International Session 8: ‘Epstein-Barr virus-associated lymphomas’

EXTRANODAL NK/T CELL LYMPHOMA, NASAL TYPE

Won Seog Kim
Samsung Medical Center, Seoul, Korea

Extranodal NK/T-cell lymphoma (ENKTL) is a rare histopathologic subtype with strong association with Epstein-Barr virus (EBV) infection. This disease is more common in Asia, especially far eastern countries, such as China, Korea, Taiwan, and Japan.

Interestingly, even inside Asia, the incidence in middle eastern Asian countries is less than 10% like Western areas. The prognosis of ENKTL was very poor for a long time. Especially, for advanced stage disease, long term survival was less than 10%. However, the natural history is significantly improved due to recent advances in the treatment. Two big progresses are concurrent chemo-radiation for localized disease and using L-asparaginase containing regimens like SMILE (steroid, ifosfamide, L-asparaginase, and etoposide) or AspaMet-Dex (L-asparaginase, methotrexate and dexamethasone). Serial monitoring of EBV-DNA copy number in plasma or whole blood is an useful tool to predict response, outcome and toxicity against chemotherapy. The role of PET is still controversial. Deletion of 6q has been most widely reported. Some genetic changes of HACE1, PRDM1, FOXO3, P53, PDGFR, PLK1, Aurora A, JAK3 have been reported. For future, knowledge on alteration of signaling and role of EBV in lymphomagenesis is necessary for biologic treatment approach.