Discussion Regarding the Method of Screening for Hepatocellular Carcinoma in Glycogen Storage Disease Type 1a

Key words: hepatocellular carcinoma, abdominal imaging, glycogen storage disease type 1a

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The Authors Reply  
We thank Dr. Bashir et al. for their comments on our case report. Their comment regarding the strategy for screening and diagnosis of hepatocellular carcinoma (HCC) in patients with glycogen storage disease is an important one.

For the detection of HCC, contrast-enhanced computed tomography, gadolinium-ethoxybenzyl-diethylenetriamine penta-acetic acid magnetic resonance imaging (EOB-MRI), and contrast-enhanced ultrasonography (CE-US) are widely used.

We agree that EOB-MRI is an excellent diagnostic modality for imaging hepatic tumors. It provides results that are not operator dependent, but the accuracy of diagnosis does depend on the quality of the MRI equipment. In addition, an MRI machine is expensive and wide-bore magnets are installed only in a limited number of clinical institutions. These social conditions affect the situation as to whether or not patients can be examined with EOB-MRI. On the other hand, CE-US results depend on the skill of the operator, and it is difficult to detect deep tumors with CE-US. However, CE-US is a non-invasive, widely available and low-cost technique. It is sensitive for the diagnosis of HCC, unless the tumor is located in a deep position. In the present case, HCC in S4 was diagnosed with CE-US, but not with EOB-MRI. In the present case, many space-occupying lesions (SOL) were detected, and it was difficult to distinguish these SOL from the hepatic adenoma. On CE-US with perflubutane, only the tumor in the 4th segment was detected in the late phase. On the other hand, in the hepatic phase of EOB-MRI, the tumor in the 4th segment was not different from the other tumors. These results suggest that if a SOL is located in a site that is easy to observe by CE-US, then CE-US can provide a more accurate diagnosis than EOB-MRI.

The advantage of CE-US using perflubutane is the ability to undertake continuous and repeated scans in the late phase without breaking the perflubutane microbubbles. Perflubutane is different than other US contrast agents, and can be used to provide a highly accurate diagnosis of HCC with CE-US.

The process of diagnosis of HCC with glycogen storage disease type 1a (GSD-1a) depends on the above-mentioned social conditions. If the patient can be examined with EOB-MRI and CE-US with perflubutane with a skilled operator, we think it is better to examine alternately CE-US with EOB-MRI.

There is a shortage of information regarding the findings of HCC and hepatic adenomas in patients with GSD-1a examined by various abdominal imaging modalities. Experience with more cases of HCC with both CE-US with perflubutane and EOB-MRI with GSD-1a is needed.

The authors state that they have no Conflict of Interest (COI).

Hironori Ochi¹, Atsushi Hiraoka² and Kojiro Michitaka²

¹Department of Gastroenterology and Metabolism, Ehime University Graduate School of Medicine, Japan and ²Department of Gastroenterology, Ehime Prefectural Central Hospital, Japan

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Correspondence to Dr. Kojiro Michitaka, c-kmichitaka@eph.pref.ehime.jp

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