Early hepatocellular carcinoma associated with fibrocystic liver disease in a 10 years old child -- a case report

Abstract

Fibrocystic liver–kidney disease is caused by a group of rare and genetically diverse disorders that are associated with kidney cysts or dysplasia and ductal plate malformation in the liver. There is a wide range of presentation for fibrocystic liver–kidney disease from relatively asymptomatic to lethality in the perinatal period due to pulmonary hypoplasia. The liver disease has characteristic pathologic findings of dilated bile ducts accompanied by dense periportal fibrosis that results from the disruption of the normally developing ductal plate in utero. There have been several reports of liver neoplasias arising in hepatobiliary fibrocystic diseases. However, most of them were cholangiocarcinomas and cases involving hepatocellular carcinoma (HCC) are rare and all reported cases are related with adults. Because there are no reports in the literature regarding the development of hepatocellular carcinoma within the context of fibrocystic liver disease in children, we describe herein one case of this association.

Keywords: ductal plate, hepatobiliary, fibrocystic, neoplasias, cholangiocarcinomas, periportal, transplantation, Porto systemic shunt, sclerotherapy, capsular area, hepatorenal fibrocystic, caroli syndrome

Introduction

Affected patients often have variable presentation and progression in their liver and kidney diseases, with patients often having more severe involvement of one organ. Some patients may develop end stage kidney disease, while others may develop complications from portal hypertension or recurrent cholangitis requiring liver transplantation. A small subset of patients requires transplantation in both organs, either sequentially or in combination. Because there are no reports in the literature regarding the development of hepatocellular carcinoma within the context of fibrocystic liver disease in children, we describe herein one case of this association.

Case report

A 10 years old girl with history of repeated gastrointestinal bleeding underwent multiple times for banding and sclerotherapy and also had history of Porto systemic shunt without any significant benefit referred to us as a case of Fibrocystic liver–kidney disease with decompensated liver disease for Liver Transplantation. Child under went Living donor liver transplantation the explanted liver showed.

I. Gross pathology

Explant liver weighed 838 g and measuring 21 x 13 x 8.5 cm with attached gallbladder measuring 7 x 3 x 0.2 cm (in wall thickness). The external surface is covered by multiple white nodules ranging in size from 0.4 to 1 cm. Serial slicing reveals an ill-defined yellow soft lesion (4 x 2.5 x 2.5 cm) localized in the sub capsular area of the left lobe (segment 4). The rest of the cut surface is green and nodular (cirrhotic).

II. Histopathology

Microscopy from largest nodule consistent with early hepatocellular carcinoma. The rest of liver is cirrhotic the morphology consistent with fibrocystic disease of liver (Figure 1&2).

Discussion

The hepatorenal fibrocystic diseases (HRFCD) are ciliopathies characterized by renal fibrocystic degeneration and abnormalities of the Porto biliary system, such as congenital hepatic fibrosis and...
caroli syndrome. They are suspected in patients who present with both renal and liver involvement. Progressions of the renal and liver involvement are independent. All patients known for ARPKD or ADPKD have some degree of liver involvement. While some will be completely asymptomatic (only micro-scopic changes), others will present with liver abnormalities and portal hypertension. Recently, it has been shown that hepatobiliary fibro polycystic diseases sometimes progress to malignant neoplasia. An association with cholangiocarcinoma was established in most cases, and HCC combined with hepatobiliary fibro polycystic diseases has rarely been reported (Table 1).

In a review of liver neoplasms associated with hepatobiliary fibro polycystic diseases, only four HCC cases without known risk factors were associated with BDH/CHF/Caroli’s disease. Bauman ME et al. reported a case of HCC arising in CHF. The case was incidentally-found multifocal HCC in an explanted liver with CHF. The renal lesion lacked the multiple large cortical cysts of ADPKD and yet cerebral and multiple splenic artery aneurysms were present. Heinke T et al. also reported two other HCC associated with BDH. Iijima M et al. reported HCC associated with Caroli’s disease recently. We report herein a case of CHF with HCC without other risk factors for HCC such as hepatitis B virus, hepatitis C virus, alcohol, and nonalcoholic steatohepatitis (NASH). Our case had multiple well-differentiated HCC, indicating multifocal development of HCC rather than intrahepatic metastasis. Therefore, it is possible that livers of hepatobiliary fibropolycystic diseases bearing HCC are in a hypercarcinogenic state. The prevalence of CHF is not clearly defined; however, it was estimated to be one in 10,000 to 20,000 based on the data of various specific ciliopathies associated with CHF. Nagano Y et al. reported that MRI was suitable for imaging diagnosis of BDH. Through the development and spread of imaging modalities including MRI, the likelihood of diagnosing hepatobiliary fibro polycystic diseases is increasing, and more patients with hepatobiliary fibro polycystic diseases will be found. Hence, the actual number of HCC arising in hepatobiliary fibro polycystic diseases might be higher.

### Table 1 List of hepatocellular carcinoma (HCC) with hepatobiliary fibrocystic diseases

<table>
<thead>
<tr>
<th>Reference</th>
<th>Year</th>
<th>Hepatic lesion</th>
<th>Grade of differentiation</th>
<th>Sex</th>
<th>Age(years)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bauman et al.</td>
<td>1994</td>
<td>CHF</td>
<td>well differentiated HCC</td>
<td>M</td>
<td>31</td>
</tr>
<tr>
<td>Heinke et al.</td>
<td>2008</td>
<td>BDH</td>
<td>well differentiated HCC</td>
<td>F</td>
<td>19</td>
</tr>
<tr>
<td>Heinke et al.</td>
<td>2008</td>
<td>BDH</td>
<td>Moderately differentiated HCC</td>
<td>M</td>
<td>39</td>
</tr>
<tr>
<td>Iijima M et al.</td>
<td>2010</td>
<td>Caroli’s disease</td>
<td>Moderately differentiated HCC</td>
<td>M</td>
<td>29</td>
</tr>
<tr>
<td>Present case</td>
<td>2010</td>
<td>CHF</td>
<td>well differentiated HCC</td>
<td>F</td>
<td>51</td>
</tr>
</tbody>
</table>

BDH, bile duct hamatoma; CHF, Congential Hepatic fibrosis; F, Female; M, Male

### Conclusion

To our knowledge, this is the first reported case of Hepatocellular carcinoma associated with fibrocystic liver disease pediatric age group. This is a rare case of HCC associated with fibrocystic liver disease. When diagnosing Fibrocystic liver disease without known risk factors, the presence of HCC must be considered and vice versa.

### Acknowledgments

None.

### Conflicts of interest

There is no conflict of interest.

### References


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