence of the disease or the syndrome is not known. However, more than 200 cases of caroli disease have been reported in the medical literature [3]. The prevalence of caroli disease has been estimated to be around 1/1,000,000, with a slightly female predisposition [4].

In this modern era of advanced diagnostics, it is usually diagnosed early, often in childhood, so that the appropriate intervention could be carried out [5, 6]. Associated clinical conditions which could prompt the clinician to suspect the diagnosis for the syndrome are cholangitis, cholelithiasis, cholecystitis, and their associated clinical signs and symptoms as well as the presence of an autosomal recessive disease.

Patients with Caroli syndrome could be asymptomatic, and a high index of suspicion is needed to diagnose it [6]. The work has been reported in line with the SCARE criteria [7].

Case Presentation

We present a case of a 70-year-old female with Caroli syndrome without any clinical symptoms. The condition was coincidentally diagnosed while undergoing a radiological investigation for a different purpose. As a known chronic obstructive disease (COPD) patient, she presented at his family physician with persistent difficulty in breathing of 7-day duration. By further probing the clinical condition, and also to rule out pulmonary artery embolism, the family physician requested for computed tomography (C.T) examination of the thorax and Abdomen. The examination was done on the 16.01.2013. It showed dilatation of the intrahepatic biliary tract with minimal hepatic fibrosis. (Fig. 1). Compared to a computed tomography of the abdomen taken 12 years previously (Fig. 2), it was noted that the intrahepatic biliary dilatation existed, but per the clinical condition during that time, Caroli syndrome was not suspected. It also showed tumors of the right and left upper and lower lobes of the lungs. The lesion of the left upper lobe was found to be new and was suspected to be a bronchial carcinoma. This therefore warranted for further investigations.

A contrast-enhanced abdominal sonography performed on 28.01.2013 showed dilatation of the biliary tract of the left liver segment II and III as well as a hepatic parenchymal cyst. A follow-up abdominal MRI taken on January 31, 2013, confirmed the dilation of the biliary tract (Fig. 3), the hepatic cysts as well as a right renal tumor of about 16 × 12 mm with concomitant cyst and a small left ovarian cyst.

Through video-assisted thoracoscopic surgery, a wedge resection of the left upper lobe of the lungs was carried out about a month later (on February 21, 2013). The histological examination of the tissue showed moderate pulmonary emphysema,

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Fig. 1 Dilatation of the intrahepatic biliary tract with minimal hepatic fibrosis

pleural fibrosis with granuloma as well as bronchial anthracofibrosis (BAF). There was no evidence of cancer cells.

A retroperitoneoscopic renal resection was carried out on August 22, 2013, to remove the right renal tumor. The histological examination showed a renal angiomyolipoma without any evidence of malignancy.

The patient was subsequently referred to our center for further evaluation and the management of the liver lesions. A critical evaluation of radiological examinations showed

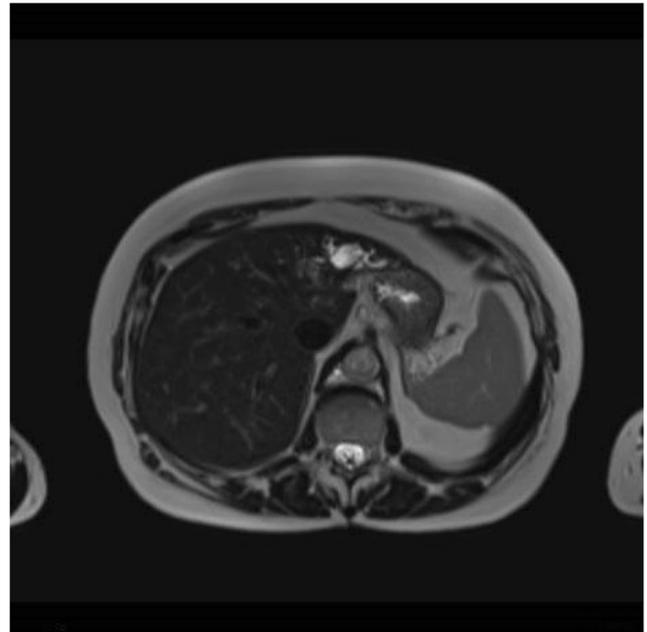


Fig. 3 Dilatation of the biliary tract

caroli syndrome. The patient was followed with serial MRI-examination over a period of 4 years. The follow-up examinations showed no significant radiological changes of the dilatations of the biliary tract (Fig. 4), and she remained throughout the course asymptomatic.

On physical examination, the patient was found to be in good general health. There were no signs of chronic liver disease. On palpation, the abdomen was soft without tenderness and organomegaly. The abdominal sounds were normal.

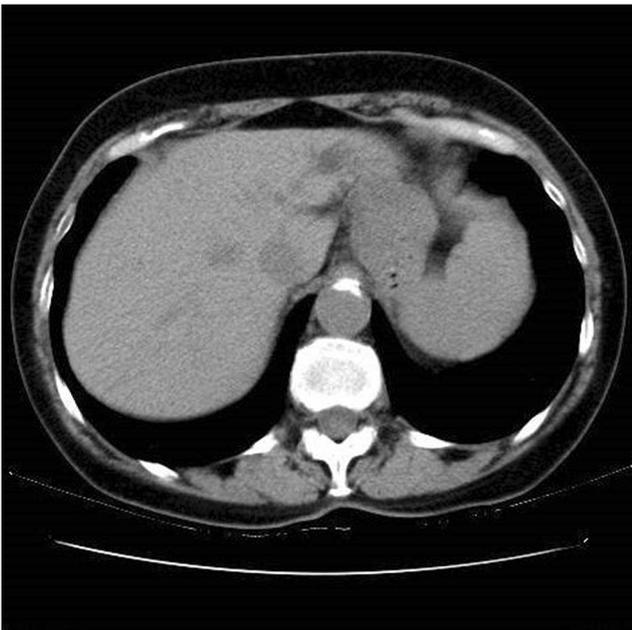


Fig. 2 Computed tomography of the abdomen taken 12 years previously

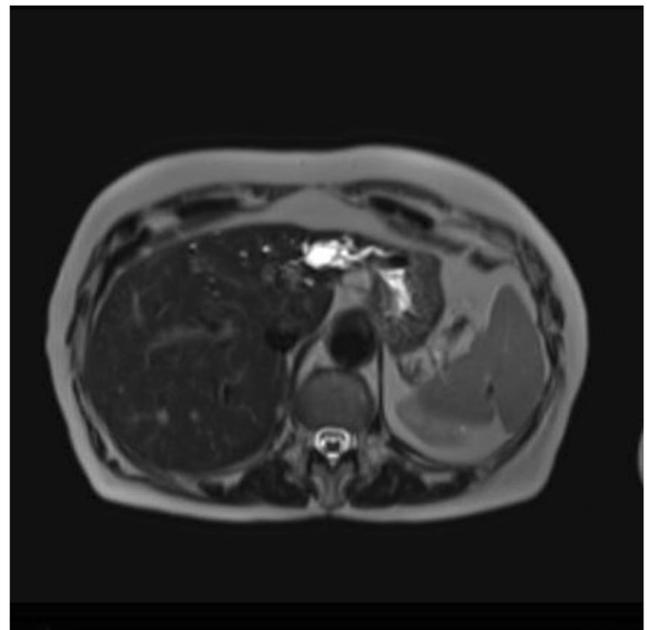


Fig. 4 No significant radiological changes of the dilatations of the biliary tract

Laboratory Results Were As Follows

Initial Laboratory Constellation

GOT(AST) 17 U/l, GPT(ALT) 28 U/l, LDH 28 U/l, Gamma-GT 41 U/l, alkaline phosphatase 46 U/l, total bilirubin 0,7 mg/dl, direct bilirubin 0,2 mg/dl, creatinine 0,8 mg/dl, INR 0,87, CA 19–9 10,3 U/ml, amylase 75 U/l, lipase 37 U/l, hemoglobin 15,3 g/dl, platelet 239 T/ μ l, WBC 16,1 T/ μ l

Laboratory Results Just Before the Surgical Operation

GOT 45 U/l, GPT 27 U/l, LDH 330 U/l, Gamma-GT 17 U/l, alkaline phosphatase 36 U/l, total bilirubin 0,6 mg/dl, direct bilirubin 0,2 mg/dl, CA 19–9 5,5 U/ml, AFP 3,2 ng/ml Amylase 62 U/l, lipase 35 U/l, hemoglobin 14,1 g/dl, platelet 256 T/ μ l, WBC 10,8 T/ μ l.

Outcome and Follow-up

Laparoscopic resection of the affected liver (segment II/III) was performed on the January 13, 2017, because of the increased risk of cholangiocellular carcinoma of patients with caroli syndrome. The histological assessment confirmed Caroli syndrome without any evidence of malignancy. The postoperative phase was uneventful and the patient was discharged on the fifth postoperative day.

Discussion

Caroli syndrome is a rare congenital disorder generally inherited as an autosomal recessive trait, and could be associated with other diseases inherited in this manner. Even though the modern era of radiological investigations makes its diagnosis somewhat early and easy, a high index of suspicion is needed in making the diagnosis as the disease may not have any distinguishing symptoms and signs and therefore patients may be asymptomatic for a long time [5]. It should be born in mind as a differential diagnosis for patients presenting with diseases which are inherited in likewise manner as well as cystic changes of other organs such as the ovaries, kidney and the lungs especially when the dilatation of the intrahepatic tract is present as was seen in our patient.

Clinician are usually prompted to further investigate for CS due to the presence of, e.g., cholangitis, hepatic fibrosis, cholecystolithiasis, choledocholithiasis, and their relevant clinical signs and symptoms [3]. However, our patient presented with dilatation of the biliary tract of the left liver segment II and III as well as a hepatic parenchymal cyst including dilation of the biliary tract, the hepatic cysts as well as a right renal tumor with concomitant cyst and a small left ovarian cyst which does not

suggest a typical CS. Caroli disease in combination with renal disease is a possibility even though it is rare as was seen in a study that showed an association with Caroli diseases and bilateral cystic renal dysplasia [8]. Our patient also presented earlier with chronic obstructive pulmonary disease (COPD) which although is not usually associated with CS was also documented in another case which was associated with emphysema [9].

The syndrome runs a variable course, but symptomatic patients need prompt intervention to curtail the unfavorable fatal outcomes. The time for implementation of therapy by asymptomatic patients is however unclear. In the case of our patient, a total of at least 16 years elapsed before a surgical intervention was carried out. The patient however remained throughout the course asymptomatic. Laparoscopic resection, which is a surgical intervention with reduced operative complications was carried out even in the absence of emergent clinical conditions because of the fatal complications such as increased risk of transformation into cholangiocellular carcinoma. Clinicians should therefore have a high index of suspicion for Caroli syndrome in patients who present with dilatation of the intrahepatic biliary tract with hepatic fibrosis even without symptoms of Caroli syndrome.

Conclusion

Caroli's syndrome association with cystic changes of other organs has not been established. The differential diagnosis of Caroli syndrome by patients with intrahepatic biliary tract dilatation even in absence of hepato-biliary symptoms should be born in mind, especially when cystic changes of other organs are present.

The syndrome may run a silent course, but the prognosis could be fatal including an increased risk of malignant transformation and liver failure. It carries about 15% malignant transformation. The time for institution of surgical intervention in asymptomatic patients is unclear. In the light of its high malignant transformation potential, a low-risk surgical procedure such as laparoscopic resection of the affected liver segment, if possible should be considered.

Author Contribution AOA conceptualized, attended the surgery, and wrote the paper. KMN conceptualized and wrote the paper. JT and GP attended the surgery. NL reviewed the radiological images and all authors read this paper.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Approval Institutional review board approval was exempt from our institution because all data were collected from clinical records and imaging systems for routine preoperative planning and follow-up.

Consent Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Guarantor Jochen Thies.

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