MACROPHAGES ACTIVATION SYNDROME AS THE FIRST SERIOUS MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS - CASE REPORT

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BACKGROUND

Macrophage activation syndrome (MAS) is a rare, serious, and potentially fatal complication of systemic inflammatory diseases, caused by excessive activation and expansion of T lymphocytes and macrophages that exhibit hemophagocytic activity, resulting in overproduction of cytokines and a hyperinflammatory state. Several studies demonstrate an association of MAS with systemic lupus erythematosus (SLE).

CASE REPORT

Female, 37 years old, recently diagnosed with SLE, started toxemic constitutional symptoms, with fever (38° C), cough with yellow sputum, dyspnea, inflammatory arthralgia, and myalgia. On laboratory examination, she had bicytopenia, complement consumption, anti-dsDNA > 200. Chest X-ray showing peribronchovascular infiltrate on the right. Treatment with levofloxacin for the pneumonic process was started, cultures were collected and prednisone 1 mg/kg/day was added. The patient evolved with a significant worsening of the clinical picture, in addition to elevated transaminases, worsening of bicytopenia, hemoglobin: 7.9 g/dL, leukocytes: $0.8 \times 10^3/\text{mm}^3$, hypofibrinogenemia (92 mg/dL), hypertriglyceridemia (439 mg/dL), hyperferritinemia > 2000 and pericardial effusion. The hypothesis of MAS was considered. Myelogram performed by Hematology, which showed hemophagocytic activity at Figure 1. Methylprednisolone was started 1 g/day for 3 days, followed by cyclophosphamide at a dose of 0.6 g/m². The patient evolved with significant clinical and laboratory improvement and was discharged. Currently, he has good disease control.

Figure 1. Bone marrow cytology showing histiocyte at conspicuous phagocytic activity of erythrocytes and platelets.

CONCLUSION

Recognition of MAS in SLE is a challenge, because it can be confused with infection or associated with current infection since in clinical rheumatologic practice infections act as triggers for disease activity. Therefore, recognizing severe rheumatologic manifestations and conducting immunosuppressive therapy in the presence of infections is essential for favorable outcomes.

KEYWORDS

Macrophage activation syndrome, Systemic lupus erythematosus, Manifestation.