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MACROPHAGE ACTIVATION SYNDROME SECONDARY TO EPSTEIN BARR VIRUS AND CYTOMEGALOVIRUS REACTIVATION IN A PATIENT WITH SARCOIDOISIS: A SUCCESSFULLY TREATED CASE REPORT

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BACKGROUND

Macrophage Activation Syndrome (MAS) is a rare life-threatening disorder among the hemophagocitic syndromes, usually triggered by rheumatologic diseases, malignancies and infections, especially Epstein Barr Virus (EBV) and Cytomegalovirus (CMV). It consists in deregulated macrophage activation, causing cytokines storm, hemophagocistosis and multiple organ dysfunctions, mimicking different conditions, such as sepsis. Treatment should be started by suspicion, given the high mortality ratio. The authors describe a successfully treated case of MAS, secondary to EBV and CMV reactivation in a patient with Sarcoidosis.

CASE REPORT

67-year-old male, priorly diagnosed with cortico-dependent Sarcoidosis, received methylprednisolone pulsotherapy in 06/2018 followed by prednisone 25 mg/day and azathioprine 150 mg/day due to disease activity (weight loss, low fever, fatigue, elevated CPR). In 09/2018 he was hospitalized with dry cough, prostration, daily 40°C fever and hypoxia. Physical exam revealed Hepatosplenomegaly with diffuse limphadenomegaly, and admission exams showed pancytopenia and hepatic dysfunction (AST/ALT/LDH – 219/304/1609 U/L). Immediate diagnosis of sepsis or relapsing Sarcoidosis was considered, and broad spectrum antibiotics along with corticosteroids were initiated. There was, however, insignificant improvement with maintained pancytopenia, leading to reproaching the differential diagnosis as other laboratory findings surprised ferritin > 1500 ng/mL, triglycerides 404 mg/dL and fibrinogen 104 mg/dL. Bone marrow biopsy was performed and caught hemophagocitosis figures, establishing the diagnosis of MAS. Treatment protocol was promptly instituted with dexamethasone and posterior introduction of cyclosporine. Further positive qualitative PCR for EBV and CMV suggested reactivation (prior positive IgG) triggering the syndrome. Ganciclovir was added to the scheme, but not rituximabe due to multiple concomitant bacterial infections. Despite the critical state, patient responded and tolerated the therapy, with improvement in triglycerides (179 mg/dL), fibrinogen (428 mg/dL), AST/ALT/LDH (69/86/770 U/L) levels and CMV/EBV quantitative PCR (undetectable/22 copies per mL), receiving clinical discharge 3 months after the admission, using cyclosporine for outpatient follow-up.

CONCLUSION

Fast recognition of MAS is the most important prognostic factor, but it can be challenging given its rarity and more common, yet severe, differentials. Some clues are hyperferritinemia, hypertriglyceridemia, hypofibrinogenemia and sustained pancytopenia. Treatment may not wait until all specific exams are performed in unstable patients, and may require administration of dexamethasone followed by cyclosporine, as well as approaching the trigger, with ganciclovir/rituximabe for CMV/EBV. Mortality, however, remains up to 46% in 6 years with treatment, and near 100% without it.