Bone marrow granulomata and hemophagocytosis in a patient with Epstein-Barr virus infection

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Introduction

Epstein Barr virus (EBV) infection is known to cause hemophagocytic syndrome in certain patients. On the other hand, granulomata in the bone marrow have also been associated with viral infections, such as EBV. However, it is very rare to have both hemophagocytic syndrome and granulomata in the bone marrow due to an EBV infection. We present a case of hemophagocytic syndrome and bone marrow granulomata after primary EBV infection in a young adult.

Clinical History

The patient is a previously healthy thirteen year old female who presented with fevers and chills for two weeks. The admission peripheral blood smear showed pancytopenia and microcytic hypochromic anemia with minimal reticulocytosis. No immature cells were seen. These peripheral smear findings were consistent with inadequate hematopoiesis of the bone marrow.

Bone Marrow Findings

A bone marrow biopsy and aspiration were performed in order to investigate the etiology of pancytopenia. The bone marrow was normocellular for age with no evidence of malignancy. There was increased erythropoiesis with a few dysplastic forms. Examination of the bone marrow aspirate demonstrated a moderate number of macrophages containing many phagocytosed erythrocytes and myeloid cells (Figures 1 and 2). In addition, focal granulomata were also seen in the bone marrow biopsy (Figures 3 and 4).

Immunohistochemistry

No acid fast bacilli or fungal organisms were identified on acid Fast and Gomori Methenamine Silver stains, respectively. However, EBV-LMP immunohistochemical stain, and EBV in-situ hybridization (EBER) performed on the bone marrow biopsy showed marked positivity within the granulomata (Figures 5 and 6). The non-granulomatous areas of the biopsy were otherwise negative. Serology for EBV indicated a latent infection, with positive EBV VCA IgG and EBVNA IgG, as well as negative EBV VCA IgM and EBVEA IgG.

Conclusion

The constellation of clinicopathologic findings are consistent with EBV-induced hemophagocytosis and granulomata resulting in pancytopenia. The patient was subsequently started on steroids and cyclosporine and is currently undergoing treatment. To the best of our knowledge, the presentation of concurrent findings of hemophagocytosis and granulomata on the bone marrow biopsy with positive for EBV-LMP and EBER in a patient with EBV infection has not been previously described.

References