LAPAROSCOPIC & ENDOSCOPIC MANAGEMENT OF LIVER CYST WITH BILIARY FISTULA IN PATIENT WITH POLYCYSTIC LIVER DISEASE

Nair, Ranjit[1]; Ginting, Karolin M.D.[2]; Allamaneni, Shyam M.D.[2]

[1]Saba University School of Medicine, [2]Mercy Health – The Jewish Hospital

Background

Isolated Polycystic Liver Disease (PLD) is a rare disease related to embryonic malformation that spreads cysts throughout the parenchyma. In most cases, patients are asymptomatic, but increasing size of cysts can cause abdominal pain and distension, which can be worsened by cyst rupture, hemorrhage, or infection. In patients with very large infected cysts, laparoscopic techniques could offer potential technical advantages in drainage. In a case of biliary fistulation to the cyst, further endoscopic treatment may be indicated.

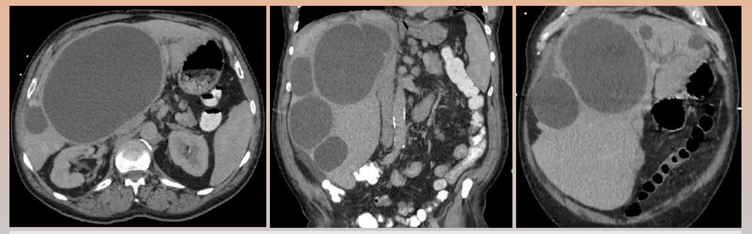


Figure 1. Multiple large hepatic cysts, the largest measuring 19 cm

Introduction

Diagnostic criteria for PLD can be loosely defined as patient with 20 hepatic cysts or more with family history or patient with more than four hepatic cysts without family history. It is pertinent to distinguish PLD from polycystic kidney disease as the latter can result in progressive kidney failure and both preventative and late-stage treatments would be different than they would be in PLD.

In abscess formation due to cyst infection, fenestration and drainage is primarily considered via image-guided percutaneous methods, open surgical methods, laparoscopic methods, or endoscopic retrograde cholangiopancreatography (ERCP). ERCP is typically utilized for abscess drainage where the infection communicates with the patient's biliary tree. Otherwise, the treatment method depends on size, number, and accessibility of infected cysts. Laparoscopy has been considered to be a more precise way to drain infected cysts. We present and discuss this initial laparoscopic drainage along with the subsequent procedures and sequelae of events involving a PLD patient with infected cysts.

Case

A 68-year-old male with polycystic liver disease (PLD) initially presented with fevers and right upper quadrant abdominal pain. Computed Tomography (CT) scan study showed multiple hepatic cysts with one large cyst containing debris (Figure 1). Primary procedure included laparoscopic fenestration, drain placement and biopsy of the cystic wall, which led to symptom resolution (Figure 2). Fluid culture was positive for *E.coli* and pathology of the cyst wall revealed fibrous stroma with one edge containing bile ductules, favoring simple cyst. Postoperatively, the drain output became bilious which was concerning for biliary fistula to the cyst. ERCP with sphincterotomy and biliary drain placement were completed (Figure 3). The patient required multiple drain exchanges due to persistent fistula that eventually resolved within a year. In the following year, the patient presented again with similar symptoms. CT scan showed fluid collection located along the previous drain tract, pushing up against the diaphragm (Figure 4). Percutaneous drainage was done at this time due to its proximity to the skin. Culture fluid was positive for *Streptococcus anginosus*. Follow up drain study showed no biliary connection. The drain was removed 4 weeks later, and the patient is currently doing well. Of note, the patient was given an extended period of antibiotic treatment as well.

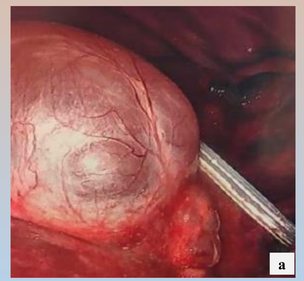


Figure 2. One of the hepatic cyst was entered and drained (a). Cyst wall was biopsied (b).

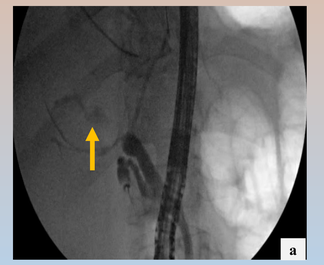


Figure 3. Biliary leak was noted during ERCP (a). Sphincterotomy and CBD stent placement were done to allow for internal drainage (b).

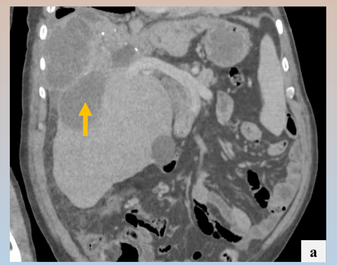


Figure 4. Hepatic cyst at the previous drain track against diaphragm (a) was drained percutaneously with resolution of symptoms (b).

Discussion

In the setting of PLD, cyst cavities could become infected, requiring external or internal drainage. Infections of cyst cavities are a serious complication, due to high risk for recurrence[1]. Infections may be due to biliary fistulation to the cyst (an endogenous route) or secondary to an exogenous transmission route. ERCP is both diagnostic and therapeutic when sphincterotomy and CBD stent placement is performed. Stent placement ultimately decreases the pressure gradient between the duodenum and common bile duct, allowing proper diversion of the bile away from the fistula and aid fistula closure[2]. Other treatment options for infected or symptomatic cysts include percutaneous aspiration, surgical cyst excision, or partial liver resection.

Our patient had initial surgical excision, deroofing, and then drain placement after finding a bile duct fistula within the cyst. Cystojejunostomy is a useful method of treatment in cases where biliary communication with the cyst exists, but there is a risk of postoperative sepsis due to enteric bacterial colonization of the cyst fluid[3]. Partial liver resection carries significant risk and morbidity as recurrence of the infection and fistula formation in cysts on other areas of the liver could continue to occur in the future.

Conclusion

History of liver cysts with biliary fistula in patients with PLD carries high risk of recurrence as illustrated in this case. In the event of biliary fistulation to the cyst, sphincterotomy and biliary stenting are feasible in allowing the biliary fistula to close. Additionally, it is important to discuss the potential need for recurrent drainage and prolonged antibiotics with patients.

References:

1. Cnossen,W.R., Drenth, J.P. Polycystic liver disease: an overview of pathogenesis, clinical manifestations and management. Orphanet J Rare Dis 9, 69 (2014). https://doi.org/10.1186/1750-1172-9-69

2. Canena, J., Horta, D., Coimbra, J. et al. Outcomes of endoscopic management of primary and refractory postcholecystectomy biliary leaks in a multicentre review of 178 patients. BMC Gastroenterol 15, 105 (2015).
https://doi.org/10.1186/s12876-015-0334-y

3. Masatsugu, T., Shimizu, S., Noshiro, H., Mizumoto, K., Yamaguchi, K., Chijiiwa, K., & Tanaka, M. (2003). Liver cyst with biliary communication successfully treated with laparoscopic deroofing: a case report. JSLS : Journal of the Society of Laparoendoscopic Surgeons, 7(3), 249–252.