MACROPHAGE ACTIVATION SYNDROME (MAS) TREATED WITH TOCILIZUMAB: CASE REPORT

Douglas Squizatto 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: squizattoleite@gmail.com

BACKGROUND

Adult Still's disease (ADS) can course (in 7–16%) with macrophage activation syndrome (MAS), an inflammatory systemic syndrome characterized by: cytopenia, organ dysfunction and coagulopathy. The use of target therapy in its treatment is still controversial, and there are no protocols. Tocilizumab (inhibitor IL-6) has been a treatment option.

CASE REPORT

A 21-year-old female patient with a history of systemic juvenile idiopathic arthritis in childhood (13 years old) presented with Still reactivation 2 months before admission (arthritis, evanescent maculopapular rash and fever), being treated in an external service with 1 mg/kg prednisone. She was admitted to our service with abdominal pain, vomiting accompanied by diarrhea (four episodes per day with blood). Laboratory tests showed anemia, leukocytosis and thrombocytopenia, in addition to hyperbilirubinemia and elevated liver enzymes and ferritin (> 2000). The hypothesis of still disease evolving with MAS (high entry H-Score) was raised with a possible infectious trigger of an abdominal focus. Hydrocortisone was started at a dose equivalent to 1 mg/kg of prednisone and piperacillin-tazobactam for possible gastroenteritis. Assessed by hematology that performed a biopsy showing hemophagocytosis and ruling out lymphoproliferative disease. On the third day of hospitalization, she presented worsening of anemia, thrombocytopenia, significant consumption of fibrinogen, drop in ESR and presence of schizocytes in the smear. Pulse therapy with methylprednisolone 1 g/day was started for 3 days and hemotherapy support with cryoprecipitate. Due to the lack of immunoglobulin in our hospital, induction with tocilizumab (8 mg/kg) was chosen, with significant improvement in the clinical condition, including normalization of the increase in liver enzymes, normalization of fibrinogen, and total resolution of MAS. She is currently in an outpatient follow-up with monthly infusions of tocilizumab, maintaining sustained remission with the medication.

CONCLUSION

Macrophage activation syndrome is a rheumatologic emergency that requires fast recognition and treatment. Although there is still controversy about the use of tocilizumab in these MAS conditions, our patient showed an excellent response to the imposed treatment, both to control the acute condition and to prevent relapses.

KEYWORDS

Macrophage activation syndrome, Tocilizumab, Adult Still's disease.