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MACROPHAGE ACTIVATION SYNDROME AND SYSTEMIC LUPUS ERYTHEMATOSUS: A CASE SERIES STUDY IN A REFERENCE HOSPITAL IN RIO DE JANEIRO

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BACKGROUND

Macrophage Activation Syndrome (MAS) is a life-threatening condition associated with many conditions, including rheumatic diseases. Due to the rarity of MAS associated to Systemic Lupus Erythematosus (SLE) most of the data available is from single cases reports.

MATERIALS AND METHODS

We performed a retrospective analysis of patients with SLE which presented MAS according to 2016 classification criteria for MAS, in a rheumatology reference hospital in Rio de Janeiro between March 2018 and February 2019.

RESULTS

In this review all patients fulfilled classification criteria for MAS with fever, low platelet count (< 181.000/µL), aspartate aminotransferase (AST) level >48units/L, triglyceride level >156mg/dL and presence of haemophagocytosis in bone marrow aspirate. Also, all patients presented severe anemia (Hb<8,0g/dL), Leukopenya (white blood cell count <4000/µL), 80% presented fall in the erythrocyte sedimentation rate (ESR), and low complement (c3<79/c4<16). 60% presented positive anti-DNAds. 60% received treatment with methylprednisolone 500mg and endovenous imunoglobulin 2g/kg, 20% received methylprednisolone 1mg/kg, and 20% received cyclosporine, prednisone 1mg/kg and endovenous immunoglobulin 2g/kg. The mortality was 0.

CONCLUSION

MAS is a severe condition related to rheumatic diseases, but its early recognizing and proper treatment may lead to low morbimortality rates.