Massive expansion of EBV+ monoclonal T-cells with CD5 down-regulation in EBV-associated hemophagocytic lymphohistiocytosis.

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摘要

Abstract

Haemophagocytic lymphohistiocytosis (HLH) comprises primary and secondary forms; the secondary form is most commonly triggered by the Epstein - Barr virus (EBV; EBV-HLH). Patients with EBV-HLH usually exhibit oligoclonal or monoclonal T cell proliferation, which may mimic T cell lymphoproliferative disorder (T-LPD). This article reports on EBV-HLH in a 17-month-old girl with an extreme surge of reactive T lymphocytosis (absolute count 167×109/l) with CD5 down regulation. Bone marrow aspirate and trephine contained florid haemophagocytosis and massive infiltration of CD3+ Epstein - Barr virus-encoded RNA+ lymphocytes, as seen by double labelling. These lymphocytes were monoclonal for EBV and T cell receptor γ chain gene rearrangement. The patient responded dramatically to intravenous immunoglobulin, interferon α2b, ganciclovir and prednisolone, suggesting restoration of her immune system and eradication of the clonal T cells through these immunoregulatory agents. Thus, careful clinicopathological correlation is warranted in the interpretation of immunophenotyping and clonality data in T cell proliferation in association with EBV-HLH to avoid erroneous diagnosis of T-LPD.