## **Pediatric Rheumatology**



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## Autoimmune hepatitis or systemic lupus erythematosus? A diagnostic dilemma

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Autoimmune hepatitis (AIH) is a chronic inflammatory liver disease of unknown etiology associated with circulating autoantibodies and hypergammaglobulinemia. Patients with AIH occasionally suffer from other autoimmune diseases. Similarly, systemic lupus erythematosus (SLE) is an autoimmune disorder affecting multiple organs and coexisting with other autoimmune diseases. Although the liver is not a major target for damage in SLE, clinical and biochemical evidence of liver abnormalities are common. However, the difference between the hepatic involvement of SLE and autoimmune hepatitis has not been clearly defined due to similarities in clinical and biochemical features.

We report a 16-year-old girl with an overlap syndrome involving AIH and SLE. She presented with failure to thrive, jaundice, non-erosive arthritis, and oral aftous lesions accompanied by hyperbilirubinemia, elevated ALT, leukopenia, hypocomplementemia, and direct Coombs positivity. Serologic tests showed that she was positive for ANA and anti-Ro antibody as well as anti-gliadin IgG and anti-endomisium IgA. Liver biopsy showed portal and periportal hepatitis with lymphocytic infiltration and piecemeal necrosis. International autoimmune hepatitis score demonstrated definite AIH. She had proteinuria at the follow-up (77 mg/m<sup>2</sup>/hour). Autoantibodies along with clinical findings fulfilled the criteria for both AIH and SLE. Arthritis and proteinuria with hematological findings attributable to SLE improved with high dose treatment with corticosteroids and azathioprine; however, remission of the liver disease could not be achieved.

In conclusion, this case suggested that AIH and SLE might be indistinguishable from each other.