MACROPHAGE ACTIVATION SYNDROME IN JUVENILE SYSTEMIC LUPUS ERITHEMATOSUS – DISEASE ACTIVITY OR INFECTION?

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BACKGROUND

Macrophage activation syndrome (MAS) is a rare and feared complication related to rheumatic diseases, usually triggered by infections, disease activity or drugs. Due to immunosuppressive treatment, rheumatologists need to be especially aware of infections. Although bacterial infections as a complication are less common in young populations than in adults, viral infection or reactivation is a potential hazard.

CASE REPORT

A 14-year-old female with juvenile systemic lupus erythematosus presented with class IV nephritis and Libman–Sacks endocarditis at disease overtone. Two months after the diagnosis, the patient was discharged from hospital using aspirin, warfarin, hydroxychloroquine, prednisolone, and mycophenolate. A week after discharge patient was hospitalized with extensive shingles lesion and treated with acyclovir for a fortnight. Two months after discharge, patient was once again hospitalized with dry cough, fever and vomiting over 3-days after antibiotic course due to a knee lesion after a fall. Mycophenolate was discontinued while infection was excluded, and empiric antibiotics initiated. Echocardiogram demonstrated an improve in systolic function and valve vegetation regression. For *Escherichia coli* in urine culture, antibiotics were stepped up to cefepime. Three days after antibiotic exchange, fever returned. Screening for MAS was performed: CPR and triglycerides increase, fibrinogen and ESR decrease as well as a ferritin 45,000 ng/mL was noted. Vancomycin was associated, new blood cultures and viral PCR collected, and methylprednisolone pulse therapy associated with immunoglobulin initiated. Two days after measures, O₂ supplementation was needed, and progressed to NIV in a 12-h period. Patient did not tolerate weaning, and intensive care/intubation was proceeded. Child presented increased in ferritin level, diffuse pulmonary infiltrate, and cardiac enlargement at ICU admission. Child evolved to cardiogenic shock with an ejection fraction of 34%. Three-days plasmapheresis associated with immunosuppressive dose methylprednisolone was instituted with little clinical improvement, and rituximab was administered. Viral PCR results came in with positive qualitative PCR of Herpes 1/2/6 as well as positive quantitative CMV PCR with 23,671 copies (log 4.37). Ganciclovir was promptly initiated, but patient developed continuous fever, worsen shock and ferritin level of 330,877 ng/mL. Despite instituted measures, patient evolved to death.

CONCLUSION

Patients with rheumatologic conditions and prolonged fever should be screened for MAS. When its treatment doesn't go as planned and step up is needed, infection investigation is essential. Our case turns on an unfavorable outcome due to a combination of prior systemic lupus erythematosus sequelae, disease activity and cytomegalovirus infection. When infection is suspected or can't be excluded, empiric treatment is recommended.

KEYWORDS

Macrophage activation syndrome, Systemic lupus erythematosus, Cytomegalovirus.