



HHV8-related hemophagocytic syndrome: diagnosis is in the eye

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A 33-year-old HIV-positive woman was admitted to the ICU for acute respiratory failure. She received immediate intubation for severe ARDS and hemodynamic instability. Physical examination reported diffuse lymphadenopathy, hepatic and splenic enlargement and a conjunctival Kaposi's sarcoma (KS). Comprehensive workup led to the diagnosis of hemophagocytic lymphohistiocytosis with fever, cytopenias, high levels of ferritin and triglycerides, and cytologic features of hemophagocytosis on bone marrow smears. No infection was documented. However, intense HHV8 replication was highlighted in the blood (>8 log), BAL and bone marrow. Flow cytometry demonstrated a circulating monotypic IgM- λ large B cell lymphocytosis consistent with the phenotype of HHV8 plasmablastic cells seen in HHV8 multicentric Castleman disease. Etoposide was administered, and liposomal doxorubicin was started for the KS. The patient was extubated at day 9 (Fig. 1).



Fig. 1 Kaposi sarcoma lesion on the left ocular conjunctiva

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