MACROPHAGE ACTIVATION SYNDROME (MAS) REFRACTORY TO IMMUNOGLOBULIN WITH EXCELLENT RESPONSE TO RITUXIMAB: CASE REPORT

Douglas Squizzato Leite^{1,*}, Matheus Zanata Brufatto¹, Sean Hideo Shirata Lanças¹, Andrea de Almeida Peduti Batista¹, Laura Maria Silva de Siqueira¹, Henrique Pereira Sampaio¹, Luiz Eduardo Valente¹, Daniela Esteves Temporim¹, João Flávio Gomes Faria¹, Sula Glaucia Lage Drumond Pacheco¹

1. Universidade Estadual Paulista "Júlio de Mesquita Filho", Botucatu (SP), Brazil.

*Corresponding author: squizzattoleite@gmail.com

BACKGROUND

Macrophage activation syndrome (MAS) is a rare condition, potentially fatal, characterized by massive production of proinflammatory cytokines that results in failure of multiple organs and can be triggered by an initial picture of ASD.

CASE REPORT

A male patient, 38 years old, previously healthy with a history of daily fever (above 39 °C), myalgia, odynophagia and dry cough for 3 weeks. He was admitted to our service prostrate, tachycardic, tachypneic, with cervical and inguinal lymphadenomegaly and erythematous cutaneous rash in the abdomen and upper chest. Pulmonary auscultation with bilaterally based crackles and presence of pericardial effusion on echocardiogram and hepatosplenomegaly on ultrasound. He presented significant leukocytosis without left deviation, increased inflammatory tests (ESR: 120). Initially entered with antibiotic and initial hypothesis of leukemoid reaction or adult still disease (DSA) was made. Hydrocortisone was started at a dose equivalent to 1 mg/kg prednisone and an inguinal LFN biopsy was requested to rule out malignancy (result compatible with reactive lymphadenitis). He evolved with acute kidney injury requiring dialysis support. On the 5th day of hospitalization, he presented an increase in liver enzymes, triglycerides and ferritin > 2000, as well as a decrease in fibrinogen and ESR, with hemophagocytosis confirmed in the MO aspirate. Diagnosis of MAS was made and pulse of methylprednisolone (1 g for 3 days) and intravenous immunoglobulin (2 g/kg divided in 5 days) were performed. After initial improvement, on the 13th day of hospitalization, he evolved with cardiac arrest, ventricular fibrillation, being reversed after 5 cycles. In the post-CPR, cardiac tamponade was found, puncture was performed with exudative fluid and it was decided to expand antibiotic therapy. After extubation, he developed fever (negative cultures) and opted for rituximab. He presented gradual clinical and laboratory improvement and hospital discharge, maintaining remission and complete resolution of the MAS.

CONCLUSION

Report of a severe case of MAS, with cardiac tamponade and multiple systemic manifestations, refractory to pulse therapy with corticosteroids and immunoglobulin and with an excellent response to rituximab, progressing with sustained remission.

KEYWORDS

Macrophage activation syndrome, Rituximab, Cardiac tamponade.