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**Title:** Lymphadenopathy in the time of Kaposi's and a tale of two cytopenias

## **Abstract**

### *Background*

Multicentric Castleman disease (MCD) is a polyclonal lymphoproliferative disorder arising idiopathically or secondarily. Immune dysregulation in MCD causes inflammatory symptoms and autoimmune cytopenias. We report a case of HHV-8+ MCD complicated by Evans syndrome (ES) - a rare disorder of autoimmune cytopenias of two or more cell lines.

### *Case Presentation*

A 49-year-old man presented with known history of HIV previously complicated by Kaposi sarcoma one week of shortness of breath, cough, and fever. His temperature measured 103°F, heart rate 140 beats/minute, and respiratory rate 24 breaths/minute. His hemoglobin was 4.3 g/μL and platelet count was 24,000 /μL. There was no evidence of hemolysis and direct antiglobulin testing (DAT) was negative. Splenomegaly and diffuse lymphadenopathy throughout the chest, abdomen, and pelvis were present on computed tomography. He required frequent red cell and platelet transfusion support. Platelet refractoriness workup confirmed the presence of alloantibodies. Enhanced DAT detected low titer IgG auto-antibodies. Excisional lymph node biopsy revealed vascular proliferation and dense plasma cell infiltrate consistent with MCD. Treatment with rituximab and liposomal doxorubicin (R-Dox) led to hematologic recovery within 6 weeks and radiographic resolution within 13 weeks.

### *Discussion*

ES is a rare complication of MCD. Enhanced DAT should be considered when there remains high clinical suspicion for autoimmune cytopenia despite a negative DAT. We present the first reported case of ES in HHV-8+ MCD, which was successfully treated with R-Dox. Further study is warranted to assess response rate and remission durability.