in RBC or platelet level is not evident, but the increase is resolved during follow-up. “Probable MPN” indicates that the increase in RBC or platelet level continues during follow-up. “Proven MPN” is diagnosed with PV or ET based on the WHO criteria. In total, 1,729 CI patients (1,003 men; 726 women) of median age 73 years (range, 19-96 yr) were reviewed. Thrombocytosis (platelets ≥450×10^9/L) was evident in 69 (4.0%) patients at diagnosis or during follow-up. Reactive thrombocytosis was the most common form of thrombocytosis (N=62, 3.6%). Three (0.2%) patients were considered to exhibit possible ET, and four (0.2%) had proven ET. The causes of reactive thrombocytosis (N=62 patients) included infection (N=59, 95.2%), bleeding (N=1, 1.6%), and iron-deficiency (N=1, 1.6%). Erythrocytosis was evident in 79 (4.6%) patients at diagnosis or during follow-up. Reactive erythrocytosis was the most common form of erythrocytosis (N=50, 2.9%), followed by possible PV (N=21, 1.2%), probable PV (N=6, 0.3%), and proven PV (N=2, 0.1%). None of the 27 patients with possible or probable PV underwent further investigations. Particularly, the JAK2 mutational status was not explored. Reactive erythrocytosis was the most common form of thrombocytosis (N=50) was detected during diagnosis and follow-up in 28 (56.0%) and 22 (44.0%) patients, respectively, and all cases were attributable to hemoconcentration. Of the four patients with proven ET, two lacked any other predisposing factor for thrombosis. All patients with proven ET and PV exhibited multifocal CI and previously undetected infarctions on CI diagnosis.

These results showed that many CI patients with erythrocytosis did not undergo further evaluation in terms of a PV diagnosis and that JAK2 mutational status should be evaluated in such patients. Stroke is a global health problem with a global lifetime risk of approximately 25% in people 25 years and older (as of 2016). People living in East Asia, Central Europe, and Eastern Europe have the highest risk of stroke [6]. In Korea, stroke accounts for roughly 1 out of every 10 deaths, and the proportion of ischemic stroke has steadily increased [7]. To effectively care for patients with PV-associated CI, hematologists should communicate well with neurologists.

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Beta-2 microglobulin as a prognostic factor of primary central nervous system lymphoma

TO THE EDITOR: Primary central nervous system lymphoma (PCNSL) is an extra-nodal non-Hodgkin lymphoma involving the brain, leptomeninges, eyes, or spinal cord and no primary malignancy outside of the central nervous system (CNS). PCNSL is a rare lymphoma and accounts for only 1% of all incident lymphomas [1, 2]. Prevalence of the disease is higher in the sixth to eighth decades of life. The median age at diagnosis of PCNSL is 65 years and the incidence is rising in the elderly population [3, 4].

Among several previously published prognostic models, the International Extranodal Lymphoma Study Group (IELSG) and the Memorial Sloan Kettering Cancer Center (MSKCC) models are the most widely used in current practice [5, 6]. Old age and poor performance status at initial diagnosis were strong poor prognostic factors commonly found in both studies. Meanwhile, serum beta-2 microglobulin (B2MG) is a well-established prognostic factor in multiple myeloma and follicular lymphoma [7, 8].

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the purpose of this study was to validate previously suggested prognostic factors and to evaluate prognostic value of serum B2MG level in PCNSL patients.

**Methods**

**Patients**

We retrospectively analyzed the PCNSL registry data for patients treated from March 1993 to May 2017 at the Asan Medical Center in Seoul, South Korea. Variables that were extracted from the medical records and analyzed included patient demographics, Eastern Cooperative Oncology Group (ECOG) performance status, tumor characteristics, treatment profiles, serum lactate dehydrogenase (LDH) level (normal range <250 IU/L), serum B2MG level (normal range <2.5 µg/dL), cerebrospinal fluid (CSF) total protein level, number of CNS lesions, existence of deep CNS lesions (brain stem, thalamus, basal ganglia, and cerebellum), date of disease progression, and survival status.

**Statistics**

Overall survival (OS) was defined as the time from the beginning of first-line therapy to death from any cause. Univariate and multivariate analyses were performed to identify prognostic factors for OS using a Cox proportional hazards model. Survival curves were estimated by the Kaplan-Meier method and compared using log-rank tests. A two-sided P-value less than 0.05 was considered statistically significant. All statistical analyses were performed using the Statistical Package for the Social Sciences (SPSS) version 21.0 (IBM Corp, Armonk, NY, USA).

**Ethical approval**

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional review board and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. For this type of study, formal consent was not required (IRB 2018-1275).

**Results**

**Clinical characteristics**

In total, 163 patients were identified and included in the analysis. The median follow-up duration was 3.5 years [95% confidence interval (CI), 2.4-4.6] and median OS was 4.0 years (95% CI, 2.1-5.9). Baseline patient characteristics are summarized in Table 1. The median age was 60 years (range, 19-83), 88 patients (54.0%) were 60 years of age or older and 89 patients (54.6%) were males. All patients received high-dose methotrexate-based chemotherapy as initial treatment. Human immunodeficiency virus (HIV) serology was checked in 67 patients at the time of diagnosis and all were negative.

**Prognostic factors**

Univariate analysis of prognostic factors revealed that poor performance status (Eastern Cooperative Oncology Group Performance scale (ECOG PS) ≥2) and elevated serum B2MG (≥1.8 µg/mL) were significantly associated with shorter OS (Table 2). Old age (≥60 yr) showed borderline association in terms of poor OS [hazard ratio (HR) 1.55; 95% CI, 0.95-2.52; P=0.068]. Multivariate analysis results were consistent with HRs of 2.5 (95% CI, 1.06-3.03; P=0.001) for ECOG PS ≥2 and 1.79 (1.06-3.03; P=0.038) for elevated serum B2MG (≥1.8 µg/mL). The survival curves of the two groups according to serum B2MG level are shown on Fig. 1.

**Discussion**

In our analysis, ECOG PS higher than 1 and elevated

| Table 1. Baseline clinical characteristics. |
|------------------------------------------|
| **Variable** | **Patients (%) (N=163)** |
|----------------|-------------------------|
| Age | |
| < 60 | 75 (46.0) |
| ≥ 60 | 88 (54.0) |
| Sex | |
| Male | 89 (54.6) |
| Female | 74 (45.4) |
| Histology | |
| DLBCL | 146 (89.6) |
| Not confirmed | 17 (10.4) |
| ECOG performance status | |
| 0–1 | 116 (71.2) |
| ≥ 2 | 47 (28.8) |
| Serum LDH level | |
| > 250 IU/L | 106 (65.0) |
| ≤ 250 IU/L | 57 (35.0) |
| N of CNS lesion | |
| Single | 77 (50.3) |
| Multiple | 76 (49.7) |
| Existence of deep brain lesion | |
| Yes | 91 (59.5) |
| No | 62 (40.5) |
| CSF protein level | |
| ≥ 68 mg/dL | 48 (35.7) |
| < 68 mg/dL | 92 (45.7) |
| Serum β2 microglobulin level | |
| ≥ 1.8 µg/dL | 39 (23.9) |
| < 1.8 µg/dL | 124 (76.1) |
| Best response after first line treatment | |
| CR | 69 (47.9) |
| PR | 58 (40.3) |
| SD | 10 (0.7) |
| PD | 16 (11.1) |
| Patients who had ASCT | |
| Yes | 58 (35.6) |
| No | 105 (64.4) |

*Not confirmed in histology means lymphoid malignancy without established exact diagnosis among NHL subtypes. Abbreviations: ASCT, autologous stem cell treatment; CNS, central nervous system; CR, complete response; CSF, cerebrospinal fluid; DLBCL, diffuse large B-cell lymphoma; ECOG, Eastern Cooperative Oncology Group; LDH, lactate dehydrogenase; PD, progressive disease; PR, partial response; SD, stable disease.
Table 2. Univariate analysis and multivariate analysis of overall survival.

| Variables                        | Univariate          |               | Multivariate          |               |
|----------------------------------|---------------------|---------------|-----------------------|---------------|
|                                  | HR (95% CI)         | P             | HR (95% CI)           | P             |
| Age                              |                     |               |                       |               |
| < 60                             | Reference           |               | Reference             |               |
| ≥ 60                             | 1.55 (0.95–2.52)    | 0.068         | 1.30 (0.96–1.73)      | 0.044         |
| ECOG performance status          |                     |               |                       |               |
| 0-1                              | Reference           |               | Reference             |               |
| ≥ 2                              | 2.24 (1.38–3.63)    | 0.001         | 2.30 (1.06–3.03)      | 0.001         |
| Serum LDH level                  |                     |               |                       |               |
| ≤ 250 IU/L                       | Reference           |               | Reference             |               |
| > 250 IU/L                       | 1.53 (0.94–2.48)    | 0.089         | 1.44 (0.94–2.20)      | 0.092         |
| Number of CNS lesions            |                     |               |                       |               |
| Multiple                         | Reference           |               | Reference             |               |
| Single                           | 0.88 (0.54–1.44)    | 0.616         | 1.10 (0.67–1.81)      | 0.688         |
| Presence of deep CNS lesion      |                     |               |                       |               |
| Yes                              | Reference           |               | Reference             |               |
| No                               | 1.15 (0.70–1.89)    | 0.583         | 1.27 (0.82–1.97)      | 0.270         |
| CSF protein level                |                     |               |                       |               |
| ≥ 68 mg/dL                       | Reference           |               | Reference             |               |
| < 68 mg/dL                       | 1.18 (0.68–2.04)    | 0.552         | 1.27 (0.75–2.14)      | 0.399         |
| Serum β2 microglobulin           |                     |               |                       |               |
| < 1.8 μg/dL                      | Reference           |               | Reference             |               |
| ≥ 1.8 μg/dL                      | 1.70 (1.01–2.88)    | 0.047         | 1.79 (1.06–3.03)      | 0.038         |

Abbreviations: CI, confidence interval; CNS, central nervous system; CSF, cerebrospinal fluid; ECOG, Eastern Cooperative Oncology Group; HR, hazard ratio; LDH, lactate dehydrogenase; OS, overall survival; PFS, progression-free survival.

Fig. 1. Overall survival from the initiation of first-line treatment.

serum B2MG level were significantly predictive of poor prognosis according to univariate analysis. Most studies defined a cutoff level of serum B2MG level between 2.0 to 3.5 and our analysis showed 1.8 as the best cutoff level to establish a significant survival benefit [7-9]. Although the mechanism underlying the negative prognostic impact of elevated serum B2MG is unclear, a widely accepted hypothesis is that it is related to high tumor burden [9, 10]. Based on this hypothesis, we might explain the relatively low cutoff level of B2MG in PCNSL by reiterating that even a small elevation of serum B2MG could reflect high tumor burden in the CNS. Our results are reliable considering the large number of enrolled patients and comparable survival outcomes of our cohort with other studies [6]. In the MSKCC study, 240 patients were enrolled, and the median OS and failure-free survival were 37 (95% CI, 31–42) months and 17 (95% CI, 12–21) months, respectively.

Our study has some limitations including the retrospective nature and also it is a single center study. Another limitation is that the cut-off value of B2MG level 1.8 μg/dL is arbitrary. However, the cutoff value in this study represents only a population from single center. Higher serum B2MG level could have some relationship with survival outcomes in PCNSL and further investigations via multi-center studies are needed. In conclusion, ECOG performance status and serum B2MG were associated with prognosis in PCNSL patients. Serum B2MG may have some association with prognosis of PCNSL.

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TO THE EDITOR: Myelodysplastic syndromes (MDS) are a group of heterogeneous hematological malignancies which demand personalized and risk-adapted clinical management [1]. Current therapeutic approaches are rather limited for patients unsuitable for allogeneic stem cell transplantation (SCT), the only realistic and potentially curative treatment measure that exists [1]. With regard to patients with high-risk MDS, the standard of care is currently represented by treatment with hypomethylating agents (HMAs), such as decitabine and azacitidine. The latter is used as initial therapy in most cases, and induces responses in 40–50% of treated patients [2, 3]. Obstacles to azacitidine administration as well as recommendations for the optimization of treatment with this agent have been reported [2, 4]. However, despite optimal management of azacitidine treatment, the duration of its clinical benefit, although variable, is usually transient and almost all patients ultimately experience loss of response to the drug, disease progression, and therefore very poor outcomes [1, 2, 5, 6]. After this loss of response or disease progression despite treatment, there are no standard care regimens available [5]. Rescue strategies including intensive chemotherapy (ICT) only provide minor benefits, whereas allogeneic SCT is feasible only in a minority of cases. With these results in mind, especially the catastrophic outcome of azacitidine-failed patients, typical concerns about decision making and clinical management in these settings can be summarized by an unusual case we observed which is reported herein. A 59-year-old woman was admitted for profound malaise due to pancytopenia on March 2015. The bone marrow (BM) and trephine biopsy revealed refractory anemia with an excess of blasts-2 (RAEB-2), remarkable multilineage dysplasia, and 18% of BM infiltrating blasts; the karyotype analysis and molecular study for typical abnormalities found in MDS were negative. She was diagnosed as having an Inter-2 MDS, according to the International Prognostic Scoring System [7]. On the basis of the patient’s overall fitness level, and given the lack of a suitable familiar donor to proceed to immediate allogeneic SCT, the only realistic and potentially curative treatment measure that exists [1]. With regard to patients with high-risk MDS, the standard of care is currently represented by treatment with hypomethylating agents (HMAs), such as decitabine and azacitidine. The latter is used as initial therapy in most cases, and induces responses in 40–50% of treated patients [2, 3]. 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