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"Primary Immunodeficiencies Inducing EBV-Associated Severe Illnesses "

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Abstract:

Epstein-Barr virus (EBV) is a ubiquitous human g -herpesvirus that infects about 95% of the adult population. The majority of primary infections occurs in early childhood and is generally subclinical; it can cause infectious mononucleosis (IM), which is usually a self-limiting lymphoproliferative disorder. However, infection of EBV occasionally results in severe, often lethal diseases, which include fatal IM, hemophagocytic syndrome, polyclonal lymphoproliferative disorders, and malignant lymphoma. These severe EBV-related illnesses occur secondary to some primary immunodeficiency diseases showing inefficient immune reaction to EBV. One example is X-linked lymphoproliferative disease (XLP), which is caused by mutations in the SLAM-associated protein (SAP) gene. The major clinical manifestations of XLP are fulminant IM, malignant lymphoma and dysgammaglobulinemia. Aplastic anemia, virus-associated hemophagocytic syndrome, and vasculitis have also been reported in XLP. We have developed a flow cytometric method using the anti-SAP monoclonal antibody to search for XLP. This clinically useful assay has successfully been used to identify XLP patients in Japan. In this review, clinical and mutational characteristics of XLP in Japan are mainly described. In addition, it is shown that the similar situations to XLP can occur in other primary immunodeficiencies involving T-cell killing function, such as autoimmune lymphoproliferative syndrome caused by Fas gene mutations or familial hemophagocytic lymphohistiocytosis caused by perforin gene mutations. Finally, the EBV-related terrible disease condition, namely chronic active EBV infection, which is common in Asian areas but its genetic background remains to be elucidated, will be toughed on.

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