MACROPHAGE ACTIVATION SYNDROME PRESENTING AS THE FIRST MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS: CASE REPORT AND LITERATURE REVIEW

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BACKGROUND

Macrophage activation syndrome (MAS) belongs to hemophagocytic lymphohistiocytosis (HLH) group of diseases, which includes familial HLH and secondary HLH, and it can be triggered by a variety of events that disrupt immune homeostasis, which includes infection, drugs, rheumatic disorder, and malignancy. In the range of rheumatologic disorders, it is most commonly associated with systemic juvenile idiopathic arthritis (SJIA). It is a rare and difficult diagnose with high mortality rate. In this article, we report a case of MAS as the first manifestation of systemic lupus erythematosus (SLE).

CASE REPORT

LGS, 20 years old, male, of black ethnicity, presented fever, headache, vomiting, inappetence, asthenia and weight loss about two months before hospitalization. The patient was admitted to the hospital for hematology care. The physical examination demonstrated hepatosplenomegaly, axillary lymph nodes and hyperchromic malar lesions. Imaging exams showed pleural and pericardial effusion. Laboratory results revealed pancytopenia, increased liver enzymes and lactic dehydrogenase, azotemia, hypertriglyceridemia, low fibrinogen and ferritin level > 7,000 ng/mL (normal = 16–300 ng/mL). A myelogram was performed without evidence of hemophagocytosis and no viral infections were detected. Immunological tests showed positive antinuclear factor, positive anti-DNA and complement consumption, configuring the diagnosis of SLE. As a result, he was transferred to the rheumatology unit one week after admission. The treatment chosen was pulse therapy with methylprednisolone for three consecutive days, meanwhile, the patient had generalized tonic-clonic seizures, acute kidney injury and pulmonary sepsis, requiring orotracheal intubation, renal replacement therapy and long stay in the intensive care unit (ICU). Due to main diagnosis of MAS, we associated IV human immunoglobulin for 5 days and, afterwards, we started IV cyclophosphamide. Gradually, the patient evolved with clinical improvement, being discharged from the hospital with hydroxychloroquine, prednisone and mycophenolate mofetil.

CONCLUSION

Macrophage activation syndrome is a syndrome caused by an abnormal downregulation by activated macrophages and lymphocytes, leading to excessive inflammation and tissue destruction. The association of SAM and SLE is a rare occurrence, the incidence varies between 0.9–4.6% and is generally related to SLE flares — our patient had SAM as a first manifestation of SLE, which is even more rare and challenging because both entities can cause similar clinical manifestations. The possibility of MAS must be urgently raised, avoiding delay in diagnosis and initiation of the goal treatment, which is critical for a successful outcome.

KEYWORDS

Macrophage activation syndrome, Systemic lupus erythematosus, Hemophagocytic lymphohistiocytosis.