

Nongastric marginal zone B-cell lymphoma: A prognostic model from a retrospective multicenter study

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Abstract

The International Prognostic Index (IPI) and Follicular Lymphoma Prognostic Index (FLIPI) are used as prognostic indices for NHL and indolent lymphoma. However, marginal zone B-cell lymphoma (MZL) evidences a distinctive clinical presentation and a natural course; thus, in this study, we attempted to devise an adequate prognostic index for MZL. Two-hundred and five patients diagnosed with MZL were retrospectively reviewed. After analysis of the prognostic factors, progression-free survival (PFS), and overall survival (OS), we constructed a prognostic index of MZL (MZLPI) via the summation of each factor. We then compared PFS and OS with IPI, FLIPI, and MZLPI. According to our multivariate analysis, nodal MZL, ECOG performance ≥ 2 , and advanced stage were composed of MZLPI. MZLPI was grouped as follows: score 0 as a low-risk group, score 1 as an intermediate risk group, and score ≥ 2 as a high-risk group. The PFS curve, according to MZLPI results, evidenced a more discriminated pattern than IPI and FLIPI, and this was especially true in the intermediate risk group. In OS, MZLPI ($P = 0.0007$) evidenced a more discriminated pattern than IPI ($P = \text{NS}$) or FLIPI ($P = 0.0044$). MZLPI, which is constructed of relatively simple

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factors, may represent a useful prognostic index for the prediction of PFS and OS in MZL, and may also be used as a substitute for IPI or FLIPI.

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1. Introduction

Non-Hodgkin B-cell Lymphoma (NHL) is an extremely heterogeneous group of disorders that vary markedly in terms of their presenting features and natural history [1].

Marginal zone lymphoma (MZL) is a distinct subgroup of NHL, which typically follows an indolent clinical course and is characterized by prolonged survival rates [2]. The most common type of MZL is mucosa-associated lymphoid tissue (MALT)-type lymphoma. MALT-type MZL has been observed to arise from a variety of extranodal sites, most commonly from the stomach [2,3]. In addition to MALT lymphoma, the entity traditionally designated as monocytoid B-cell lymphoma has been classified by the World Health Organization (WHO) Classification of Hematological Malignancies as nodal marginal zone B-cell lymphoma [4].

In 1993, the International Prognostic Index (IPI) was published [5]. Although originally intended for use in patients with aggressive lymphoma, this index has utility for all types of NHL, and has been extensively applied even in indolent lymphomas [6]. However, it appears to have limited discriminating power, as the majority of patients are allocated to the favorable risk group [6,7]. Recently, a Follicular Lymphoma International Prognostic Index (FLIPI) has been proposed by a collaborative international concern focused on indolent-nature lymphomas [8].

Indolent lymphomas are a heterogeneous group of lymphoproliferative disorders with specific hallmarks consisting of a relatively long natural history, a continuous tendency to relapse, and difficulty to achieve disease eradication [9–11]. This heterogeneity also applies within each histological category, making an accurate assessment of prognostic features at presentation mandatory in these NHL subtypes, due to the broad variability of effective therapeutic options.

Although MZL and FL evidenced the same indolent-nature, each of them is considered to be a distinct disease entity. Nodal involvement has

been observed in a minority of MZL patients, and no evidence has been reported regarding the effects of low hemoglobin values on progression-

Table 1
Patients characteristics ($N = 205$)

Features	Number of patients (%)
Age median (range)	49 (13–82)
Sex (male/female)	104 (50.7)/101 (49.3)
Performance status (ECOG)	
0–1	195 (95.1)
2–4	10 (4.9)
Ann Arbor stage	
I/II	157 (76.6)
III/IV	48 (23.4)
B symptoms	
Absent	201 (98)
Present	4 (2)
Serum LDH level	
Normal	176 (85.9)
Elevated	23 (11.2)
Unknown	6 (2.9)
Hemoglobin	
≥ 12	158 (77.1)
< 12	47 (22.9)
HCV antibody serologic test	
Negative	122 (59.6)
Positive	3 (1.4)
Unknown	80 (39.0)
Bone marrow involvement	
Absent	183 (89.3)
Present	19 (9.3)
Unchecked	3 (1.4)
International Prognostic Index (IPI)	
Low	167 (81.5)
Low-intermediate	18 (8.8)
High-intermediate	12 (5.8)
High	8 (3.9)
Follicular lymphoma IPI	
Good	162 (79.1)
Intermediate	21 (10.2)
Poor	22 (10.7)

ECOG, Eastern Cooperative Oncology Group; LDH, lactic dehydrogenase; HCV, Hepatitis C virus.

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