Commentary

Liver Diseases and Autoimmunity

Harry Hua-Xiang Xia

Novartis Pharmaceuticals Corporation, East Hanover, NJ, USA

Based on the principal clinical and pathological features, autoimmune diseases can be broadly divided into localized (involving only a single organ or tissue) and systemic (involving multiple organs). The liver is a lymphoid organ that not only participates in the immune response, but also a target of autoimmune reactions. Autoimmune hepatitis (AIH), primary biliary cirrhosis (PBC) and primary sclerosing cholangitis (PSC) are three major types of autoimmune diseases localized in the liver or autoimmune liver diseases, whereas granulomatous diseases such as sarcoidosis and connective tissue diseases such as systemic lupus erythematosus, rheumatoid arthritis and primary Sjögren syndrome (PSS) are the major systemic autoimmune diseases with liver involvement.

Over the past decade, advances have been achieved in the diagnosis and treatment of autoimmune liver diseases and liver involvement in systemic autoimmune diseases.[1–5] Accordingly, in this issue of *Journal of Clinical Translational Hepatology*, the aim of the review articles is to update the knowledge and current management of autoimmune liver diseases and liver involvement in systemic autoimmune diseases.

AIH is a chronic inflammatory liver disease of unknown cause, with an estimated prevalence of 0.5–1.0 per 100,000; it is more common in women than in men. There have been extensive studies on this important disease, and remission of the disease is now achievable by immunosuppression; however, the prognosis is very poor if the patients are left untreated. In this issue, Kapila *et al.* present the clinical manifestations of AIH, update pathogenesis of liver-directed immune injury and new concepts in the understanding of immune tolerance, and finally provide insight into the development of novel therapeutic approaches.[6]

Sarcoidosis is a systemic, granulomatous disease that usually affects multiple organs including the liver. The estimated prevalence of sarcoidosis is 2–60 per 100,000. So far, little is unknown about its etiology. Liver involvement in sarcoidosis is very common. In this issue, Tadros *et al.* introduce the epidemiology and clinical spectrum of hepatic sarcoidosis of the disease and explore the underlying mechanism for liver injury in hepatic sarcoidosis.[7] Moreover, they provide state-of-the-art knowledge on the diagnosis, differential diagnosis (with PBC and PSC) and treatment of the disease.

PSS is a systemic autoimmune disorder with secretory gland dysfunction. The estimated prevalence is 0.1–3.3%, with significant female preponderance, and it is reportedly associated with increased risk of cancers. Liver involvement is one of the commonly reported extraglandular manifestations of PSS, and two main causes of liver disease, namely chronic viral infections and autoimmune liver diseases contribute to liver involvement in PSS. Diagnosis of the causes of liver involvement in PSS is very important but challenging. In this issue, Briton-Zerón *et al.*[8] specifically describe the causes, and provide the diagnostic strategy accordingly.

There is no doubt that the new data provided in this issue will enrich our understanding and improve clinical treatment of autoimmune liver diseases and liver involvement in systemic autoimmune diseases. This is consistent with the objective of *Journal of Clinical Translational Hepatology* to deliver recent advances in pathogenetic mechanisms, and developments in basic, translational and clinical studies.

Conflict of interest

None.

References

[1] Zachou K, Muratori P, Koukoulis GK, Granito A, Gatselis N, Fabbri A, *et al*. Review article: autoimmune hepatitis – current management and challenges. Aliment Pharmacol Ther 2013;38:887–913.
[2] Piga M, Vacca A, Porru G, Cauli A, Mathieu A. Liver involvement in systemic lupus erythematosus: incidence, clinical course and outcome of lupus hepatitis. Clin Exp Rheumatol 2010;28:504–510.
[3] Brito-Zerón P, Sisó-Almirall A, Bové A, Kostov BA, Ramos-Casals M. Primary Sjögren syndrome: an update on current pharmacotherapy options and future directions. Expert Opin Pharmacother 2013;14:279–289.
[4] Selmi C, De Santis M, Gershwin ME. Liver involvement in subjects with rheumatic disease. Arthritis Res Ther 2011;13:1–6.
[5] De Santis M, Crotti C, Selmi C. Liver abnormalities in connective tissue diseases. Best Pract Res Clin Gastroenterol 2013;27:543–551.
[6] Brito-Zerón P, Retamozo S, Bové A, Kostov BA, Sisó A. Ramos-Casals M. Diagnosis of Liver Involvement in Primary Sjögren Syndrome. J Clin Transl Heaptol 2013;1:94–102.
[7] Tadros M, Forouhar F, Y. Wu GY. Hepatic sarcoidosis. J Clin Transl Heaptol 2013;1:87–93.
[8] Kapila N, Higa JT, Longhi MS, Robson SC. Autoimmune hepatitis: clinical review with insights into the purinergic mechanism of disease. J Clin Transl Heaptol 2013;1:79–86.

☆ DOI of original article: 10.14218/JCTH.2013.00026.
Correspondence to: Dr. Harry Hua-Xiang Xia, Novartis Pharmaceuticals Corporation, One Health Plaza, East Hanover, NJ 07936, USA. Email: harry.xia@novartis.com