Introduction to Epstein-Barr Virus and Lymphoproliferative Diseases in Immunodeficient Individuals

David T. Purtilo and George Klein

EBV is an ubiquitous B-cell-tropic virus responsible for infectious mononucleosis (5) and is one possible etiological agent of Burkitt lymphoma and nasopharyngeal carcinoma (6, 7). When the virus infects a young child, silent seroconversion is the rule. In contrast, primary infection of young adults results in infectious mononucleosis in about one-half of the cases (6). The observations of fatal infectious mononucleosis, agammaglobulinemia, and aplastic anemia associated with infectious mononucleosis and malignant B-cell lymphoma in 6 maternally related males in the Duncan kindred (11) and other similar families (1-4, 8, 9) had suggested to Purtilo (10) that immunodeficient responses to EBV were responsible for a variety of diseases in males with the X-linked lymphoproliferative syndrome. Primary infection by EBV normally provokes potent immune responses which efficiently cope with the B-cell proliferation and provide lifelong latency (6). Persons with inherited or acquired immune defects, depending on the type and degree of the deficiency, are more or less vulnerable to life-threatening B-cell proliferation.

The articles published in this symposium focus on the possible roles of EBV and immunodeficiency in lymphoproliferative disorders. Some of the major objectives of the reports are to: describe various diseases possibly associated with EBV in immunodeficient patients; demonstrate inherited or acquired cellular and humoral immune defects to EBV-specific antigens; and document the presence of viral genome in lymphoid tissues from the immunodeficient patients. The authors have sought to develop criteria for establishing involvement or noninvolvement of EBV in the lymphoproliferative diseases in immunodeficient patients.

References

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