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Original Research Article

An Observational Study Was to Assess Laparoscopic Fenestration in Patients with Polycystic Liver Disease and Long-Term Outcome

Sanjay Kumar[1], Tushar Saini[2], Nitesh Kumar[3], Saket Kumar[4], Manish Mandal[5]

[1]Additional Professor, Department of Surgical Gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India

[2]Senior Resident, Department of Surgical Gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India

[3]Additional Professor, Department of General Surgery, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India

[4]Additional Professor, Department of Surgical Gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India

[5]Professor and Head, Department of Surgical Gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India

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Corresponding Author: Dr. Tushar Saini

Conflict of interest: Nil

Abstract

Aim: The aim of the present study was to assess laparoscopic fenestration in patients with Polycystic liver disease and long-term outcome.

Methods: The present study was conducted in patients with PCLD were referred to the Department of surgical gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India. 50 patients were included in the study. Unfenstrated patients were 35 and 15 were fenestrated patients. It is retrospective observational study.

Results: 35 patients had no surgical treatment for their liver cysts. 23 of these patients were asymptomatic or had mild pain that was relieved with an oral analgesic. Cholangitis secondary to biliary lithiasis and an unresectable cholangiocarcinoma were present in one patient each. The remaining 15 patients underwent wide fenestration of the cysts and so form the basis of this retrospective review. One patient had suppurative cholangitis secondary to compression of the left intrahepatic bile ducts by the cysts. Pain was never the result of an acute complication such as hemorrhage, superinfection, or acute distension of cysts.

Conclusion: In conclusion, deroofing should be performed only in patients with PCLD who have significant related disorders and minimal renal dysfunction. Because of the high incidence of this disease, surgery is probably rarely indicated. Fenestration is a safe, effective procedure when the noncystic liver is normal and when the cysts may be anticipated to collapse after operation. Rarely, symptomatic patients seem to have highly advanced disease, resulting in a rigid, noncystic liver. These patients seem to be at a higher risk of postoperative complications (mainly ascites) and shorter efficacy of treatment.

Keywords: Operative Outcomes, Polycystic Liver Disease (PCLD). ADPKD (Autosomal Dominant Polycystic Kidney Diseases)

Introduction

Polycystic liver disease (PCLD) exists as the most common extrarenal manifestation in polycystic kidney disease, or less commonly, in an isolated fashion. [1–3] The natural history of the disease results in progressive development in number and size of liver cysts. [1–4] While a majority of patients remain asymptomatic, some patients develop incapacitating hepatomegaly with resultant digestive or respiratory symptoms like pain, inability to eat, weight loss, dyspnea etc. In advanced cases, patients may develop portal hypertension or hyperbilirubinemia. Treatment of PCLD thus covers wide range of therapeutic options ranging from non interventions to liver transplantation, including needle aspiration evacuation with injection of sclerosant, laparoscopic fenestration and fenestration by laparotomy combined with liver resection. Choice between these different treatment depends on symptomatology, the intrahepatic extension of lesions and patients general condition. Treatment remains focused on volume reduction for

symptomatic management, with medical therapy aimed toward reducing cystic secretions and limiting cystic growth. [4,5]

Recent randomized controlled trials have shown that somatostatin analogues, such as octreotide and lantreotide, result in short-term liver volume reduction in selected patients, [5–8] though the long term effect has yet to be investigated.7 Surgical therapy is dictated by size and distribution of cystic lesions. [4,9] Large hepatic cysts with favorable anatomy are treated with fenestration either by laparotomy or laparoscopic, which is associated with long term better clinical outcome. [4,6,10] Hepatic resection and/or liver transplantation are reserved for patients with diffusely distributed small, numerous cysts. [4,11,12]

PCLD is a rare (incidence < 0.01%) dominantly inherited disorder characterized by multiple diffuse cystic lesions of the liver parenchyma. An asymptomatic enlarged liver is usually the hallmark of the disease. However with more effective treatment of renal disease, increasing numbers of patients are living long enough to experience symptoms from their associated polycystic liver disease. Significant symptoms or complications from liver involvement can occur in up to 20 percent of cases. [13,14] Although there is an isolated form of polycystic liver disease, knowledge concerning the pathogenesis of hepatic cysts was gained from the study of hepatic cysts in ADPKD. These lesions have been attributed to bile duct overgrowth after the arrest of embryogenesis and failure of the intralobar bile ducts to involute. This involutional failure results in cystic dilations that are known as biliary microhamartomas or von Meyenburg complexes (VMC). [15]

The aim of the present study was to assess laparoscopic fenestration in patients with Polycystic liver disease and long-term outcome in patients who had multiple large cyst, symptomatic hepatomegaly, laparoscopically reachable location and favorable hepatic anatomy.

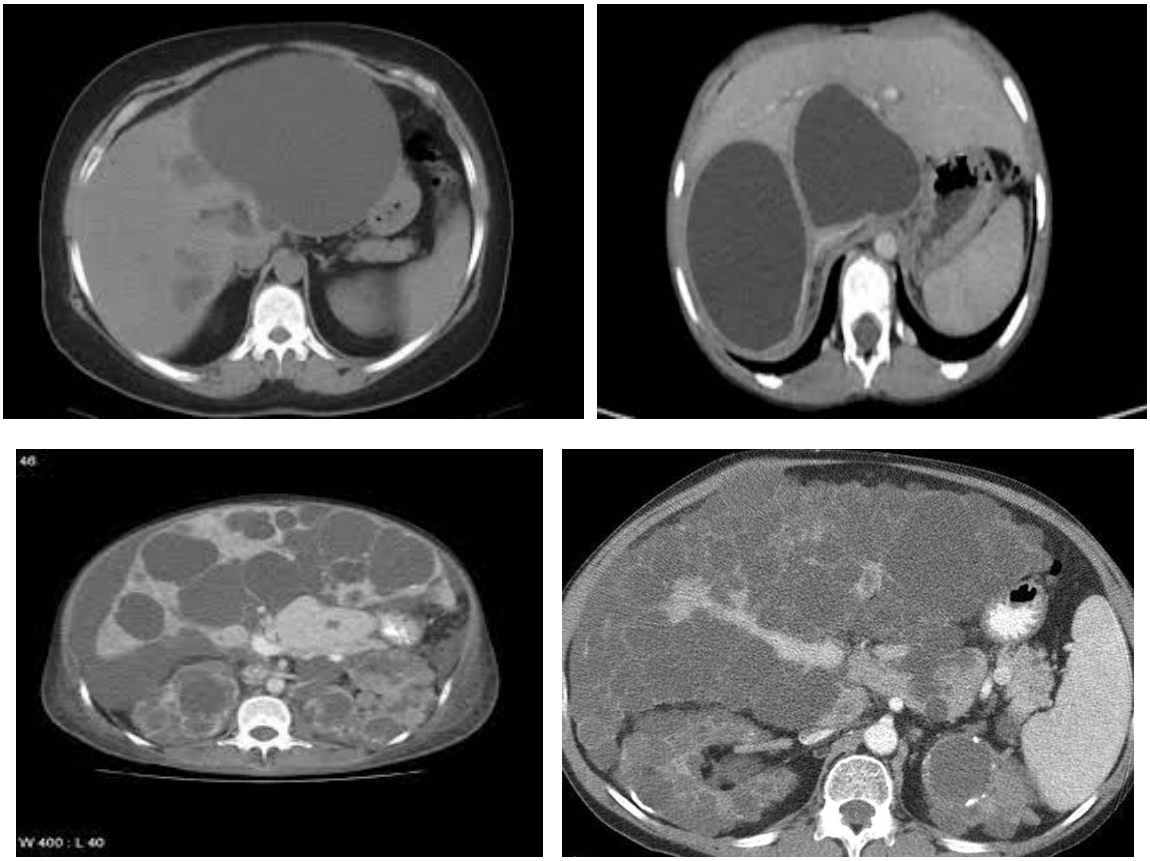
Materials and Methods

The present study was conducted in patients with PCLD were referred to the Department of surgical gastroenterology, Indira Gandhi Institute of Medical Sciences, Sheikhpura, Patna, Bihar, India for 3 year (2020 to 2023). 50 patients were included in the study, out of which 28 were female and 22 were male, belonging to ages between 42 to 60 years. Unfenstrated patients were 35 and 15 were fenestrated patients. PCLD was differentiated from multicystic liver disease (MCLD)- though the two are probably variants of the same disease- according to the following criteria: (1) liver enlargement, an inconstant feature in patients with MCLD; (2) cysts too numerous to be counted (patients with MCLD, in comparison, seldom have more than five cysts); (3) the diameters of the cysts being within a comparable range (patients with MCLD often have one large cyst and multiple smaller ones).

Morphologic Pattern of PCLD and Classification of Patients Cysts were diffusely scattered in both lobes of the liver in all patients, and most were smaller than 5 cm. According to the number of the cysts and the volume of the noncystic liver, out of the 15 fenestrated patients were 10 patients had a significant number of cysts but still had large areas of normal-appearing liver. In the five(5) patients, the liver was completely replaced by cysts, and on computed tomography (CT) scans the parenchyma appeared as thin bands compressed between the cysts.

Operative Procedure

The operative approach was laparoscopic fenestration. Adhesions found between the liver and the diaphragm in those patients who had previously undergone USG guided aspiration or had episodes of cyst infection. As many cysts as possible were widely deroofed, starting from the most superficial ones and then stepwise opening the deeper ones. Preoperative CT scans proved useful for locating the involved liver segments and determining the route of access to the deep cysts and also the port positions for laparoscopic fenestration. The cystic content was always clear. The average volumes of fluid aspirated during laparoscopy were not different in the two groups. The abdomen was drained prior to closure in four patients. spirinolactone was given to one patient during the postoperative period to reduce the volume of ascites [7]. Kinesitherapy was always performed during the postoperative period to improve breathing kinetics and the abdominal girdle.



Results

Table 1: Complaints

Unfenestrated patients (n=35)	
Moderate pain or symptoms	23
Cysts, infection	7
Clinical ascites	3
Cholangiocarcinoma	1
Cholangitis	1
Fenestrated patients (n=15)	
Incapacitating RUQ pain	13
Respiratory distress	7
Clinical ascites	4
Renal cyst	2
Leg edema	1
Cholangitis	1

35 patients had no surgical treatment for their liver cysts. 23 of these patients were asymptomatic or had mild pain that was relieved with an oral analgesic, 7 patients had cyst infection leading to right quadrant pain and fever, 3 patients had ascites. Cholangitis secondary to biliary lithiasis and an unresectable cholangiocarcinoma were present in one patient each. Female Patients are more symptomatic than male.

The remaining 15 patients underwent wide fenestration of the cysts and so form the basis of this retrospective review. One patient had suppurative cholangitis secondary to compression of the left intrahepatic bile ducts by the cysts. Pain was not the result of an acute complication such as hemorrhage, superinfection, or acute distension of cysts.

Discussion

Polycystic liver disease (PCLD) is an autosomal dominant disorder resulting in numerous cyst spread throughout the liver parenchyma. The cysts develop from bile ducts malformations originating from peripheral biliary tree which are called Von Meyenburg complexes (VMC) that failed to establish connections embryologically with the larger interlobular bile ducts.[16] The prognosis is

classically determined by the renal cystic involvement that is associated in up to 50% of the patients and may result in impaired renal function.[16,17] Liver cysts, which develop in most patients later than renal cysts[14,18], are usually clinically silent[18,19] but may lead to symptoms as a result of cyst infection; the pressure effect on hepatic ducts, liver vessels, or adjacent structures; or malignant transformation, especially in the patients with polycystic kidney disease whose lives have been prolonged by dialysis or renal transplantation.[14]

35 patients had no surgical treatment for their liver cysts. 23 of these patients were asymptomatic or had mild pain that was relieved with an oral analgesic. Cholangitis secondary to biliary lithiasis and an unresectable cholangiocarcinoma were present in one patient each. The remaining 15 patients underwent wide fenestration of the cysts and so form the basis of this retrospective review. One patient had suppurative cholangitis secondary to compression of the left intrahepatic bile ducts by the cysts. Pain was not the result of an acute complication such as hemorrhage, superinfection, or acute distension of cysts. Polycystic liver disease has been previously classified according to the diameter of the cysts.[20] In the present study, in contrast, the morbidity and long-term results of fenestration have appeared to be at least partly related to the diffusion of the cysts in the liver or to the morphologic aspect of the noncystic liver. Most patients, though having a significant number of cysts, still had large areas of noncystic liver on preoperative CT scans. In these patients, fenestration was a safe procedure; it provided long lasting relief of the symptoms and significantly reduced the size of the liver and the size and number of the cysts. This result closely resembles our experience and that of others who have performed fenestration of solitary or multiple cysts of the liver[21] and is compatible with the progressive collapse of the unroofed cysts.

Conclusion

In conclusion, laparoscopic fenestration should be performed only in patients with PCLD who have significant symptoms related to the disorder and minimal renal dysfunction. Fenestration is a safe, effective procedure when the noncystic liver parenchyma is normal and when the cysts may be anticipated to collapse after operation. Long term outcome is very good without any significant complications. Rarely, symptomatic patients seem to have highly advanced disease, resulting in a rigid, noncystic liver. These patients seem to be at a higher risk of postoperative complications (mainly ascites) and shorter efficacy of treatment.

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