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• BRIEF REPORTS •

Combined hepatic resection with fenestration for highly symptomatic polycystic liver disease: A report on seven patients

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Abstract

AIM: To evaluate the immediate and long-term results in a series of patients with highly symptomatic polycystic liver disease (PLD) treated by combined hepatic resection with cystic fenestration.

METHODS: We reviewed our recent experience with a combined hepatic resection-fenestration procedure in seven highly symptomatic patients with PLD. Clinical data, liver manifestation of computed tomography (CT), and morbidity were recorded pre- and post-operation. Follow-up was made by clinical and CT examinations in all patients.

RESULTS: Symptomatic relief and reduction in abdominal girth were obtained in all patients during an average follow-up period of 20.4 mo. CT scans confirmed post-resection hypertrophy of the spared liver and lack of significant cyst progression. All patients had mild to severe ascites. Two patients were complicated with pleural effusion.

CONCLUSION: Some highly symptomatic patients with massive PLD may benefit from combined hepatic resection and fenestration at acceptable risk. To stitch the dissected hepatic ligaments could prevent the instable remnant liver from kinking and collapsing.

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INTRODUCTION

Polycystic liver disease (PLD) is a rare, benign inherited condition, frequently associated with polycystic disease of the kidney[1]. Unusually as liver failure appears, some patients may be highly symptomamtic due to the compression of liver enlargement or liver complications, therefore requiring treatment.

Highly symptomatic patients with PLD almost exhaust all conservative therapeutic options, and surgery is considered as the best possible procedure[2]. The purpose of this study was to evaluate the immediate surgical results and the short- and long-term outcome in patients treated with hepatic resection combined with cyst fenestration on the basis of our preliminary understanding and experience.

MATERIALS AND METHODS

From October 1995 to September 2002, we examined and operated on 6 women and 1 man with highly symptomatic PLD at our hospital. All operations were performed by a single surgeon (G.S.Yang). In this retrospective study, the medical records were reviewed. The mean interval from diagnosis of PLD to surgical treatment was 4.3 years, ranging from 2 to 11 years. Patient age ranged from 36-54 years, averaging 44.1 years. Specific symptoms included pain, massive abdominal distention, early satiety, regurgitation and supine dyspnea. Six patients were complicated with polycystic kidney disease but had no clinical evidence of abnormal renal function. Four patients had undergone percutaneous cyst aspiration with alcohol sclerotherapy before operation.

Preoperative workup included hematologic, hepatic and renal laboratory tests, and determinations of both respiratory and cardiovascular performance. CT scan was performed to delineate cyst distribution to assess portal vein patency and to serve as a baseline for follow-up comparison in each patient. No patient underwent angiography. The clinical features of these patients are summarized in Table 1.

Preoperative preparations of each patient were an overnight lavage bowel preparation and venous catheterization. The abdomen was explored through a bilateral subcostal incision. The hepatoduodenal ligament was exposed to provide access to a vascular clamping and identification of major vascular and biliary structures. The liver was mobilized by the division of hepatic peritoneal attachments, which was facilitated by sequential fenestration of accessible cysts according to the Lin technique[3]. Liver segments spared of cystic involvement were identified to define limits to resection. Non-anatomic segmental or lobar resection was executed due to cystic distortion of normal anatomy. Significant islands of functional liver parenchyma were preserved as many as possible. After resection of major cystic segments, extensive fenestration of residual cysts was performed by excision of the cyst walls. Finally, cyst cavities exposed to the peritoneum were fulgurated by argon beam coagulation (Bard Electromedical Systems, USA) in an attempt to ablate secretory epithelium and reduce postoperative peritoneal fluid losses. Cholecystectomy in conjunction with hepatectomy was performed in three patients. The hepatic resection bed was drained by two large closed suction drains. After operation, patients were sent to the surgical intensive care unit for close monitoring.

All patients were followed up through telephone calls or at clinic. Special data included hepatic and renal function, symptomatic relief, the patients' working capacity and CT scans.

RESULTS

The surgical outcome of our patients is presented in Table 2. The histologic examination showed von Meyenburg's complexes in all cases. There was no hospital death. All patients had ascites, the majority of them were successfully managed with diuretics and drainage within one week. The average duration of drainage was 11 d, with a range of 5-26 d. Drainage

Table 1 Clinical findings of patients with PLD

Patient no.	Age (yr)	Gender	Family history	Symptoms	Other involved organs	Previous treatment
1	48	Female	No siblings	Abdominal distention, regurgitation	None	None
2	41	Female	Father, two sisters, brother	Abdominal pain and distention	Kidney	PCA+AS (5 times)
3	54	Female	Sister	Early satiety, regurgitation	Kidney	PCA+AS (2 times)
4	45	Male	Sister	Abdominal pain and distention	Kidney	PCA+AS (2 times)
5	43	Female	Sister, brother, grandmother	Abdominal pain and distention, early satiety, dyspnea	Kidney	PCA+AS (2 times)
6	36	Female	Mother, sister	Abdominal pain and distention, early satiety, supine dyspnea	Kidney	None
7	42	Female	Father	Abdominal distention, regurgitation	Kidney	None

PCA+AS, Percutaneous cyst aspiration with alcohol sclerotherapy.

Table 2 Operative data, surgical outcome, and follow-up of patients with PLD

Patient no.	Hepatic resection	Blood loss (mL)	Cystic fluid aspirated (mL)	Days of hospital stay (after operation)	Postoperative complications	Symptomatic relief	Reduction in size of liver
1	III, IVb, V	400	1 500	21 (12)	Acites	Marked	Marked
2	II, III, V	800	3 000	25 (14)	Acites	Marked	Marked
3	Right semi hepatectomy, cholecystectomy	200	1 200	16 (8)	Mild acites	Marked	Marked
4	Right smi hepatectomy, cholecystectomy	300	1 800	24 (16)	Moderate acites	Marked	Marked
5	II, III, IVb	1 500	6 500	83 (29)	Severe acites, left pleural effusion	Moderate	Moderate
6	II, III, VI, VII	3 400	4 000	42 (16)	Severe acites, right pleural effusion	Marked	Marked
7	II, III, VI	300	2 500	27 (11)	Acites	Marked	Marked

Table 3 Review of the literature: mortality, morbidity and outcome of patients with PLD treated by combined hepatic resection an cystic fenestration

Authors	No. of patients	Mortality (no.)	Morbidity (no.)	Follow-up (mos)	No. of symptom-free patients
Armitage *et al*.[12] (1984)	1	0	0	12	1
Iwatsuki *et al*.[13] (1988)	6	0	0	-	-
Newman *et al*.[2] (1990)	9	1	6	17	7
Sanchez *et al*.[8] (1991)	2	0	0	48	2
Vauthey *et al*.[1] (1992)	5	0	4	14	5
Henne-Bruns *et al*.[14] (1993)	8	0	3	15	6
Soravia *et al*.[15] (1995)	10	1	2	68	6
Que *et al*.[16] (1995)	31	1	18	29	28
Vons *et al*.[17] (1998)	12	1	10	34	10
Johnson *et al*.[18] (1999)	1	0	0	18	1
Our series	7	0	7	0	6
Total	92	4 (4.3%)	50 (54.3%)		72 (78.3%)

was continued by needle aspiration in Patients 5 and 6 immediately after a two-week closed suction drainage. The change was an attempt to prevent ascites from infection. Furthermore Patient 5 required thoracentesis due to left pleural effusion.

All patients experienced relief of abdominal symptoms and had normal hepatic and renal laboratory tests. Abdominal girth was reduced markedly in six patients and moderately in one. Repeated abdominal CT scans in these patients failed to show significant development of cysts in the previously spared liver segments during a mean follow-up period of 20.4 mo, with a range from 2 to 86 mo. Indeed, as shown in Figure 1, CT scans confirmed post-resection hypertrophy of the spared liver and lack of clinically significant cyst progression.

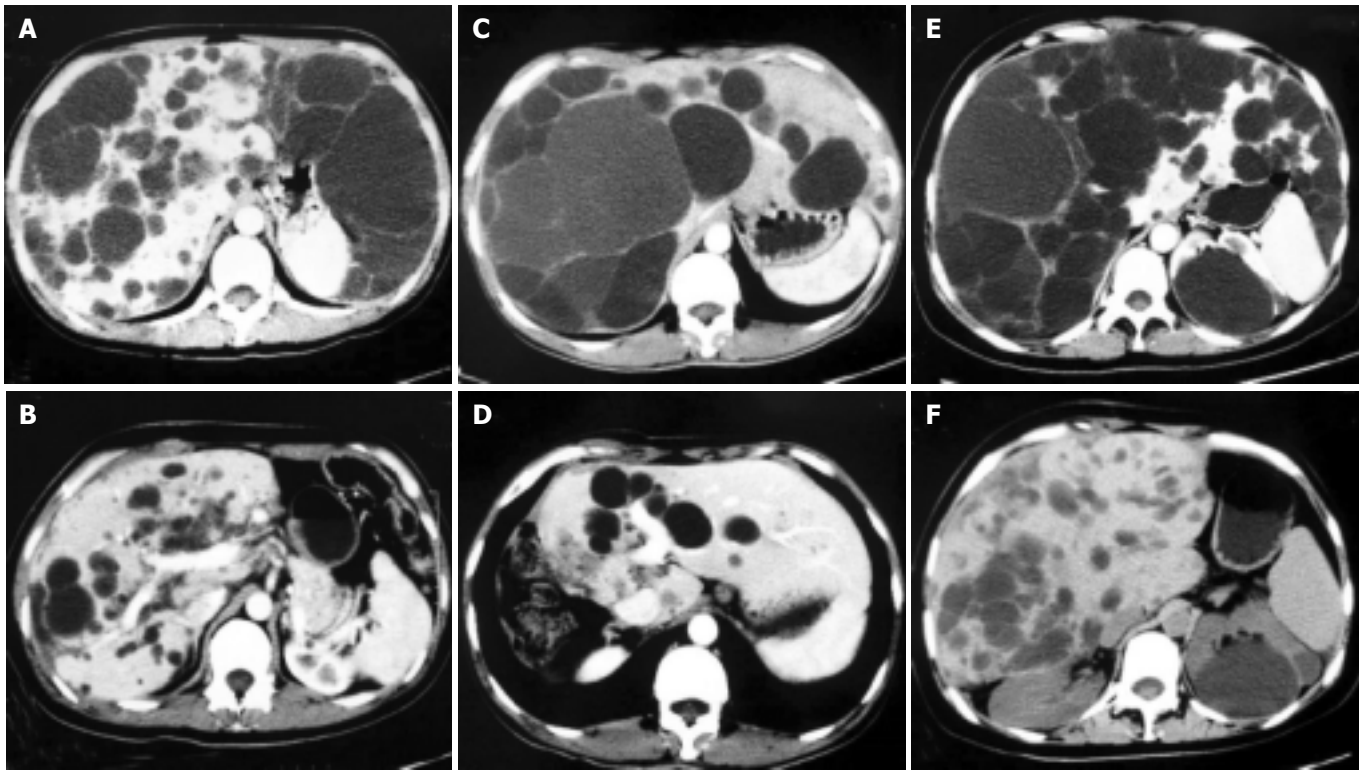

Figure 1 CT scans of livers in PLD patients before (left panels) and 3 mo (right panels) after operation, show that hypertrophy of spared liver and lack of clinically significant cyst progression, and that operative methods totally depend on cystic distribution in different patients, e.g., lateral segment removal from A to B, right semi-hepatectomy resection from C to D, whereas extensive fenestration of posterior and interior cysts from E to F.

DISCUSSION

PLD is a rare disorder usually associated with autosomal dominant polycystic kidney disease, with an increasing prevalence associated with age and female sex[4]. Symptoms arise from liver enlargement and compression of adjacent structures. Most symptomatic patients complained of increasing abdominal girth and a chronic dull abdominal pain. Liver enlargement may cause early satiety, weight loss, respiratory compromise, and physical disability. Complications such as ascites, esophageal varices, jaundice and hepatic failure, are rare[5-7].

A variety of treatment have been advocated for the few patients with incapacitating symptoms. Percutaneous aspiration with alcohol sclerotherapy seems valuable for solitary cysts but does not provide relief for patients with PLD because cyst collapse may not be sufficient[8]. Cyst fenestration with internal drainage into the peritoneal cavity, described by Lin in 1968, may fail to achieve long-term favorable outcome[9]. In practice, the efficacy of the Lin decompression for PLD is limited by extent of cysts, access to central cysts, postoperative walling off of cysts, and the rigid architecture of the fenestrated liver, which does not completely collapse[2]. Liver transplantation might be considered for patients with a "syndrome of lethal exhaustion" from PLD[7,10,11].

Armitage and Blumgart[12] described in 1984 a patient with PLD who underwent partial hepatic resection and cyst fenestration. This procedure allowed the excision of most prominent cysts with minimal resection of liver tissue; liver parenchyma was preserved despite the polycystic disease. Other reports have shown the feasibility of such an approach (Table 3)[1,2,8,12-18]. We reported a similar favorable outcome. During a mean follow-up of 20.4 mo, six of seven patients were symptom free. Herein, we emphasize that the result might be achieved by a careful selection of patients and by an experienced surgeon. The head surgeon of our surgical team has abundant experiences of more than 1400 hepatectomies for liver tumors during the same study period.

The postoperative morbidity rate in reviewed reports was 54.3%. The majority of immediately postoperative complications in our series were ascites and transient pleural effusion. Ascites was easily treated by diuretics and effective drainage or followed by needle aspiration. None of our patients had kidney failure requiring dialysis before or after operation. Ascites occurred in the patients with renal dysfunction may be refractory and even increased the chance to be infected or bring forth malnutrition[17]. The elucidation for the mechanism of hepatic cystic epithelium secretion may be a key to solve this problem[19].

The literature review showed an overall mortality rate of 4.3%. There is only one report that the cause of death was related to this procedure for patients with PLD[15]. Acute Budd-Chiari syndrome developed shortly after fenestration of posterior cyst, which provoked a liver collapse. In practice, to stitch on the dissected triangular, falciform and round ligaments easily kept the stability of the remnant liver, which was weakened by extensive fenestration of posterior and interior cysts. Patient 6 is a good example in our series. In patients 3 and 4, we found that the left lobes twisted towards right due to gravity after right semi-hepatectomy in operation. Connecting the falciform and round ligaments easily enabled the remnants fixed, avoiding occurrence of kinking. It may be unnecessary to avoid fenestration of posterior cysts and propose a frontal hepatectomy. Two patients in literature died from postoperative intracerebral bleeding, suggesting that evaluation of selected patients' cerebral vasculature to detect aneurismal disease is essential[2,16].

Our follow-up duration is limited and our preliminary study includes just seven selected patients. Therefore, the long-term benefit of combined hepatic resection and fenestration should be further evaluated. On the basis of our experience and review of the available literature, however, this approach

could be feasible, with an acceptable risk as well as a favorable long-term outcome at follow-up. To stitch the dissected hepatic ligaments could prevent the instable remnant liver from kinking and collapsing.

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