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<td>Author(s)</td>
<td>Furukawa, Masato; Furui, Junichiro; Otsubo, Mitsuji; Fujio, Toshiyuki; Miyazaki, Kunihisa; Yasaka, Takahiro; Shirahama, Satoshi; Kusano, Toshiomi</td>
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Classification of Primary Hepatolithiasis According to Morphology of the Liver, Especially Atrophy of Hepatic Parenchyma

Masato Furukawa, MD, PhD, Junichiro Furui, MD, PhD, Mitsuji Otsubo, MD, Toshiyuki Fujio, MD, Kunihisa Miyazaki, MD, Takahiro Yasaka, MD, Satoshi Shirahama, MD, PhD, and Toshiomi Kusano, MD, PhD

1) Department of Surgery, Nagasaki Chuo National Hospital
2) Department of Surgery, Kamigoto Hospital
3) Department of Medicine, Kamigoto Hospital
4) 1st Department of Surgery, Ryukyu University School of Medicine

One hundred twenty one patients with primary hepatolithiasis (PHL) was encountered for the last 7 years. In order to elucidate how to follow and treat PHL, PHL was classified into five types (I-V) on the basis of morphological findings obtained by CT. It depends mainly on the presence of atrophy of hepatic parenchyma (AHP) and dilatation of intrahepatic bile ducts (DIBD).

Methods of treatment were selected for each patient according to the classification.

Type I patients comprised of 43.8% in observation group. These patients were asymptomatic and just followed since neither AHP nor DIBD were observed.

The remaining patients in the observation group, operation was refused or contraindicated. Most of them were asymptomatic.

Surgical treatments were done on 47 patients (hepatic resection in 32 patients, surgical lithotomy in 10 and others in 5). In Type III, IV and V, hepatic resection was often performed to remove all of stones and bile stasis since improvement in hepatic function could not be expected in view of AHP.

All patients were followed-up 79 months or less.

In the surgically treated patients, recurrence was not observed. In observation group, no new symptoms developed during the follow-up period.

It was suggested, therefore, that the present classification of PHL was helpful in determining therapeutic strategy for PHL.

I. Introduction

Primary hepatolithiasis (PHL) is the most refractory and complicated condition among the various benign biliary tract diseases. There are still controversies on therapeutic choice for PHL.

Recently, exact conditions of the liver in PHL has been diagnosed by computed tomography (CT), which also contributed to detect asymptomatic PHL.

PHL was classified, on the basis of morphological findings obtained by CT, atrophy of hepatic parenchyma in particular.

In this study, we report that this classification of PHL is helpful in determining therapeutic strategy for PHL.

II. Patients and Methods

A total of 121 patients with PHL encountered at our hospital between June, 1985 and December, 1991.

Plain CT and CT cholangiography were performed using a Toshiba 70A initially with a slice width of 1.0cm and then with a width of 0.5-1.0cm at sites showing stones.

PHL was classified into five types (Type I-V) by CT.

The classification on depends mainly on the presence of atrophy of hepatic parenchyma (AHP) and dilatation of intrahepatic bile ducts (DIBD) (Table 1). (Fig. 1-5).

Type I comprised cases without AHP or DIBD, in which small stones were found in the periphery of intrahepatic bile ducts (53 patients).

Type II comprised cases without AHP but with DIBD (12 patients).

Type III comprised cases with AHP but without DIBD (8 patients).

Type IV comprised cases with moderate AHP and DIBD (25 patients).

Type V comprised cases with severe AHP and marked DIBD, in which major and peripheral bile ducts were filled with large stones (23 patients).

The intrahepatic type confined to the liver (type I) was observed in 90 patients, and the intrahepatic predominant type showing more intrahepatic than extrahepatic stones (type IE) in 31, which classification has been followed the rules proposed by the Research Group for the Study of Damage to Intrahepatic Bile Ducts. Intrahepatic stones were located in the right lobe alone in 49 patients (40.5%), left lobe alone in 62 (51.2%), and both lobes in the other 10 (8.3%) (Table 1).

Methods of treatment for PHL were selected for each patient according to this classification (Table 2).

In Type I, most of these patients were asymptomatic and they were not treated but just followed, since neither AHP
### Table 1. Classification of PHL and Patient Distribution according to Findings of CT

<table>
<thead>
<tr>
<th>Classification</th>
<th>APH</th>
<th>DIBD</th>
<th>No. of patients</th>
<th>Location of stones</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>I</td>
</tr>
<tr>
<td>I (-) (-)</td>
<td>53</td>
<td>51</td>
<td>2</td>
<td>38</td>
</tr>
<tr>
<td>I I (-) (+)</td>
<td>12</td>
<td>7</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>III (+) (-)</td>
<td>8</td>
<td>6</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td>IV (+) (+)</td>
<td>25</td>
<td>12</td>
<td>13</td>
<td>0</td>
</tr>
<tr>
<td>V (++) (+-)</td>
<td>23</td>
<td>14</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>121</td>
<td>90</td>
<td>31</td>
<td>49</td>
</tr>
</tbody>
</table>

APH: Atrophy of Hepatic Parenchyma  
DIBD: Dilatation of Intrahepatic Bile Ducts  
I: Intrahepatic bile ducts  
IE: Intrahepatic > Extrahepatic bile ducts  
R: Right lobe of the liver  
RL: Right and Left lobes of the liver  
L: Left lobe of the liver

**Fig. 1.** Classification of PHL, Type I  
Type I: APH(-), DIBD(-), S.T. 50 y.o. Male  
Small stones were found in posterior segment of liver. But neither APH nor DIBD were observed.

**Fig. 2.** Classification of PHL, Type II  
Type II: APH(+), DIBD(+), I.T. 39 y.o. Male  
In plain CT, stones were observed in anterior segment. In DIC-CT, anterior branch of bile duct was dilated, but parenchyma was not atrophic.

**Fig. 3.** Classification of PHL, Type III  
Type III: APH(+), DIBD(-), S.F. 50 y.o. Female  
Stones were observed in anterior segment. Hepatic parenchyma of anterior segment was atrophic and posterior and medial segment were hypertrophic. Anterior branch of bile duct was not dilated.

**Fig. 4.** Classification of PHL, Type IV  
Type IV: APH(+), DIBD(+), F.Y. 62 y.o. Female  
Stones were observed in lateral segment. Hepatic segment was atrophic, and lateral branch of bile duct was dilated.
nor DIBD were observed.

In Type II, APH was not found, so that improvement of liver function could be expected by removal stones and bile stasis. Of the patients with type II, 2 showing stones only in the left lobe were treated by left lateral segmentectomy for complete removal of stones, 2 by choledochojejunostomy for extrahepatic bile duct dilatation, and the other 4 by lithotripsy under cholangioscopic guidance via an external bile duct fistula and percutaneous transhepatic cholangioscopic lithotomy (PTCSL).

In Type III, IV and V showing liver atrophy, hepatic resection was performed in 30 patients, to remove all of stones since improvement of hepatic function could not be expected in view of hepatic parenchymal atrophy, and the other 8 in whom the function of the remaining liver was decreased due to multiple operations were treated by liver-preserving operation.

Thus, hepatectomy was performed in 32 of the 47 patients treated (68.1%), liver-preserving operation in 10, and endoscopic removal of stones in the other 5. Stones in intrahepatic bile ducts were completely removed by surgical and/or endoscopic procedures.

III. Results

The mean observation period was 39.3 months in the untreated patients, 28 months in the patients treated by PTCSL, 34.6 months in those treated by liver-preserving operation and 34.5 months in those treated by hepatectomy (Table 3).

Table 2. Choice of Treatment for PHL according to the Classification

<table>
<thead>
<tr>
<th>Type</th>
<th>No. of patients</th>
<th>Observation only</th>
<th>Procedures of treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td>PTCSL</td>
</tr>
<tr>
<td>I</td>
<td>53</td>
<td>53</td>
<td>0</td>
</tr>
<tr>
<td>II</td>
<td>12</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>III</td>
<td>8</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>IV</td>
<td>25</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>V</td>
<td>23</td>
<td>6</td>
<td>0</td>
</tr>
<tr>
<td>Total</td>
<td>121</td>
<td>74</td>
<td>5</td>
</tr>
</tbody>
</table>

PTCSL: Percutaneous Transhepatic Cholangioscopic Lithotripsy

Table 3. Results of the Patients with PHL

<table>
<thead>
<tr>
<th>Choice of treatments</th>
<th>No. of patients</th>
<th>Follow-up periods (mos)</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Observation only</td>
<td>74</td>
<td>66.5 (5.0–79.0)</td>
<td>Cholangitis (4) Death (6*)</td>
</tr>
<tr>
<td>PTCSL</td>
<td>5</td>
<td>39.8 (4.0–63.0)</td>
<td>Bile leak (2) Death (1***)</td>
</tr>
<tr>
<td>Hepatectomy (-)</td>
<td>10</td>
<td>50.8 (4.0–75.0)</td>
<td>Cholangitis (1) Death (2****)</td>
</tr>
<tr>
<td>Hepatectomy (+)</td>
<td>32</td>
<td>49.6 (3.0–73.0)</td>
<td>Death (2****)</td>
</tr>
</tbody>
</table>

(): No. of Patients
*: Cause of death
**: Cause of death
***: Cause of death
****: Cause of death

Bile duct ca.(1), Renal ca.(1), Lung ca.(1), Adult T-cell Leucamia (1), Esophageal ca.(1), Heart failure (1)

Cholangitis (1), Death (1), Hepatic failure (1)

Intrahepatic bile duct ca.(1), Gall bladder ca.(1)
As postoperative complications in the treated group, a transient bile leakage was observed in 2 patients and cholangitis in 1 treated by liver-preserving operation with additional operation on the bile duct. Causes of death were intrahepatic bile duct cancer in 2 patients, gallbladder cancer in 1 and suicide in 1, but all the others had favorable postoperative courses and resumed ordinary lives. Recurrence was not observed.

In the group observed without treatment, 1 each died of bile duct cancer, renal cancer, lung cancer, esophageal cancer, adult T-cell leucemia, hepatic failure and renal failure. Four patients developed symptoms of gallstones due to falling of stones to the extrahepatic bile duct or other factors and underwent removal of stones.

IV. Discussion

PHL was considered to be intractable because localization of stones was difficult to diagnosis, and their complete removal was impossible. However, recent advances in imaging techniques, especially CT and US, have allowed detection of many cases of asymptomatic intrahepatic stones (especially, peripheral intrahepatic bile duct type). Various classifications have been proposed for PHL, mostly according to the site of stones. We classified PHL into five according to the morphology of the liver, especially liver atrophy, and selected different treatment methods according to the disease types. It was thought that Type I was the early stage of PHL, Type V the late stage.

Patients with type I were observed without treatment because most of them were asymptomatic and showed no AHP or DIBD.

In patients with type II who showed DIBD but not AHP, the liver was preserved, and stones were completely removed by methods such as PTCSL.

Patients with type III, IV, or V showing AHP, especially those with stones in one lobe alone were actively treated by hepatectomy.

Hepatectomy for intrahepatic stones has been considered to be indicated in patients with liver atrophy and stenosis or cystic dilatation of the intrahepatic bile duct. However, with recent advances in endoscopic removal of stones and the development of non-operative techniques such as release of bile duct stenosis, the treatment modalities for intrahepatic stones including indications for hepatectomy have been changed.

In case 1 showing AHP, fibrosis was observed in the hepatic parenchyma, and hepatocytes were rarely present (Fig. 6). The viability of the liver was lost. It was considered that complete removal of stones may only result in the formation of a diverticulum in which bile, produced another sites and regurgitated, stagnates. Therefore, hepatectomy was performed. CT clearly demonstrated atrophy of hepatic parenchyma and was very useful for classifying PHL.

On the other hand, bile duct dilatation was accurately visualized by CT combined with DIC. As shown in case 2, such dilatation sometimes improved after complete removal of stones (Fig. 7). This finding was consistent with the report by Nimura that suspected stenosis disappeared, and suspected bile duct dilatation improved to the normal diameter following removal of intrahepatic stones. Since it was difficult to determine whether bile duct dilatation is reversible or not, it was excluded from indications for hepatectomy.

The results of our treatment based on the above treatment criteria are shown in Table 3. Postoperative cholangitis was observed only in 1 patient. In addition, stones did not recur during the observation period (maximum, 79 months) in any patient.

These results suggest that our classification of PHL is very useful for determining therapeutic approaches. However, malignant tumors in the biliary tract have been re-
ported to often complicate intrahepatic stones. In this series, malignant tumors (gallbladder cancer and bile duct cancer) were also observed in 3 patients. The possible presence of malignant tumors should be taken into consideration during observation of clinical course.

V. Conclusion

One hundred twenty one patients with PHL were classified into five types, detecting by CT according to the morphology of the liver, especially liver atrophy, and discussed appropriate treatment methods for each PHL type.

In Type I, most of these patients were asymptomatic and they should not be treated but just followed, since neither APH nor DIHBD were observed. In Type II, APH was not found, and they should be treated by liver-preserving operation, since improvement of liver function could be expected by removal stones and bile stasis.

In type III, IV, or V showing liver atrophy, hepatectomy should be primarily performed.

It was suggested, therefore, that the present classification of PHL was helpful in determining therapeutic strategy for PHL, although careful attention should always be paid to the association of malignant lesions during the follow-up of this disease.

(An abstract of this paper was presented in the 3rd meeting of the World Association of the Hepatic, Pancreatic and Biliary Surgery.)

References