Primary Effusion Lymphoma

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The known causative factors of malignant lymphomas include viruses, bacteria, environmental factors, and genetic factors. In particular, information on the association between viruses and malignant lymphoma has been accumulating: human T-cell lymphotropic virus type I and adult T-cell leukemia/lymphoma; Epstein Barr virus (EBV) and Burkitt’s lymphoma, Hodgkin’s lymphoma, immunodeficiency-related lymphoproliferative disorders and pyothorax-associated lymphoma (PAL) (1); human herpesvirus-8 (HHV8) and primary effusion lymphoma (PEL) (2); and hepatitis C virus (HCV) and lymphoplasmacytic lymphoma (3). Recently simian virus 40 has been reported to be a new candidate etiologic factor for non-Hodgkin’s lymphoma (NHL) (3).

B-NHL occasionally develops in the body cavity of human immunodeficiency virus (HIV)-positive individuals, thus it is called body cavity-based lymphoma (BCBL) (2). In addition to the presence of HHV8, these BCBLs possess certain unusual clinical, immunophenotypic, and molecular genetic characteristics that suggest they represent a distinct clinicopathologic entity. They grow exclusively or mainly in the pleural, pericardial, and peritoneal cavities as lymphomatous effusions, usually with no identifiable tumor mass, throughout their clinical course and frequently exhibit indeterminate immunophenotypes but B cell genotypes with clonal immunoglobulin gene rearrangements, nearly always contain EBV, and consistently lack c-myc gene rearrangements. PAL is B-NHL that also arises in the pleural cavity after long-standing pleural inflammation resulting from therapeutic artificial pneumothorax or from tuberculous pleuritis. Although PALs present as solid tumor masses, they are otherwise similar to BCBL in that they are B cell lymphomas, usually exhibit immunoblastic morphology, and contain EBV. Then presence of HHV8 sequences was investigated in 14 PAL, giving negative results in all cases. This finding strongly suggests that BCBLs and PALs are distinct clinicopathological entities and further strengthens the association between the presence of HHV8 and an effusion phenotype. Based on these findings, the term BCBL was replaced with the term PEL (5). In this issue, a case of PEL which developed in a patient with a history of lymphanginoma of forearm, protein losing enteropathy, systemic edema, repeated chylous ascites and chylothorax is described (6).

Unlike the ordinary PEL, the examinations revealed that this PEL did not contain HIV, HHV-8, or EBV sequences. After speculating mutual relationships among the above-mentioned findings, the authors claimed that their PEL case has a different etiology from HHV8-positive PEL and might be classified in a different category than classical HHV-8 positive PEL. More findings, especially positive findings characteristic for this tumor, including identification of etiologic factors should be provided to confirm their proposal. In this regard, the accumulation of well-characterized, similar cases is expected.

Katsuyuki AOZASA, MD
Department of Pathology,
Osaka University Medical School, Suita, Osaka 565-0871

References