<https://doi.org/10.47660/CBR.2020.16999>

EXTREMELY HIGH FERRITIN LEVELS IN A PATIENT WITH MACROPHAGE ACTIVATION SYNDROME ASSOCIATED WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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BACKGROUND

Hyperferritinemia can be a result of inflammation, infection, chronic iron overload, or other uncommon pathologies, including macrophage activation syndrome (MAS). We herein present a case of severe MAS with an extremely high level of ferritin.

CASE REPORT

A 25-years-old female patient with systemic lupus erythematosus (SLE) since 2017, with previous history hemolytic anemia and nephritis, attended a medical consultation presenting nodular lesions in both thighs (with superficial phlogistic signs) and fever initiated one week before. She was taking chloroquine diphosphate 250 mg, mycophenolate mofetil 2g, prednisone 20 mg and enalapril 20 mg every day. She was sent to the emergency department, and lab evaluation demonstrated pancytopenia (hemoglobin, 9.0 mg/dL; leukocytes, 2020; segmented neutrophils, 1505; lymphocytes, 101; platelets, 79,000), acute renal failure (serum urea, 219 mg/dL; creatinine, 3.48 mg/dL; potassium, 7.1 mEq/L), hepatic disfunction [aspartate aminotransferase (AST), 218 U/L; alanine-aminotransferase (ALT), 55 U/L; gamma-glutamyltransferase (GGT), 1093 U/L; alkaline phosphatase, 712 U/L; total bilirubin, 3.21 mg/dL; albumin, 2.0 g/dL; lactate dehydrogenase (LDH), 2622 U/L]. Vital signs were normal. Empirical antibiotic coverage was started and the use of mycophenolate was interrupted. Intravenous methylprednisolone was prescribed at the dose of 60 mg/day. After 48 h of treatment, the patient presented copious melena and worsened neutropenia (790 segmented neutrophils and undetected lymphocytes). Upper endoscopy demonstrated multiple Cameron lesions, with negative search for cytomegalovirus (by biopsy and PCR). Considering the diagnosis of MAS, intravenous hyperimmune immunoglobulin (IVHI) was infused over 5 days (400 mg/kg/day). Hemodialysis was initiated due to acidosis and hyperkalemia refractory to clinical management. There was no apparent response to IVHI, and worsening of laboratory exams was observed (ferritin > 100,000 ng/mL; fibrinogen, 79 mg/dL; AST, 931 U/L; ALT, 155 U/L; triglycerides, 186 mg/dL; LDH, 9590 U/L). Pulse steroid dose (1000 mg/day of methylprednisolone for 3 days) and oral cyclosporin (up to 3 mg/kg) were added to the treatment. Bone marrow biopsy showed medullary hemophagocytosis. Ferritin remained in extremely high levels in 2 dosages (> 100,000 and equal to 90,069 ng/mL) over the next 3 days. Nine days after admission to the hospital room, the patient presented multiple bleeding sites (digestive, insertion of catheters, and biopsy wound) leading to hemodynamic instability. Etoposide treatment was indicated, but patient evolved with multiple organ failure and death.

CONCLUSION

We report here a case of extreme hyperferritinemia in MAS related to SLE. Reports of such high levels of ferritin are unusual, and are associated with poor clinical outcome.