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Basic principles and emerging concepts in Epstein-Barr virus-positive marginal zone lymphoma

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Abstract

Epstein-Barr virus (EBV) is well known to be associated with various types of B cell, T cell, and NK cell lymphomas. The roles of EBV in lymphomagenesis are now better elucidated through decades of research. Marginal zone lymphomas (MZL) are a heterogeneous group of small B-cell lymphomas and traditionally EBV negative. However, recent studies have identified EBVpositive extranodal MZLs particularly in transplant recipients and at least partially responsive to reduced immunosuppression, suggesting that these should be regarded as a form of post-transplant lymphoproliferative disorders (PTLD). We expanded the spectrum of EBV+ MZLs by identifying the first case of nodal MZL, and more cases of extranodal MZL but in non-transplant settings that included iatrogenic immunosuppression, congenital immune deficiency, and increased age as the only potential cause of immune dysfunction. These cases were either EBV latency I or II, with a typical plasmacytoid and/or monocytoid B cells positive for EBV in all cases. Unlike published series that were predominantly IgA-positive, our cases were either positive for IgG or IgM. Cases arising from cutaneous sites and salivary glands demonstrated differing characteristic features in morphology. Our data show that EBV+ MZLs can arise in a variety of clinical settings and are most often extranodal. Most patients had a clinically indolent disease with response to reduction of immune suppression, or immunochemotherapy. As these lymphomas warrant different management strategies compared to EBV-negative cases, a high suspicion must be kept, and relevant workup should be performed for nodal and extranodal MZLs in post-transplant and non-transplant but immunocompromised patients.

Biography

Shunyou Gong is the director of Hematology and Hematopathology at the Ann & Robert H Lurie Children's Hospital of Chicago and assistant professor at Northwestern University Feinberg School of Medicine. His clinical interests and areas of expertise include pediatric hematopoietic malignancies, inherited bone marrow failure syndromes, and bleeding or thrombotic disorders. As first-author or corresponding author, he has published many landmark papers on prestigious journals including Cell, Blood, and American Journal of Surgical Pathology.

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