Clinics and Research in Hepatology and Gastroenterology (2019) 43, e7—e8

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LETTER TO THE EDITOR

Answer to challenging issues in the management of Wilson's disease

Dear Sir,

Thank you for your relevant comments concerning the challenging issues in the management of Wilson disease (WD) [1]. We agree with you that acute liver failure or end stage liver disease could reveal WD and that diagnosis could be extremely difficult. In these specific situations, REC (ratio exchangeable copper/serum copper) is of valuable interest as shown in our publications. As you outlined it, data from the first paper is scant as it concerned only sixteen patients [2] but following publications have enriched the data with confirmation of the interest of the REC. In an animal model of WD, authors tested and confirmed the validity of this new biomarker throughout the course of the liver disease, from a presymptomatic stage to the beginning of chronic liver failure [3]. Two recent papers studied 57 de novo and untreated WD patients and confirmed that the REC was elevated (> 18.5%) in all patients at diagnosis while REC was below 16.3% in other hepatic diseases studied [4,5]. In view of the results, we think that this rapid and reliable biological test should be largely available in hospitals that take care of patients with liver and neurological diseases, this especially as the technique is easy to set up.

As you mentioned, outcome of neurologic manifestation after LT for WD is difficult to anticipate. In our experience, no worsening of neurological symptoms was described after LT for neurologic indication but the situation is different in LT for liver indication as reported in your experience and in a few cases in the literature. Central pontine myelinolysis (CPM) is a well-known complication in patients with neurological symptoms receiving LT because of liver failure. Deterioration of pre-existing neurological symptoms (encephalopathy and CPM excluded) after LT have also been reported in 3/141 patients transplanted because of liver disease [6—8]. Another hypothesis of the rapid neurological worsening after LT for liver indication is the development of extensive liver tissue necrosis during the peri-transplant period, with subsequent copper release to the circulation and the brain causing irreversible neuronal damage [9]. As worsening of neurological symptoms and new onset of neurological symptoms are not well understood, one may recommend to assess systematically the neurological status before LT with the specific UWDRS score and a brain MRI.

Concerning the risk of hepatocellular carcinoma (HCC), we agree that this situation is rare in WD but need to be detected. In the French registry (604 patients), 1.2% developed a liver carcinoma during the course of their disease. In the cited paper, HCC represented 14% of the indication of liver transplantation in a liver surgery center that takes care of Wilsonian patients.

Disclosure of interest

The authors declare that they have no competing interest.

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Available online 27 August 2018