

Early Macrophage Activation Syndrome Triggered by Liver Biopsy in a Patient with Systemic Lupus Erythematosus: A Case Report (Abstract ID 024)

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Introduction

Systemic lupus erythematosus (SLE) is a complex autoimmune disorder with multi-organ involvement. Management involved immunosuppressive therapy which can be limited by complications such as transaminitis. This case report highlights a rare complication of macrophage activation syndrome (MAS) in a patient with SLE, triggered by a liver biopsy.

Case report

- 23-year-old lady with SLE (mucocutaneous, musculoskeletal, and haematological involvement) suffered frequent SLE flares.
- Persistent transaminitis hindered initiation of second-line immunosuppressants.
- Viral and autoimmune hepatitis serologies negative; US abdomen revealed fatty liver.

Electively admitted for liver biopsy to further investigate cause of persistent transaminitis

Biopsy successful and uneventful, however developed persistent high - grade fever (up to 40°C) within 24 hours post biopsy

Bone marrow aspirate and trephine (BMAT): positive CD163, increased histiocytes with hemophagocytic activity, confirming MAS in

Liver biopsy: steatohepatitis, no evidence of hemophagocytosis.

Empirically treated for hospital acquired infection and SLE flare with broad-spectrum antibiotics (meropenem) and IV dexamethasone but still deteriorated

Blood, sputum & urine cultures : no growth

CMV & EBV PCR: Not detected

Repeat US: hepatosplenomegaly

Worsening transaminitis, bicytopenia, hyperferritinemia, hypertriglycerideamia and low fibrinogen

Hscore 272: high probability of HLH

Treatment : high-dose pulse methylprednisolone (500 mg daily), oral cyclosporine (3 mg/kg twice daily), and intravenous immunoglobulin (IVIG) (0.5g/kg daily for 2 days).

Post treatment: fever, cytopenia resolved, liver function normalised

Discussion

- MAS a form of secondary hemophagocytic lymphohistiocytosis (HLH), is a rare but life-threatening complication in SLE¹.
- Early signs include sustained fever, bi/ pancytopenias, deranged liver function, organomegaly, and hyperferritinemia².
- Hscore is helpful in estimating an individual's probability of having reactive hemophagocytic syndrome³
- Bone marrow biopsy is key for diagnosis³.
- This case is unique for:

MAS onset within 24 hours of biopsy Diagnosis supported by **HScore** and **BMAT** No MAS histological features in liver biopsy

Conclusion

- This case underscores the importance of maintaining a high index of suspicion for MAS in SLE patients with acute deterioration post-procedure.
- Early recognition and timely treatment can be life-saving.

References

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- Grom AA et al. Nat Rev Rheumatol. 2016;12(5):259-
- 3. Fardet Let al. Arthritis Rheumatol. 2014;66(9):2613-