

A Case of Hemophagocytic Lymphohistiocytosis confused with Stevens-Johnson Syndrome

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Introduction: Stevens-Johnson syndrome(SJS) is a severe cutaneous adverse reaction(SCAR) characterized by epidermal detachment and extensive necrosis. It is mainly caused by immune-mediated reactions to medications. In comparison, hemophagocytic lymphohistiocytosis(HLH) is an uncommon yet life-threatening syndrome characterized by immune dysregulation. It can be classified as either a primary(genetic) or secondary(reactive) disorder. In a clinical setting, typical manifestations include prolonged fever, cytopenia and hepatosplenomegaly.

Case: Recently, we experienced a case of SJS complicated with HLH in a 77-year old man who was admitted with fever and a rapidly worsening maculopapular rash. At this point, he had been undergoing antibiotic treatment(ceftriaxone) for the past two days. On examination, the patient was febrile; additionally, maculopapules and blisters with positive Nikolsky sign were scattered over the eyelid, trunk, and thigh. SJS, most likely due to the ceftriaxone or an infectious cause, was the differential diagnosis. Skin punch biopsies showed subacute vacuolar interface dermatitis with superficial perivascular lymphocytic and eosinophilic infiltration, consistent with drug eruption, and the patient was administered treatment with methylprednisolone. Despite the steroid, however, the patient's fever and skin lesions worsened. It was then found that the patient satisfied the criteria for HLH, including fever, cytopenia(anemia and thrombocytopenia), hypofibrinogenemia(64mg/dl), hemophagocytosis in the bone marrow, elevated ferritin(6325ng/mL), and an elevated soluble IL-2 receptor level(11395U/mL). He was in a state of cytokine overproduction consistent with HLH. Treatment with IV immunoglobulin was initiated; however, the patient continued to deteriorate with progressive multiple organ failure. He died on hospital day 15.

Conclusion: SJS is rarely observed in conjunction with, but the combination can be life-threatening. SJS and HLH share similar clinical and laboratory features in that both are associated with severe inflammation reactions. This can make it difficult to differentiate SJS combined with HLH from deterioration due to underlying SJS.

