MACROPHAGE ACTIVATION SYNDROME IN SYSTEMIC-ONSET JUVENILE IDIOPATHIC ARTHRITIS – A DRAMATIC CASE OF SUCCESS

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BACKGROUND

Macrophage activation syndrome (MAS) is a rare and feared complication related to rheumatic diseases, especially systemic-onset juvenile idiopathic arthritis (s-JIA), resulting from uncontrolled activation and proliferation of T lymphocytes/macrophages. This syndrome can be triggered by disease flare, infections, or drugs. Although rare, it can be life threatening and is considered a rheumatologic emergency.

CASE REPORT

We present a 3-year-old boy with month-long prostration, lack of appetite, daily vespertine fever, arthralgia, and abdominal pain. After antibiotic courses without improvement, the child was hospitalized. His mother referred ongoing fever and arthralgia in the last 3 months, with continuous fever and worsen general condition over the last month. After infectious disease exclusion, the pediatric rheumatology team was called and rash during fever, arthritis of the wrists, knees and ankles, leukocytosis, anemia, high ESR, CRP, hyperferritinemia and no lymph nodes enlargement were noted. Echocardiogram, abdominal ultrasonography, and chest X-ray were normal. While awaiting hematologic evaluation, patient developed tachypnea, respiratory effort and O2 supplementation need. Increase in ferritinemia and triglycerides, as well as decrease in fibrinogen was noted, but only support measures were taken. After 3 days, a new diffuse interstitial alveolar infiltrate developed, and patient evolved to respiratory failure. Intensive care support was needed. During intubation, patient presented a 4-min cardiac arrest, and was successfully resuscitated. No optimal oxygenation was guaranteed, and venoarterial extracorporeal membrane oxygenation (ECMO) was installed. Immunoglobulin and pulse therapy with methylprednisolone were initiated followed by maintenance with immunosuppressive methylprednisolone. Due to no improvement in hemodynamic and respiratory parameters, oral cyclosporine was initiated. Although stability, an accidental decannulation from ECMO induced hypovolemic shock and another cardiac arrest for 27 min. After resuscitated, brain protection measures were instituted. As cardiac function worsened and X-ray demonstrated acute respiratory distress syndrome, 3-days plasmapheresis was executed. Forty-eight hours after the first infusion, patient was extubated and showed global improvement. Cyclosporine was held as maintenance therapy with no MAS relapse over a 6-month period. Although motor and cognitive sequelae (hemiplegia, hemispatial neglect and vocabulary loss), the patient presented important motor and cognitive improvement with physiotherapy and is rehearsing his first steps and phrases back after the dramatic outcome.

CONCLUSION

Although rare, MAS is a life-threatening condition. Lung and cerebral involvement are especially dangerous and intensive care is needed until drugs effects are obtained. Children without prolonged fever should be screened to MAS. Besides high dose corticosteroid therapy, immunoglobulin, cyclosporine, and biologics (canakinumab and tocilizumab), plasmapheresis can be considered in severe cases.

KEYWORDS

Macrophage activation syndrome, S-JIA, ECMO.