Acute-Onset Autoimmune Hepatitis

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Autoimmune hepatitis (AIH) presents as a chronic inflammatory liver disease which responds to steroid therapy. The histological feature of AIH shows multilobular necrosis, interface hepatitis and mononuclear cell infiltration with predominantly plasma cells in the portal tract. The presence of autoantibodies, hypergammaglobulinemia, and elevated serum transaminase is helpful for diagnosis of AIH. An AIH scoring system has been proposed by the International Autoimmune Hepatitis Group (1). AIH scoring system has been useful for the diagnosis of typical AIH, however, some patients who do not fulfill the criteria present atypical features. One such type of patient has acute-onset AIH, the other type has clinicopathological features of both AIH and primary biliary cirrhosis or primary sclerosing cholangitis. Furthermore, acute-onset AIH includes two types of AIH. One is clinically diagnosed “acute-onset AIH”, which is pathologically shown as an acute exacerbation with chronic hepatitis. The other type is truly “acute-onset AIH” which shows clinically and histologically acute hepatitis.

Lefkowitch et al first reported two AIH cases presenting histologically acute hepatitis (2). Burgart et al reported that only one of 26 recent-onset AIH patients showed lobular hepatitis without portal inflammation (3). In a Japanese nationwide survey study, 5.6% of patients with AIH were found to have a feature of acute hepatitis upon histological examination (4). The histological characteristics of acute-onset AIH revealed centrilobular necrosis, none or mild inflammatory infiltration in the portal area, and no portal fibrosis (5, 6). The serum gammaglobulin or IgG concentrations were often lower than those of chronic hepatitis (5). These data may gradually increase during the clinical course. The relationship between clinical course and clinical manifestation are proposed as shown in Fig. 1. It is difficult to diagnose these patients based on the international criteria, because they assume chronic hepatitis. There have been reports stating that a response to corticosteroid therapy should help to establish the diagnosis of acute-onset AIH.

In the prior issue of Internal Medicine, Takahashi et al report a case of acute-onset autoimmune hepatitis as fulminant hepatic failure (7). This case was rescued by living donor-liver transplantation. It was reported that fulminant hepatic failure is due to AIH in 4-10% of patients. It could be that acute-onset AIH results in subacute or late-onset hepatic failure rather than acute hepatic failure. These patients often have resistance to corticosteroid therapy including pulse therapy (8). It was reported that there is a 20-46% survival rate without liver transplantation (9, 10). However, the prognosis remains unclear. Liver transplantation is one of the therapeutic options for these patients with a fatal outcome. A case of severe acute hepatitis of AIH was also treated with liver transplantation, when corticosteroid therapy was not effective (11).

The clinical presentation of acute-onset AIH is often non-specific and diverse. For the diagnosis of AIH, virus infection, drug-induced hepatitis, and other causes need to be
ruled out. Patients sometimes have some drugs including dietary supplements. It is difficult to distinguish acute-onset AIH from drug-induced hepatitis, because their clinical features and laboratory data are similar. In this report, the patient had taken weight-loss supplements. We should carefully record the drug history and exclude the possibility of drug-induced hepatitis (5).

The period of initial symptoms to the diagnosis of fulminant or severe acute hepatitis are occasionally longer than that of acute hepatitis (Fig. 1) (12). These patients may indicate that there are several weeks between disease onset and hospitalization. As a result, liver injury has already been in progress, and their prognosis may be poor. It is possible that the prognosis is improved by starting corticosteroid therapy early. For early diagnosis of patients, we need to develop the network system for severe hepatitis in the hospital area.

Some acute-onset AIH patients have severe liver injury and there is an urgent need for an immediate diagnosis in order to design an appropriate management scheme. The histological examination of the liver is necessary for early diagnosis. We should pay attention to the possibility that AIH can cause acute hepatitis and fulminant hepatic failure.

References


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