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An unusual cause of stupor in a febrile patient

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The case study begins with a 45yr old, Filipino lady, presented to the hospital with history of fever with chills, myalgia, and generalized rash for five days. Observation in hospital and further workup was suggestive of Adult onset Still's disease (AOSD). She was started on oral steroids, but within 24 to 48 hrs of starting steroids, she had a febrile spike and her GCS dropped from 15/15 to 9/15. Urgent workup was sent for a possible undiagnosed infection, which could have flared up on steroids. Though she had a systemic inflammatory response syndrome (SIRS), the lab parameters were not suggestive of infection, and the possibility of macrophage activation syndrome (MAS) was suspected and work up was sent. Clinical and laboratory features were suggestive of MAS. She was started on pulse steroids, and IVIG. Her clinical condition improved over the next five days, with sensorium returning to normal, resolution of the SIRS and laboratory parameters showing a trend towards normalization. At discharge, she was well, afebrile, and the rash had resolved. She stayed a total 10 days in the hospital, was discharged on oral Prednisolone 1 mg/kg and advised rheumatology to follow up. This case is remarkable, for highlighting, the not commonly known fact that macrophage activation syndrome, can present with a decreased level of consciousness and prompt diagnosis and management can avert a potentially calamitous outcome. It is also significant to note that, the course of AOSD can become complicated despite steroids being initiated when the patient is clinically stable and early in the course of the disease. Learning points: AOSD can be diagnosed early when a high index of suspicion is entertained for patients with the classical rash that may show variation (though not complete disappearance) from time to time, quotidian high grade fever with neutrophilic leukocytosis and high ferritin. MAS should be strongly considered in patients with AOSD developing SIRS. MAS can develop despite starting steroids for the patient and relatively early in the presentation of the disease. A fall in GCS is one of the less known presenting features of MAS. Classically described and highlighted features like pancytopenia may be absent early in the course of the disease. Prompt initiation of definitive treatment is crucial for improved outcome. Our case substantiates the claim that early treatment and combination of IVIG has a successful outcome.

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