

**Fatal HHV8-associated Hemophagocytic
Syndrome in an HIV-negative
Immunocompetent Patient with Plasmablastic
Variant of Multicentric Castleman Disease
(Plasmablastic Microlymphoma)**

莊世松

Li CF;Ye H;Liu H;Du MQ;Chuang SS

摘要

Abstract

Virus-associated hemophagocytic syndrome (VAHS) triggered by HHV-8 is extremely rare and has been reported only in 9 immunocompromised patients. We report the first case of HHV-8-associated VAHS in an HIV-negative, immunocompetent patient with plasmablastic variant (plasmablastic microlymphoma) of multicentric Castleman disease (MCD). This 61-year-old man presented with fever, cough, and bilateral inguinal lymphadenopathy. Biopsy of the right inguinal lymph node revealed plasmablastic MCD with nodular aggregates of plasmablasts expressing IgM, MUM1, HHV-8 latency-associated nuclear antigen, and viral interleukin-6. These plasmablasts were monotypic for Iglambda light chain expression but not Igkappa. All the B-cell clonality assays, including IgH-FR2, IgH-FR3, DH-JH, Igkappa, and Iglambda PCR, showed a polyclonal pattern. His serum human interleukin-6 level was markedly elevated and was negative for EBV acute infection/reactivation. The marrow aspirate showed florid hemophagocytosis. His disease progressed rapidly to multisystemic illness, and he died of acute respiratory failure in 1 month. Our case showed that HHV-8 might trigger VAHS in an immunocompetent patient with plasmablastic MCD. We speculated that our patient developed VAHS under the cytokine storm associated with the proliferating HHV-8-infected plasmablasts, similar to the EBV-triggered VAHS in patients with EBV-associated T-cell lymphoma.