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Laparoscopic liver resection in Caroli disease: A single-centre case series

Simon Wabitsch¹, Wenzel Schoening¹, Julia-Sophia Bellingrath¹, Christian Brenzing¹, Alexander Arnold², Uli Fehrenbach³, Moritz Schmelzle¹, Johann Pratschke¹,¹ Department of Surgery, Charité – Universitätsmedizin Berlin, Campus Charité Mitte and Campus Virchow-Klinikum, Berlin, Germany² Department of Pathology, Charité – Universitätsmedizin Berlin, Campus Charité Mitte and Campus Virchow-Klinikum, Berlin, Germany³ Department of Radiology, Charité – Universitätsmedizin Berlin, Campus Charité Mitte and Campus Virchow-Klinikum, Berlin, Germany

Correspondence Address:

Dr. Wenzel Schoening

Department of Surgery, Charité – Universitätsmedizin Berlin, Campus Charité Mitte and Campus Virchow-Klinikum, Berlin Germany

Abstract

Background: Liver resection is the treatment of choice for patients with localised Caroli disease. While liver resection was traditionally performed as open procedure, this case series aims to evaluate the safety and efficacy of minimally invasive, laparoscopic liver surgery in these patients. **Methods:** A systematic review of electronic case files of patients seen between April 2015 and December 2017 at the Department of Surgery, Charité University Hospital Berlin, was conducted. Patients with Caroli disease in whom laparoscopic liver resection had been performed were identified and analysed in this single-centre case series. **Results:** Seven patients who underwent laparoscopic liver surgery for Caroli syndrome were identified and presented with a median age of 49 (range = 44–66) years, of which four (57%) were female. Preoperatively, six patients were classified as the American Society of Anaesthesiologists (ASA) 2 and one patient as ASA 3. Two operations were performed as single-incision laparoscopic surgery, whereas the others were done as multi-incision laparoscopic surgery. One patient required a conversion to an open procedure. The length of operation varied between patients, ranging from 128 to 758 min (median = 355). The length of stay in the intensive care unit ranged from 0 to 2 days. Two patients presented with post-operative complications (Clavien–Dindo Grade $\geq 3a$), whereas no patient died. In histopathological analysis, all patients demonstrated characteristic findings of Caroli disease and no cholangiocarcinoma was found. **Conclusion:** These results indicate that minimally invasive, laparoscopic liver surgery is a safe and efficacious treatment option for patients with Caroli disease who require liver resection.

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Full Text

Introduction

Caroli disease is a congenital disorder of the intrahepatic bile ducts, characterised by multifocal, segmental saccular or cystic dilatation and ectasia of the intrahepatic biliary tree.[1] In addition to biliary duct ectasia, the more common, autosomal recessive Caroli syndrome is characterised by congenital hepatic fibrosis (CHF). In 60% of cases,

it is associated with renal involvement, in particular autosomal recessive polycystic kidney disease, and on more rare occasions, with autosomal dominant polycystic kidney disease.[1],[2] Disease pattern can be either diffuse or limited to a liver segment or lobe. The complications of both Caroli disease and Caroli syndrome are caused by biliary stasis that ensues as a result of the intrahepatic dilation as well as the portal hypertension resulting from CHF.[3],[4] They include intrahepatic cholelithiasis, severe cholangitis, sepsis, hepatic abscess as well as an increased risk of developing intrahepatic cholangiocarcinoma (iCC).[5],[6] Surgical treatment has been shown to offer a definitive treatment option for patients with Caroli disease.[7] Surgical treatment options are liver resection for localised disease and liver transplantation for diffuse liver involvement.[8] Complete resection of localised Caroli disease can be curative and therefore has the potential to eliminate the risk of iCC.[9],[10] While liver resection was traditionally performed as open surgery, laparoscopic resection recently was found to be a safe and efficacious technique.[11],[12] Due to its minimally invasive nature, laparoscopic resection generally leads to a shorter and less painful recovery and mobilisation, leading to a decreased length of hospital stay.[13],[14] This study aims to investigate the use of minimally invasive, laparoscopic liver resection as a safe and efficacious tool in the surgical treatment of patients with Caroli disease. While single case reports for laparoscopic liver resection in these patients do already exist,[9],[15] showing the feasibility of laparoscopic resection, to the best of our knowledge, this present study is the largest case series to date.

Methods

Study design and collection of patient data

A systemic review of electronic case files of patients seen between April 2015 and December 2017 at the Department of Surgery, Charité University of Berlin, was conducted. Patients with Caroli disease, who underwent laparoscopic liver resection, were analysed, and the results summarised in this single-centre case series. Informed written consent was obtained from each patient before the surgery, and approval from the ethics committee (EA2/006/16) was sought before the usage and publication of these retrospectively analysed data.

Primary endpoints include safety and efficacy of laparoscopic liver resection measured by rate of surgical complications, length of stay in the intensive care unit (ICU) and duration of hospitalisation as well as histopathological proof of complete resection.

Pre-operative risk assessment was done with the American Society of Anaesthesiologists (ASA) classification.

Patient records contained the variables age, gender, reason for surgery, body mass index, duration of surgery, length of stay in the ICU, duration of hospital stay as well as complications classified according to Clavien–Dindo (CD).[16]

Surgical technique

Every patient was informed of potential risks and complications associated with laparoscopic liver resection and gave written consent. After anaesthesiologic preparation (intubation, central venous line, arterial line, etc.), the patient was placed in French position on the operation table and the operative field was washed and covered in a sterile manner. Detailed description of our single-incision laparoscopic surgical (SILS) approach is published by Struecker et al.[17] Concerning the other five patients of this series, following team time out, the first trocar was placed through a (supra-) umbilical minilaparotomy and pneumoperitoneum created with insufflation of CO₂. When a pressure of 14 mmHg was reached, two or three additional trocars (12.5 mm and 5 mm in diameter) were subsequently placed depending on the location of resection [Figure 1].{Figure 1}

After laparoscopic exploration of the abdominal cavity with macroscopic inspection of the liver, intraoperative ultrasound was performed to confirm the radiological diagnosis and extent of disease as well as the final planning of the resection. The respective liver lobe was mobilised with UltraCision™ (Harmonic ACE, Ethicon, USA). In patients without prior cholecystectomy, a gallbladder resection was performed. For right or left hemihepatectomy, the hepatic veins and their entry point into the inferior vena cava were visualised by dissection of the triangular ligament. In preparation for an extracorporeal Pringle manoeuvre, the hepatoduodenal ligament was isolated and threaded, and 250 mg methylprednisolone was given intravenously. The left and right hepatic arteries were visualised, and the artery belonging to the lobe to be resected was triple clipped and dissected. Then, the portal vein was visualised, and its respective portal vein branch was isolated and dissected with a 45 mm Endo-GIA stapler (Medtronic, MN, USA). The resection lines of segments or lobes to be removed were marked with a monopolar hook on the liver capsule guided by intraoperative ultrasound. Parenchymal liver dissection was performed with UltraCision™ for the more superficial tissue and with water jet (Erbe, Tuebingen, Germany) for the deeper parenchyma. Crossing structures were double clipped and dissected. Intraoperative bleeding was minimised by intermittently applied Pringle manoeuvre (each 15 min Pringle followed by 5 min reperfusion). Finally, the corresponding hepatic vein was dissected with a 45 mm Endo-GIA stapler (Medtronic, MN, USA) at the end of resection. Haemorrhage control was performed

with infrared and bipolar coagulation throughout parenchymal resection and afterwards with an intra-abdominal pressure reduced to 8 mmHg. The resected liver was retrieved either by a renewed incision of a pre-existing scar or a 'Pfannenstiel incision' [Figure 1].

After retrieval of the specimen, the abdomen was re-insufflated to rule out residual bleeding or biliary leakage; a silicon drain was placed, the intraperitoneal space was rinsed with isotonic fluid and the incisions were closed layer by layer. Finally, a sterile dressing was placed on the wound, and the patient was taken to the ICU or recovery room and later on regular surgical ward, depending on his condition.

Data analysis

Microsoft Excel (Microsoft, Redmond, WA) was used to collect the patient data and generate a patient database as well as for data analysis. Mean, median and standard deviation were calculated with Microsoft Excel. Continuous variables are displayed as medians with range.

Results

Patient characteristics

Of the seven patients, four (57%) were female and three (43%) were male. Patients' age ranged from 48 to 66 years (median = 49). The median body mass index (BMI) was 21.6 (range: 20.7–29.7). According to the WHO classification, five patients had a normal BMI (18–25) and one was overweight (25–29) but not obese [Table 1]. All patients had clinical and radiological signs of Caroli disease (representative imaging of patient 2 is demonstrated in [Figure 2]) before the surgery, and histopathological results of resected liver tissue confirmed the diagnosis in all cases.{Table 1}{Figure 2}

Caroli-related complications and co-morbidity

Six patients had complications related to their underlying disease, whereas one patient record lacked information about complications related to Caroli disease. The most common of these complications were intrahepatic gallstones, which were diagnosed in 86% of patients. Further complications were cholangitis, present in three patients, from which one patient had previously contracted systemic sepsis. Pancreatic and liver cysts, a possible extrahepatic manifestation of Caroli disease, were present in one patient. Four patients had concurrent disease of the extrahepatic bile ducts in the form of cholecystolithiasis and cholecystitis.

Apart from these complications related directly to Caroli disease, two patients exhibited typical metabolic disorders: primary hypertension and type II diabetes. Two further patients had hypothyroidism, whereas one patient had paroxysmal atrial fibrillation. Conn syndrome due to a cortical adrenal adenoma was present in one patient, and adrenal resection was performed concurrently. This patient also had the highest burden of co-morbidities which included primary hypertension, chronic obstructive pulmonary disease, chronic gastritis, type II diabetes mellitus, arthritis and an increased activity of factor VIII which had led to a deep-vein thrombosis and pulmonary embolism in the past.

Previous surgeries

Three of the patients had no previous surgeries and three had a history of abdominal surgery. Of the three patients with previous abdominal surgery, one underwent a cholecystectomy, the second underwent a cholecystectomy as well as an appendectomy and the third underwent surgical correction of a rectal prolapse as well as cataract operation on both eyes and additionally a stripping of lower-limb varices.

Extent of Caroli disease

All seven patients presented with unilobular disease. Five patients showed exclusively left liver involvement, whereas two patients had exclusive disease of the right liver lobe. The most common liver segments affected by Caroli disease were segments II and III, with five of seven patients showing the involvement of these segments. Of these five patients, two had additional involvement of segment IVb. The remaining two patients had liver lesions confined to segments VI and VII [Table 2].{Table 2}

Pre-operative risk assessment

Six patients were classified as ASA 2 and one patient as ASA 3. General laboratory values of AST, ALT, bilirubin, albumin, platelets and INR were normal. The median MELD score was 7 (range: 5–9).

Operation

Two patients presenting with bile duct dilations exclusively in segments II and III underwent a left lateral bisegmentectomy using a SILS approach. The third patient with lesions in the left lateral lobe was resected using a conventional multi-incision laparoscopic surgery (MILS). A left hemihepatectomy was performed on the patients with lesions in segments II, III and IV. Two patients presented with Caroli typical alterations in segments VI and VII. One of these patients was resected with a posterolateral hepatectomy, whereas the other patient underwent a complete right hemihepatectomy. In this patient, one of the lesions was adherent to the right hepatic vein, making a right hemihepatectomy the only feasible approach to safely remove the lesion.

The length of operation varied widely between patients, ranging from 128 min to 758 min (median = 355 min). The considerably longer operative time in one of our patients (758 min) can be explained by a 'non-emergency' conversion from laparoscopic to open left hemihepatectomy. This patient's disease was particularly severe and extensive both clinically and histopathologically with involvement of segments II, III and IVb. The conversion to open surgery was necessary because, intraoperatively, a massive dilation of the left hepatic bile duct extending up to the main bile duct bifurcation as well as a biliary variant with posterolateral duct draining through the left system was found. Thus, biliary reconstruction had to be performed with two bilio-jejunal anastomoses and insertion of a T-tube and a Neuhaus drain.

As mentioned previously, two operations were performed with a SILS port, whereas the others were performed by MILS. The number of trocars used in each individual operation ranged from 3 to 5 trocars. Sixty-five per cent were 12 mm trocars and 35% 5 mm trocars. Post-operative cosmetic results of patients that received MILS and SILS are shown in [Figure 3].{Figure 3}

Perioperative course

The total length of hospitalisation ranged from 4 to 27 days (median = 13). The length of stay in the intensive care ward ranged from 0 to 2 days (median = 1). Two patients (29%) did not require a stay in the ICU, three patients (42%) were observed in the ICU for 1 day and two patients remained in the ICU for 2 days [Table 2]. The above-described patient (patient no. 5), who underwent conversion to open surgery, remained in the hospital for 27 days. In addition to the longer recovery associated with the open approach, the patient developed an intra-abdominal seroma requiring irrigation through intraoperatively placed drains.

Post-operative complications were observed in three of seven patients: two patients had CD 3a and one CD 2. In detail, two patients developed a seroma in the plane of resection and were successfully treated with percutaneous drainage and irrigation. Patient no. 1 was treated with computed tomography-guided post-operative drain insertion and subsequent irrigation, classified as CD 3a. As mentioned above, patient no. 5 also developed a seroma in the plane of resection which was treated with irrigation of an intraoperatively placed drain, classified as CD 2. Neither of these patients showed signs of biliary leakage or abscess. Patient no. 6 developed a cholangitis and a left-sided pleural effusion, which were treated with ERCP-guided papillotomy, intravenous antibiotics and pleural catheterisation, classified as CD 3a.

Histopathology

All patients demonstrated characteristic histopathological findings of Caroli disease: cystic dilation of small, intrahepatic bile ducts (ranging from 7 to 20 mm) with varying degrees of chronic fibrosing and active cholangitis as well as portal fibrosis. Patient no. 5 had the most severe findings on histopathology: apart from severe cholestasis, a low-grade biliary intraepithelial dysplasia was seen, whereas resection lines were found to be tumour free. The same patient presented with hepatic steatosis (15%) and thrombosis of portal vein branches as well as sinusoidal obstruction [Figure 4].{Figure 4}

Discussion

Altogether nine (including our seven patients) cases of laparoscopic liver resections for Caroli disease are published so far.[9],[15] Concurrent with the literature, no gender predominance was found in our patient cohort.[18] Apart from Caroli disease, and some cases of metabolic disorders such as diabetes and hypertension or other common diseases such as hypothyroidism, our patients were overall in good health, which is reflected in the pre-operative ASA score of 2 in six of seven patients. Although some patients had standard previous abdominal surgery, such as appendectomy or cholecystectomy, almost half of the patients had never undergone abdominal surgery, making the conditions for laparoscopic liver resection favourable. The most commonly affected liver lobe was the left, predominantly segments II and III. The length of operation as well as length of hospital stay varied widely reflecting

the varying difficulty of surgery. Conversion to open hemihepatectomy was required only in one case due to severe disease manifestation masking a biliary variant in pre-operative imaging. This might indicate that laparoscopic resection is particularly well suited for patients with moderate to mild cases of Caroli rather than severe manifestations of the disease. Nevertheless, all patients might profit from primary laparoscopic evaluation, if conversion in difficult cases is performed in a non-emergent manner.

To our knowledge, this study represents the first larger case series of minimally invasive surgical treatment of Caroli disease. Two single case reports have previously been published.[9],[15] One patient underwent a left hemihepatectomy[9] and the other a resection of segments II and III.[15] Both operations were performed in multiport technique, and no conversions were described. The post-operative course was uneventful in one case and not described at all in the other.[15] Unfortunately, very little information was provided on operative and perioperative details and outcome (operation time, length of ICU and general hospitalisation as well as complications according to CD) making a comparison with our results difficult.

This presented case series is limited by a non-standardised protocol as well as a small patient number. To definitively assess the efficacy of laparoscopic liver resection in Caroli disease, a prospective trial using standardised operative protocols for laparoscopic and open performed surgery would be appropriate.

Nevertheless, to the best of our knowledge, this is the largest case series of laparoscopic liver resection for Caroli disease reported so far. Thus, the results of this study are an important contribution to further development of laparoscopic liver resection, showing that it is a safe and efficacious surgical treatment for patients with Caroli disease.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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