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NCCN Guidelines Panel: T-Cell/Primary Cutaneous Lymphomas Panel

On behalf of Sun Yat-sen University Cancer Center, I respectfully request the NCCN (T-Cell/Primary Cutaneous Lymphomas Panel) to review the enclosed data for inclusion of the new staging system for extranodal natural killer T-cell lymphoma (ENKTL).

Specific Changes: Recommend the CA staging system as a routine staging for the initial diagnostic evaluation of ENKTL (changed from “Workup, essential”).

FDA Clearance: Not available.

Rationale: As is well-known, ENKTL is an aggressive malignancy, and patients suffering from this type of disease have poor survival outcomes. However, using the Ann Arbor staging system (AASS), most ENKTL patients are categorized as early stage, which is inconsistent with their poor survival. AASS has been shown in many studies to be unsuitable for the staging of ENKTL. We developed the CA staging system to predict overall survival in patients with newly diagnosed ENKTL on the basis of data from the largest study population published so far. We also validated the system in an independent international cohort. The CA system exhibited a more balanced patient distribution and a much more reasonable discrimination of survival outcomes than the AASS, it also might inform treatment decisions for ENKTL. We believe that CA could be used to develop risk-adapted treatment approaches for patients with ENKTL and could be useful for future clinical practice and clinical trial design. This study has been published at Leukemia 2020 Feb 17, DOI: 10.1038/s41375-020-0740-1. Attached is the paper of CA system. We hope that NCCN guidelines could consider to adopt the CA staging system.

The following article are submitted in support of this proposed change. We are grateful to NCCN members for evaluating our request.

1. Huangming Hong et al. A proposal for a new staging system for extranodal natural killer T-cell lymphoma: a multicenter study from China and Asia Lymphoma Study Group. Leukemia 2020 Feb 17, DOI: 10.1038/s41375-020-0740-1. This paper supported the superior of CA staging system compared with Ann Arbor staging system. CA demonstrated with a more balanced patient distribution and a much more reasonable discrimination of survival and might inform treatment decisions for ENKTL.
2. Yong Yang et al. Prognostic Nomogram for Overall Survival in Previously Untreated Patients with Extranodal NK/T-Cell Lymphoma, Nasal-Type: a Multicenter Study. Leukemia 2015 Jul; 29(7). This paper revealed that the predictive accuracy of Ann Arbor staging system has been shown to be

limited for ENKTL.

3. Xin Li et al. DDGP versus SMILE in Newly Diagnosed Advanced Natural Killer/T-Cell Lymphoma: A Randomized Controlled, Multicenter, Open-label Study in China. Clin. Cancer Res. 2016 Nov 01; 22(21). This paper used a modified Ann Arbor new staging system, which is similar with CA staging system, the CA staging system would be useful for future clinical practice and clinical trial design.

Sincerely,

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