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Recurrent Hepatolithiasis and Hepatic Abscess Secondary to Caroli's Disease – Case Report

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Abstract
Background
Hepatolithiasis is the presence of gallstones in the intrahepatic ducts due to primary, idiopathic or infectious causes (lithogenic bile) or secondary causes such as congenital cysts and strictures or past hepatobiliary surgery. Typically, hepatolithiasis patients presents with the Charcot's triad (abdominal pain, jaundice, fever) suggestive of cholangitis. It is not uncommon for severe cholangitis to be accompanied with a hepatic abscess. The treatment consists in supportive measures for cholangitis, subsequent stone extraction and surgical removal of strictures and bile drainage. Patients should be closely monitored because of the high risk of recurrence, liver cirrhosis or cholangiocarcinoma.
Case presentation
Our patient is a 61 years old male with the following medical history and presentation: Right nephrectomy before approximately 15 years, for polycystic renal disease. Seven years ago, he underwent the surgical procedure of cholecystectomy and choledocho-duodenal anastomosis for gallbladder and CBD stones. A year later, he undergoes an urgent surgical procedure for an intra-abdominal abcess (right subphrenic). For the next five years the patient had no complaints. Three months prior to the surgery he is admitted to the hospital for persistent, recurring episodes of fever (38.5 °C), which did not respond well to antibiotic therapy. Radiology confirms intrahepatic stones, bilateral cystic dilations of bile ducts (Caroli's Disease) and an abscess of the VIIth liver segment. The patient continues a conservative treatment until optimal parameters are reached for the eventual procedure. The VIIth liver segment is partially resected, the hepatic calculi are extracted and a hepatico-jejunostomy performed, joining a Roux limb with the biliary confluence.
Discussion
Depending on patient history and current presentation in terms of the severity of the disease and the classification of hepatolithiasis, the surgeon has to weigh and decide upon possible treatment options. Among them we mention supportive measures to treat acute cholangitis, pharmacologic therapy (statins), percutaneous transhepatic cholangioscopic lithotomy, peroral cholangioscopic lithotripsy and surgery.
Conclusion
Patients with hepatolithiasis recurrence experience chronic cholangitis and eventually develop cirrhosis over a period of 10 to 20 years. Furthermore, the incidence of intrahepatic cholangiocarcinoma is about 5% to 10%. These patients require a long-term follow-up because of an increased risk of cancer after 10 to 20 years.
Keywords: General Surgery, Hepatobiliary Surgery, Hepatolithiasis, Caroli's Disease, Congenital Biliary Cysts.
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1. Introduction

Hepatolithiasis is the presence of gallstones in the intrahepatic ducts due to primary, idiopathic or infectious causes (lithogenic bile) or secondary causes such as congenital cysts and strictures or past hepatobiliary surgery.
According to stone location and presence of strictures, hepatolithiasis can be classified in five types (Takada et al.):
I – Mild dilation of the biliary system without strictures.
II – Stricture in the distal choledochus or ampulla of the duodenum.
III – Stricture at the hepatic hilum.
IV – Unilateral hepatic lobe stricture.
V – Multiple biliary strictures in both lobes or congenital biliary cysts (Caroli's Disease).

The presence of bile duct cysts indicates definitive surgical management based on an observed increase in the risk of cholangitis and pancreatitis, as well as malignant degeneration into cholangiocarcinoma (12-16%).

Typically, hepatolithiasis patients presents with the Charcot's triad (abdominal pain, jaundice, fever) suggestive of cholangitis. It is not uncommon for severe cholangitis to be accompanied with a hepatic abscess.

A diagnosis of hepatolithiasis is made by blood tests, with signs of systemic inflammation and cholestasis, in addition to imaging studies, with possible findings of intrahepatic calculi and biliary strictures or dilations. Useful imaging methods include ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI), endoscopic retrograde cholangiography (ERC) or percutaneous transhepatic cholangiography (PTC).

The treatment for hepatolithiasis consists in the supportive management of cholangitis, subsequent stone extraction and surgical removal of strictures and bile drainage. Patients should be closely monitored because of the high risk of recurrence, liver cirrhosis or cholangiocarcinoma.

2. Case presentation

2.1 Medical history

Our patient is a 61 years old male with the following medical history and presentation:

Right nephrectomy before approximately 15 years, for polycystic renal disease.

Seven years ago, he underwent the surgical procedure of cholecystectomy, choledochotomy (bile duct exploration) and choledocho-duodenal anastomosis for gallbladder and CBD stones.

A year later, he undergoes an urgent surgical procedure for an intra-abdominal abcess (right subphrenic).

For the next five years the patient had no complaints and did not show up for medical consultation.

Three months prior to the surgery he is admitted to the hospital for persistent, recurring episodes of fever (38.5 °C), which did not respond well to antibiotic therapy.

An MRI (Figure 1) confirms intrahepatic stones, bilateral cystic dilations of bile ducts (Type V of bile duct cysts, Caroli's Disease) and an abscess of the VIIth liver segment.

The patient continues a conservative treatment until optimal parameters are reached for the eventual procedure.

"Mother Teresa" University Hospital Center
Central Service of Imaging

Patient name: L... N...
Age: 61
Examination date: 21.09.2022

Abdomen MRI

Technique: Images were obtained with T2 TSE, T2 Spair, dual FFE, BTDE, e-THRIVE sequences, on axial and coronal planes, with IV contrast.

Results

Liver, with a 54×68 mm formation on segment VII, with hypersignal in T2, signal restriction in DWI, which is contrasted in an irregular annular pattern; an aspect in favor of an abscess on the terrain of cholangitis. Hypersignal on the hepatic parenchyma surrounding this lesion.
Left liver lobe hypoplasia and cystic dilations of intrahepatic biliary ducts, more notable on the left side; which have signal restriction in DWI.
A signal defect is noticed, compatible with the presence of calculi in the left and right hepatic ducts.
Cholecystectomy. Common bile duct with a diameter of 5 mm.
Portal vein and suprahepatic veins free.
Atrophic pancreas with normal signal intensity.
Wirsung duct with normal aspect.
Surrenal glands with normal morphology and signal intensity.
Right nephrectomy.
Left kidney with normal dimensions and parenchyma, without dilation of the pyelocaliceal system.
Cortical cysts of diameters up to 10 mm.
No free abdominal fluid. No aggressive bone lesions.

Dr. Ergisa Toska
Electronic signature

Figure 1. MRI report.

2.2 Details of the procedure

A chevron incision was chosen for adequate exposure of the liver.

Immediately, numerous adhesions are encountered, more notable on the upper abdominal compartments. They are dissected and released carefully, taking up a considerable amount of time (*Figure 2*). The diaphragm is divided from its tight attachment with the liver, which in turn exposes the latters.

Careful dissection of at the hepatic hilum follows. An aberrant right hepatic artery is identified and a vascular loop is passed (*Figure 4*). The common bile duct is identified and separated with a loop (*Figure 5*). A vascular loop is also passed around the entire hepatic peduncle in case a Pringle manoeuvre is needed.

On the right hepatic lobe, segment VII, a semi-calcified abscess of 4 × 4 cm dimensions is evidenced. A partial resection of the VIIth lobe is executed, until we come upon healthy hepatic tissue (*Figure 6*).

Next, the hepatic bifurcation is prepared, where the right and left ducts are visible. At the bifurcation level, where the common hepatic duct was the widest, an amputation is done. Subsequently, a considerable number of intrahepatic gallstones are extracted from both the right and left ducts using Desjardin's forceps and a copious amount of isotonic saline lavage (Figure 7, 8).

The jejunal loop is prepared and an anastomosis between the hepatic bifurcation and the intestine is performed in a Roux-en-Y fashion (Figure 9, 10).

A Kehr drain (T-tube) is left on the right hepatic duct to protect the anastomosis (*Figure 11*).

2.3 Post-operative period

The patient tolerated the procedure well, with uneventful post-operative course. He was discharged in good health. The T-tube was removed two months after the surgery.

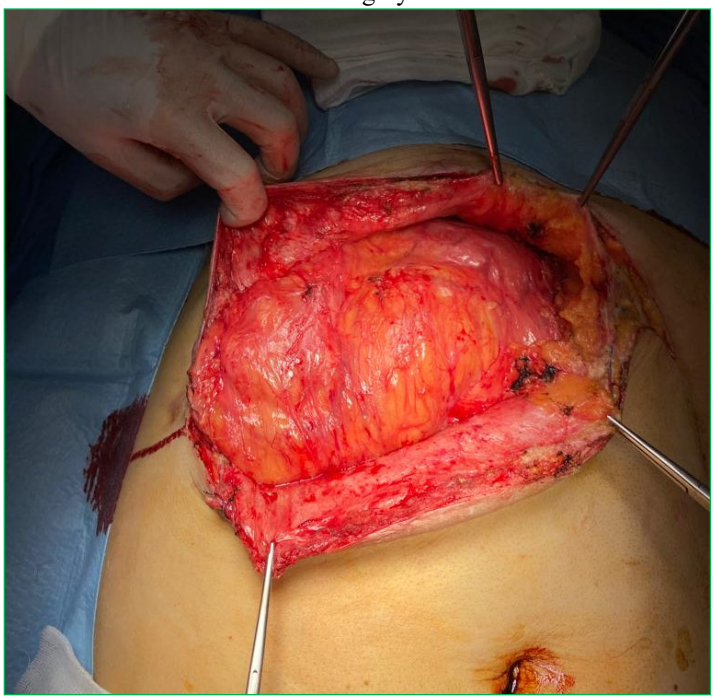

Figure 2. Notable adhesions upon entering peritoneal cavity.

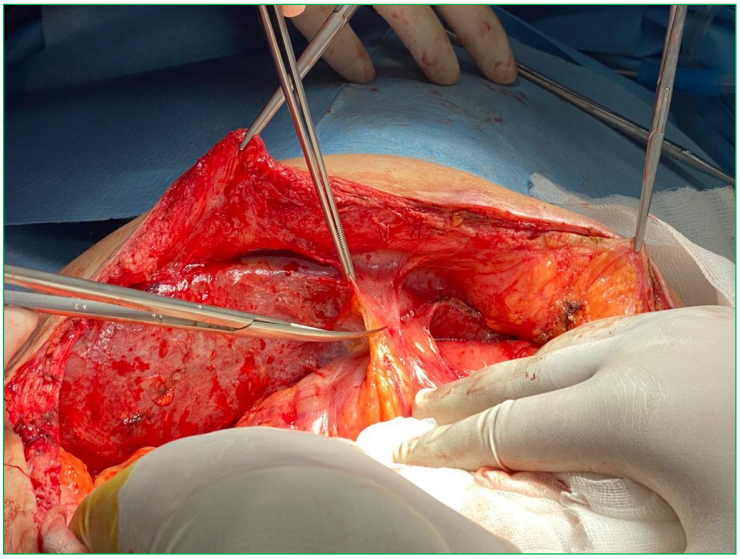

Figure 3. Dissecting the adhesions.

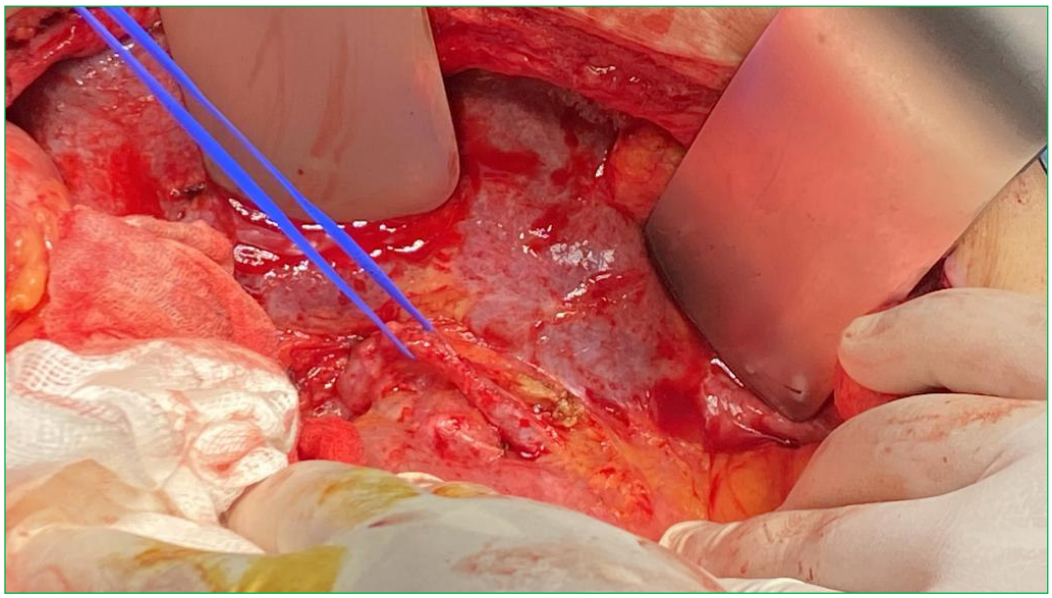

Figure 4. Exposing the right hepatic artery (aberrant variation).

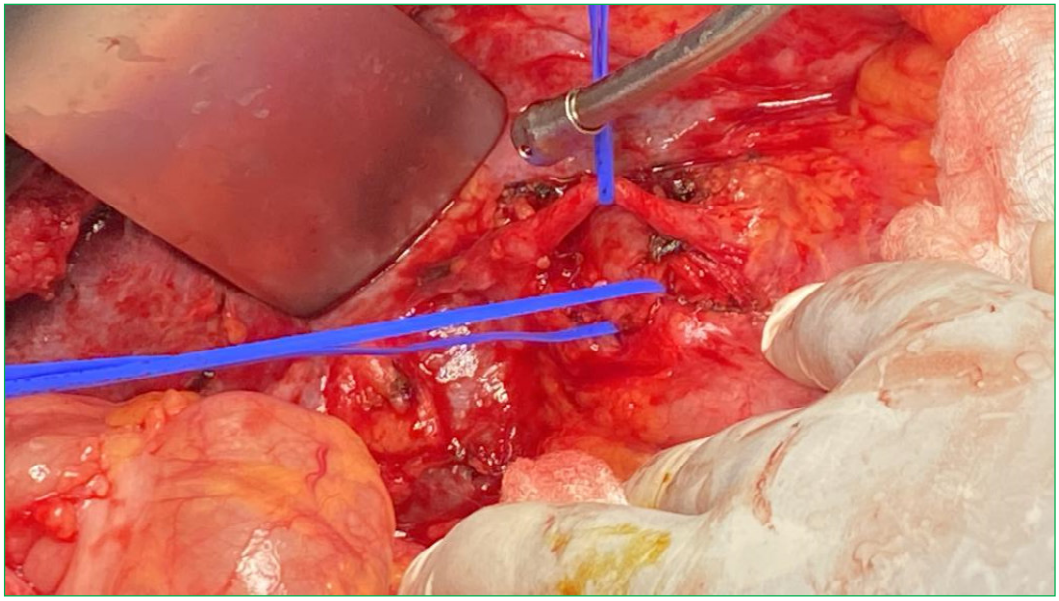

Figure 5. Exposing the common bile duct (left loop) and right hepatic artery (upper vascular loop).

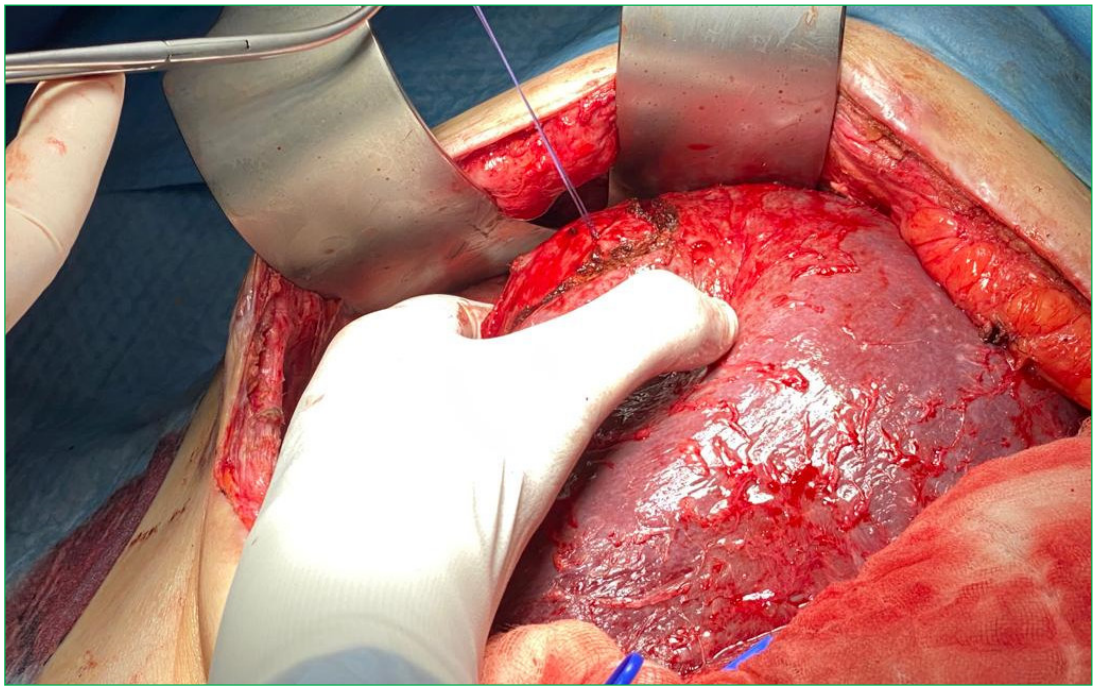

Figure 6. Partial resection of segment VII.

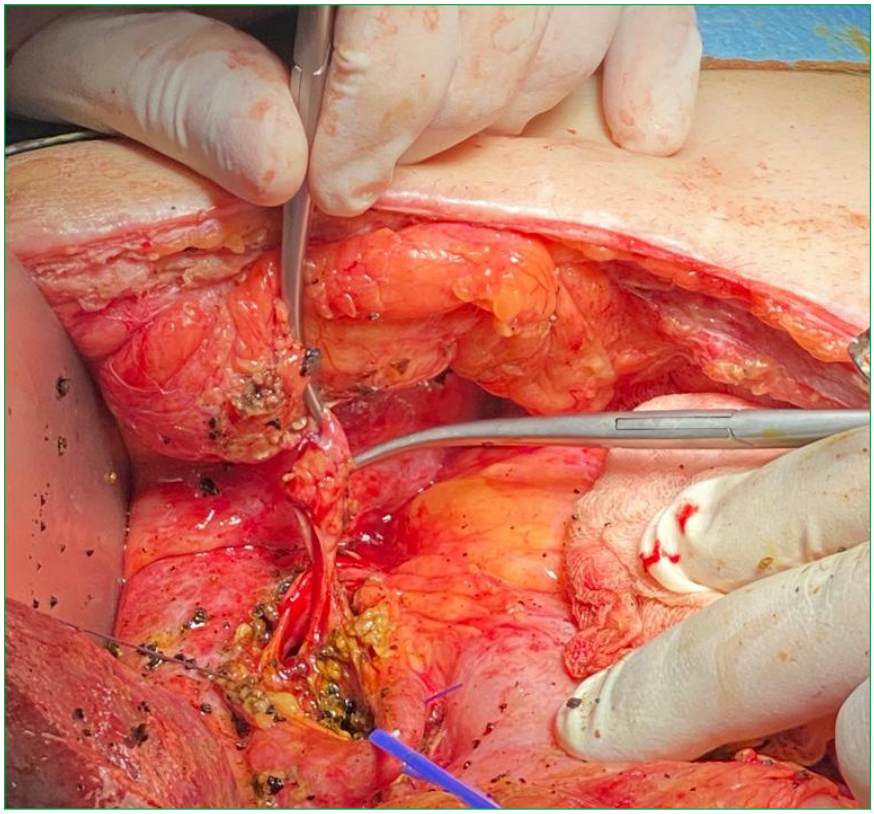

Figure 7. Exploration and lavage of the hepatic ducts.

Figure 8. A considerable number of gallstones removed from hepatic ducts. (Diameter up to 1.5 – 2 cm)

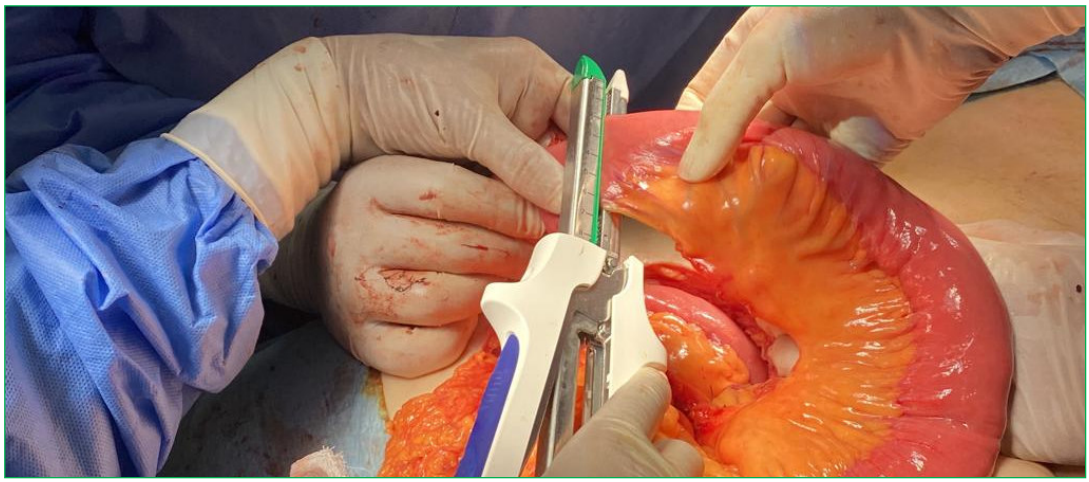

Figure 9. Preparing the Roux jejunal limb.

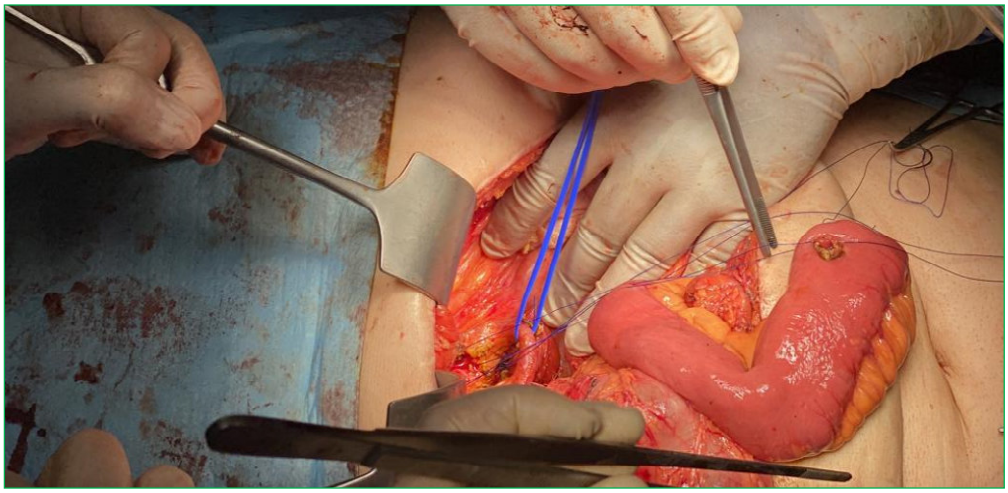

Figure 10. Hepatico-jejunal anastomosis.

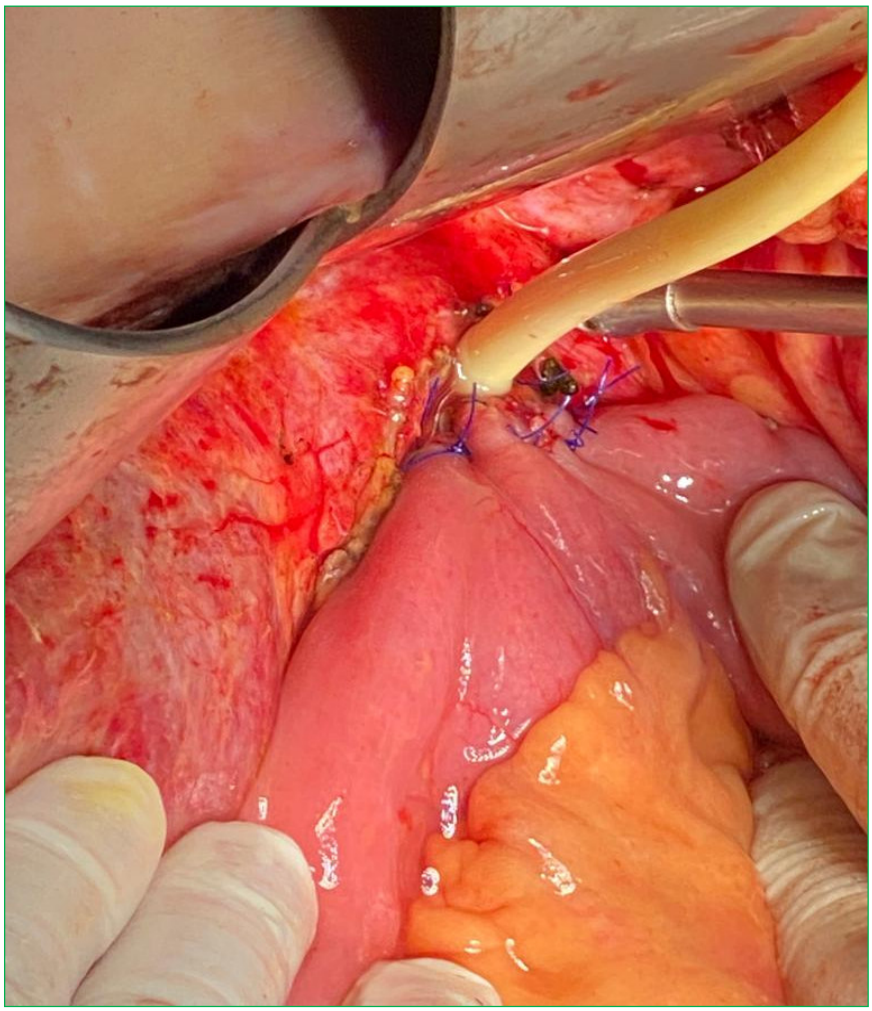

Figure 11. Completed hepatico-jejunal anastomosis, protected with Kehr drain.

3. Discussion

Depending on patient history and current presentation in terms of the severity of the disease and the classification of hepatolithiasis, the surgeon has to weigh and decide upon possible treatment options. Among them we mention supportive measures to treat acute cholangitis, pharmacologic therapy (statins), percutaneous transhepatic cholangioscopic lithotomy, peroral cholangioscopic lithotripsy and surgery.

As most hepatolithiasis patients presents with signs and symptoms of cholangitis, their initial management should include fluid and electrolyte resuscitation, along with broad spectrum antibiotics.

The ultimate treatment of hepatolithiasis consists in complete removal of calculi and the possible causative agent, mostly biliary strictures. Surgeons must consider hepatectomy when one side is atrophied with significant bile duct dilations and strictures.

Until now, there is no consensus on a medical drug therapy for hepatolithiasis, since the causative factors have not yet been established, but there are studies to support the role of statins (simvastatin) in decreasing the risk for gallstones.

Several surgical options include: cholecystectomy and bile duct exploration with T-tube insertion, choledocho-duodenal anastomosis, hepatico-jejunostomy (with or without anchoring the jejunal limb to the abdominal wall for stone extraction), right/left hepatectomy and liver transplant.

4. Conclusion

Hepatolithiasis is a challenging condition that requires careful evaluation. Determining the correct type and extent of hepatolithiasis leads to better decision-making in terms of operative strategy.

Caroli's disease has no specific signs or symptoms, which makes diagnosis of this rare condition difficult. However, knowing its association with conditions such as intrahepatic calculi, cholangitis, pancreatic cysts and renal cysts should raise our suspicion.

Patients with hepatolithiasis recurrence experience chronic cholangitis and eventually develop cirrhosis

over a period of 10 to 20 years. Furthermore, the incidence of intrahepatic cholangiocarcinoma is about 5% to 10%. These patients require a long-term follow-up because of an increased risk of cancer after 10 to 20 years.

For unilateral hepatic cystic disease with liver fibrosis and atrophy there is a general consensus that liver resection is a preferred treatment. Meanwhile, for bilateral hepatic cysts, a hepatico-jejunostomy is an effective option, before the necessary criteria are met for liver transplantation.

Conflict of interest

The author(s) declare(s) that there is no conflict of interest. The authors alone are responsible for the content and writing of the paper.

Financial disclosure

There is no financial support to this study.

Ethical aspect

Informed consent was obtained from all participants in the study and all procedures were conducted in accordance with the Declaration of Helsinki.

References

1. Jarnagin WR, Allen P, D'Angelica M, DeMatteo R, Kinh Gian Do R, Vauthey JN – Blumgart's Surgery of the Liver, Biliary Tract, and Pancreas, 6th ed, Elsevier 2017, p762, p642. ISBN: 978-0-323-34062-5.
2. Al-Sukhni W, et al: Recurrent pyogenic cholangitis with hepatolithiasis – the role of surgical therapy in North America, J Gastrointest Surg 12:496–503, 2008.
3. Bodmer M, et al: Statin use and risk of gallstone disease followed by cholecystectomy, JAMA 302:2001–2007, 2009.
4. Nishioka T, et al: Administration of phosphatigylcholine-cholesterol liposomes partially reconstitutes fat absorption in chronically bile diverted rats, Biochim Biophys Acta 1636:90–98, 2004.
5. Kim MH, et al: Primary intrahepatic stones, Am J Gastroenterol 90:540–548, 1995.
6. Guglielmi A, et al: Hepatolithiasis-associated cholangiocarcinoma: results from a multi-institutional national database on a case series of 23 patients, Eur J Surg Oncol 40:567–575, 2014.
7. Suzuki Y, et al: Predictive factors for cholangiocarcinoma associated with hepatolithiasis determined on the basis of Japanese multicenter study, Hepatol Res 42:166–170, 2012.
8. Takada T, et al: Classification and treatment of intrahepatic calcali, Jap J Gastroenterol Surg 11:769–774, 1978.
9. Akiyama T, et al: Recurrence of intrahepatic stones after an end-to-side choledochojejunostomy, Surg Today 24:599–605, 1994.
10. Liu CL, et al: Primary biliary stones: diagnosis and management, World J Surg 22:1162–1166, 1998.