

## **Abstract**

Systemic lupus erythematosus may first present in pregnancy as pregnancy causes exacerbation of SLE. SLE itself causing liver disease is rare. However it can overlap with other autoimmune liver diseases. SLE is known to complicate with Macrophage activation syndrome which is potentially fatal.

A previously healthy 31 years old of lady presented in her second pregnancy at 11 weeks of gestation with fever for 3 days. There were no other systemic symptoms . Ultimately she was diagnosed with Systemic lupus erythematosus with Autoimmune Cholangitis. Her disease course was further complicated with Macrophage activation syndrome (MAS). Her disease course was complicated with multiple diagnostic and therapeutic dilemmas.

This is an unusual presentation of SLE in pregnancy. MAS should be suspected in patients with persistent fever with cytopenia . Therapy resistant MAS in rheumatological diseases is infrequently described and deciding on management need further evidence.