Laparoscopic Cholecystectomy And Unroofing Of A Hepatic Cyst In A Patient With Polycystic Liver Disease

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INTRODUCTION

Polycystic liver disease (PLD) represents a group of genetic disorders presenting with multiple hepatic cysts. PLD may occur in isolated form as autosomal dominant polycystic liver disease (ADPLD) or in association with autosomal dominant polycystic kidney disease (ADPKD). Estimated incidences are 0.01% and 0.2%, respectively,[1] and both entities present a autosomal dominant hereditary pattern. The natural history of PLD is characterized by a continuous increase in the volume and the number of cysts.[2] Symptoms will appear in 20% of patients with PLD,[3] generally due to the expansion of hepatic volume.

Mild common bile duct dilation has also been reported,[4] but unlike the much rarer autosomal recessive form of polycystic kidney disease, which is associated with nonobstructive intrahepatic duct dilation (Caroli disease) and recurrent cholangitis,[5] clinically significant biliary tract complications are less well recognized in patients with polycystic disease.

Upon literature review in PubMed, biliary complications in patient with polycystic liver or kidney disease is sparse. Pathology included obstructive jaundice due to enlarged cysts, and less so, symptomatic cholelithiasis. In a single center study of greater than 1,000 patients with autosomal dominant polycystic diseased, compared with non-ADPD hospital controls, patients had higher rates of admission for biliary tract disease (rate ratio [RR], 2.24; 95% confidence interval [95% CI]. Overall, biliary tract disease seems to be a distinct and important extrarenal complication.[6]

Our case highlights the complexity of performing a laparoscopic cholecystectomy for symptomatic biliary disease in the setting of polycystic liver disease.

CASE REPORT

A 48-year-old woman with a past medical history of asthma, polycystic kidney disease, polycystic liver disease, brain aneurysm with clipping presented to our surgical clinic for right upper quadrant abdominal pain. She states that she has been experiencing abdominal discomfort for the past 3-4 months consistent with fat intolerance. She was worked up outpatient, including liver enzymes, and a right upper quadrant ultrasound supporting the diagnosis of biliary colic. Pre-operative CT of her abdomen-pelvis was showing a large liver cyst abutting the gallbladder infundibulum, amongst innumerable other surrounding cysts. She was sent for pre-operative testing and taken for an elective laparoscopic cholecystectomy.

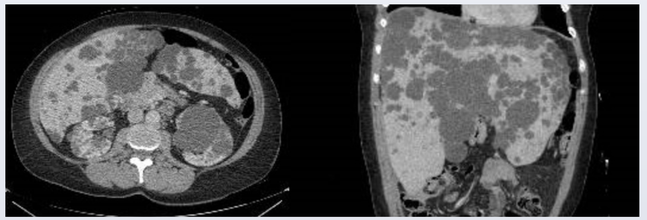


Figure 1. Pre-operative CT showing multiple liver and kidney cysts

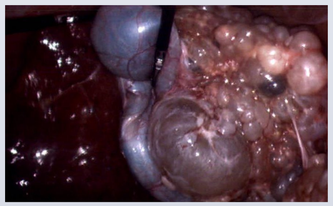




Figure 2,3. Intraoperative view of dominant hepatic cyst abutting gallbladder edge.

In the operating room, her liver was palpable below the right costal margin, thus laparoscopic ports placed were placed in a inferior position to avoid organ injury. The abdomen was entered via Hassan technique. Upon visual inspection, the liver was noted to be completely covered with thin walled cysts [Figure 2]. A large cyst was overlying the infundibulum and cystic duct. After attempting dissection of the lower third of the gallbladder, aspiration of the cyst was deemed necessary to visualize the critical view of safety. Despite aspiration, the cystic plate could not be adequately visualized and unroofing of the cyst wall was carried out. The biliary structures were then visualized and transected after endoclipping. The gallbladder was removed with adequate hemostasis and without bile leak. The edges of the cyst were then electrocauterized. A #10 Jackson-Pratt drain was left in place in the gallbladder fossa. She was successfully extubated and discharged from PACU in stable condition.

DISCUSSION

This case illustrates the rarity and complexity of biliary pathology with concomitant polycystic liver and polycystic kidney disease. Benign biliary disease can be diagnosed in standard fashion. Tokyo guidelines have been created describing standard techniques for laparoscopic cholecystectomy in additional to risk factors associated with prolonged operative time and open conversion, but do not include cystic liver.

The management of hepatic cysts range from percutaneous procedures and sclerotherapy, to hepatic resection and liver transplantation. Although no standardized approach, intraoperatively we decided to use aspiration and deroofing. This is predominantly used for symptomatic cysts, but this dominant cyst hindered progression and deemed necessary. In one prospective study, laparoscopic deroofing was regarded as a safe and functional treatment, with a 9% average recurrence rate and 4.5% symptomatic recurrence.[7] The most common post-operative complication for cyst fenestration are ascites, pleural effusion, and bile leaks.

CONCLUSION

Cystic lesions of the liver consist of a heterogeneous group of disorders and might present a diagnostic and therapeutic challenge during laparoscopic cholecystectomy. The anatomical localization might present an additional risk factor in the incidence of bile duct injury. When working up a patient for biliary disease found to have PLD or PKD, it is important to know the varied options when encountering obstructing simple cysts, as there is no standardized approach.

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