AUTOIMMUNE HYPOGLYCEMIA EXPANDS THE BIOLOGICAL SPECTRUM OF HHV8+ MULTICENTRIC CASTLEMAN DISEASE.

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Castleman disease (CD) is a rare non-clonal lymphoproliferative disorder. Diagnosis is based on histological features (1). Some cases of multicentric CD (MCD) are related to a polyclonal proliferation of HHV8-infected plasmablasts (called "HHV8+ MCD"), mostly encountered in immunocompromised hosts. The infected cells are uniformly IgM and λ–positive and locate in the mantle zone of the lymph node. These cells are associated with a marked polyclonal plasma cell infiltrate. Hemophagocytic lympho-histiocytosis (HLH) and other cytokine release complications make this condition life-threatening (2,3). Numerous auto-antibodies can develop during MCD flares, and autoimmune thrombocytopenia (ITP), autoimmune hemolytic anemia (AIHA) and Thrombotic Thrombocytopenic Purpura (TTP) (4) have been described. We here report on autoimmune hypoglycemia as a new autoimmune complication occurring in 5 patients with active HHV8+ MCD. We demonstrate that hypoglycemia is caused by autoantibodies directed towards the insulin receptor and that rituximab is an effective and safe treatment of this condition.

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