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Autoimmune liver disease - are there spectra that we do not know?

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Abstract

ABSTRACT: Autoimmune liver diseases (AILDs) are common leading causes for liver cirrhosis and terminal stage of liver disease. They have variable prevalence among patients with liver disease and have two major clinical and biochemical presentations. Autoimmune hepatitis (AIH) is the typical example of hepatocellular AILD, but it can also be presented under a cholestatic pattern. AIH has a scoring diagnostic system and respond in most cases to the treatment with prednisolone and azathioprine. Primary biliary cirrhosis (PBC) is the second most common AILD, with a cholestatic presentation and characterized by positive antimitochondrial antibody (AMA). It has an excellent response and long term outcome with the administration of ursodeoxycholic acid (UDCA). Another AILD that is thought to be a variant of PBC is the autoimmune cholangitis, being a disease that has biochemical and histological features similar to PBC; but the AMA is negative. Primary sclerosing cholangitis (PSC) is a rare entity of AILD that has a cholestatic presentation and respond poorly to the treatment, with the ultimate progression to advance liver cirrhosis in most patients. Other forms of AILD include the overlap syndromes (OS), which are diseases with mixed immunological and histological patterns of two AILD; the most commonly recognized one is AIH-PBC overlap (AIH-PSC overlap is less common). The treatment of OS involves the trial of UDCA and different immunosuppressants. Here we present three case reports of unusual forms of chronic liver diseases that most likely represent AILD. The first two patients had a cholestatic picture, whereas the third one had a hepatocellular picture at presentation. We discussed their biochemical, immunological and histological features as well as their response to treatment and their outcomes. Then, we compared them with other forms of AILD.