

Preface: why this special series on marginal zone lymphomas?

Marginal zone lymphoma (MZL) is a relatively uncommon form of indolent non-Hodgkin lymphoma currently defined anatomically as extranodal, nodal and splenic. The therapeutic approaches are variable, ranging from antimicrobial approaches to monoclonal antibodies, chemoimmunotherapy, kinase inhibitors and cellular therapies. From April 17–18, 2019, the Lymphoma Research Foundation (LRF) gathered international experts including members of the International Extranodal Study Group in the field of MZL in New York City for the inaugural International Scientific Workshop on Marginal Zone Lymphoma. This group collectively realized that there was far more to be learned and accomplished in MZL.

One impediment to therapeutic progress in MZL is that, MZL and follicular lymphoma (FL) patients are often included in the same protocols despite their differences in biology and natural history. Thus, a number of questions were addressed: within MZL is the biology of extranodal, nodal and splenic subtypes different? Can broader epidemiology studies contribute to a better understanding of these diseases? How can further understanding of the biology of transformation aid in the management strategies of MZL? Would it be preferable for MZL and FL patients be included in the different clinical trials? In extranodal MZL, should refinements in measurable disease criteria such as orbital disease be developed in certain subsets? What is the optimal radiation therapy approach in the different subsets of MZL? What future directions can enhance design and accrual to clinical trials?

A number of conclusions were drawn from this Workshop. Given the differences in biology and natural history of MZL and FL, clinical protocols should not lump these entities together as low-grade lymphoma. Because the rarity of the diseases, collaborative efforts are imperative. Further delineation of the cell of origin of MZL cells is critical to further development of therapeutic strategies to improve outcomes in the various subsets of MZL. Since MZL is currently considered incurable, patients are subjected to a series of treatment options. A better understanding of the molecular etiology will help individualize therapy and direct patients to the optimal sequence of the novel targeted treatment options becoming available.

One outcome of this Workshop is this focused issue on MZL which serves as the most extensive and comprehensive review of individual topics in MZL by international leaders in the field. We hope that it will provide a roadmap taking us from where the field currently is, to where it is going, but, more importantly where it will need to go to lead to the cure of patients with MZL.

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