haematological malignancies

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CLINICAL FEATURES AND OUTCOMES OF HUMAN IMMUNODEFICIENCY VIRUS (HIV)-NEGATIVE PATIENTS WITH MULTICENTRIC CASTLEMAN’S DISEASE

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Aim: Castleman’s disease (CD) is a rare polyclonal lymphoproliferative disease that presents a variety of symptoms and clinical courses. Multicentric Castleman’s disease (MCD) is defined by the involvement of at least two non-contiguous lymph node areas and commonly associated with poor clinical outcomes. We performed a retrospective analysis to reveal the clinical features and prognostic factors for patients with MCD.

Methods: Between 1990 and 2013, 80 patients of biopsy proven CD were identified from the database of Asan Medical Center, Seoul, Korea. The data were collected retrospectively by reviewing medical records. Among them, 32 patients were classified into MCD. With the exclusion of 4 patients with unknown HIV infection status, a total of 28 HIV-negative patients with MCD were included in this analysis.

Results: Most patients were male (76%) and median age was 54 years (range, 13-75) at diagnosis. Hyaline vascular variant was the most common subtype (39%) and followed by plasma cell variant (36%) and mixed cell variant (25%). Frequently reported symptoms and signs at diagnosis were hepatosplenomegaly or splenomegaly (57%), fever (39%), edema (29%) and ascites (18%). As an initial therapy, cytotoxic chemotherapy, glucocorticoid alone, and interferon-α were given in 11 (39%), 6 (21%), and 1 (0.4%) patients, respectively. Remaining 10 (36%) patients were managed with ‘watch and wait’ strategy. With median follow-up of 67 months (range, 1-162), 10 (36%) patients died and 5-year overall survival rate was 77% (95% CI, 61%-93%). Patients who showed extravascular fluid accumulation (peripheral edema, ascites, pleural effusions) were significantly associated with poorer survival than those without those signs (5-year survival rate, 94% vs 56%; p = 0.04). Extent of involvement (same side vs both sides of diaphragm) was marginally associated with survival (p = 0.11). However, other clinicopathologic factors, including gender, age, histopathologic types, hepatosplenomegaly, anemia, thrombocytopenia, serum albumin and lactate dehydrogenase were not associated with survival (p > 0.1 for all comparisons).

Conclusions: Clinicopathologic features of MCD in our cohort were consistent with previous reports. Our analyses showed that the signs of extravascular fluid accumulation at diagnosis were associated with poor overall survival in patients with MCD.

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