Original article

Primary testicular lymphoma—A single center experience and review of literature

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ABSTRACT

Objective: The purpose of this study is to evaluate the clinical characteristics, pathology, treatment outcomes, and survival of primary testicular lymphoma (PTL) patients treated at our hospital after 1990. Related literature was reviewed.

Materials and methods: We retrospectively enrolled patients diagnosed with PTL between January 1990 and September 2013 in our institute. Clinical features, pathology, and overall survival were analyzed.

Results: 24 patients were enrolled. They had a mean age at diagnosis of 65.0 years (range 10–84 y), mean follow-up duration 57.0 months (range 3–182 mo), and median overall survival of 38 months (range 4.0–184.7 mo). The most common pathology subtype was diffuse large B-cell lymphoma (n = 21, 87.5%). Fourteen patients (58.3%) achieved complete remission of disease and five patients (20.8%) achieved partial remission after treatment. Three patients had a relapse of disease after complete remission: one in the brain parenchyma, one in the pelvis soft tissue and omentum, and one in the left pyriform muscle. Three patients had metastasis after partial remission of disease: two in the brain parenchyma and one in the cauda equina. By univariate analysis, the factors significantly associated with superior overall survival were primary tumor diameter < 7.5 cm, serum lactate dehydrogenase ≤ 250 U/L, Ann-Arbor Stage IE/II, and International Prognostic Index (IPI) ≤ 1 (p < 0.05). Rituximab-containing treatment did not show overall survival benefits in our series. By multivariate analysis, IPI ≤ 1 showed statistical significance (p = 0.019), suggesting a potential prognostic value of IPI in evaluating PTL patients.

Conclusions: The overall survival of PTL patients is poor, especially those with extensive disease (Stage III/IV). The IPI have a prognostic role in PTL. The use of rituximab in the treatment regimen of PTL does not seem to improve survival in our series.

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1. Introduction

Primary testicular lymphoma (PTL) is a rare but aggressive malignant tumor. It constitutes only 1–2% of non-Hodgkin lymphoma, and population-based studies have estimated the incidence at 0.09–0.26/100,000 per year.1 However, lymphoma remains the most common testicular neoplasm in men > 60 years of age. The usual presentation is a painless testis mass which grows slowly over weeks or months, with either unilateral or bilateral involvement. Typically, PTL is diagnosed after radical orchiectomy, and the most frequent histological subtype is diffuse large B-cell lymphoma (DLBCL), although other histological subtypes have been described.2

Currently, there are no gold standard treatment guidelines for PTL. The available data to date are from nonrandomized studies or retrospective series. Generally, the common treatment strategies include radical orchiectomy, followed by chemotherapy and/or radiotherapy. Rituximab, a chimeric monoclonal antibody against
the protein CD20, has proven therapeutic effects in nodal DLBCL. However, its impact on outcomes in PTL remains unclear, and may be less definitive than in nodal DLBCL.1

In this retrospective study, we evaluated the clinical characteristics, pathology, and treatment outcomes of 24 PTL patients treated at our hospital after 1990. Related literature was also reviewed for a better understanding of this rare malignancy.

2. Materials and methods

We retrospectively enrolled patients diagnosed with PTL between January 1990 and September 2013 in our institute (Taipei Veterans General Hospital, Taiwan). The diagnosis of PTL was made if the testes were the primary site of the disease or at least the main site of involvement, without or with only minor nodal involvement.1 Patients with a lymphoma history and those lost to follow-up were excluded. All patients underwent computed tomography (CT) scan from the neck to pelvis, whole-body bone scan, and bone marrow biopsy for staging. Potential prognostic factors, including age, primary tumor diameter, serum lactate dehydrogenase (LDH), Ann-Arbor stage, International Prognostic Index (IPI) score, and the use of rituximab-containing regimen, were evaluated. IPI is a clinical tool developed by oncologists to aid in predicting the prognosis of patients with aggressive non-Hodgkin lymphoma. One point is assigned for each of the following risk factors: age > 60 years, Stage III/IV disease, elevated serum LDH, Eastern Cooperative Oncology Group (ECOG) performance status > 1, and more than one extranodal site. Based on IPI scores, patients could be classified into low-risk group (IPI < 1) or intermediate—high-risk group (IPI ≥ 2). The Ann-Arbor staging is the staging system for lymphomas. Stage I indicated lymphoma involving only a single lymph node (Stage I) or a single extralymphatic organ (Stage IE); Stage II indicated lymphoma involving two separate regions, with both the affected areas being confined to one side of the diaphragm; Stage III indicated that lymphoma has spread to both sides of the diaphragm; and Stage IV indicated diffuse or disseminated involvement of one or more extralymphatic organs, including any involvement of the liver, bone marrow, or lung. Patients received further treatment including chemotherapy with or without rituximab, intrathecal chemotherapy, and radiotherapy to the contralateral testes. The standard chemotherapy regimen is CHOP before the rituximab era and R-CHOP after the rituximab era. The standard R-CHOP regimen is as follows: rituximab 700 mg, cyclophosphamide 750 mg/m², doxorubicin 50 mg/m², oncovin 1.4 mg/m², and prednisolone 40 mg by mouth twice daily from Days 2 to 5; the regimen was given every 3 weeks. Doxorubicin may be replaced by etoposide 50 mg/m² if the patient’s cardiac function is poor. The standard intrathecal regimen for radiation is 30 Gy. The overall survival (OS) was calculated from the date of diagnosis to the time of mortality, or the time of last follow-up for survivors. The Kaplan–Meier survival curve was plotted for patients of each group, and the differences between the curves were examined using the log-rank test. Statistical analyses were performed using SPSS version 19.0 (SPSS, Chicago, IL, USA).

3. Results

From January 1990 to September 2013, 24 patients with PTL were diagnosed and treated in our institute. Their mean age at diagnosis was 65.0 years (range 10—84 y), mean follow-up duration was 57.0 months (range 3—182 mo), and median OS was 38 months (range 4.0—184.7 mo). Radical orchiectomy was performed in 23 patients, and only one patient received open testis biopsy for diagnosis. The most common pathology subtype is DLBCL (n = 21, 87.5%). Patient’s clinical characteristics are summarized in Table 1.

3.1. Treatment response

The treatment modalities and outcomes are summarized in Table 2. Of the 24 patients, two (Cases 13 and 22) did not receive further treatment due to poor performance status and rapid disease progression resulting in mortality. The remaining 22 patients received further systemic chemotherapy, with or without intrathecal central nervous system (CNS) prophylaxis or contralateral testis prophylactic irradiation. Fourteen patients (58.3%) had complete remission of disease, but three patients had disease relapse, including one with brain metastasis after 7 years, one with left pyriform muscle metastasis at 11 months, and one with metastasis to the pelvis soft tissue and omentum at 16 months. Five patients (20.8%) had partial remission of disease, with three patients had disease progression, including two with brain metastasis at 9 months and 13 months, respectively, and one with cauda equina metastasis at 6 months.

3.2. Prognostic factors

Patients were grouped according to age, primary tumor diameter, serum LDH level, Ann-Arbor stage, IPI scores, and rituximab-containing regimen. Based on the Ann-Arbor staging system, patients were separated into those with limited disease (Stage IE/II) or extensive disease (Stage III/IV). According to IPI scores, patients could be classified into low-risk group (IPI ≤ 1) or intermediate–high-risk group (IPI ≥ 2). The statistical results are summarized in Table 3 and Figure 1. By univariate analysis, the factors significantly associated with a superior OS were primary tumor diameter ≤ 7.5 cm, serum LDH ≤ 250 U/L, and limited and low-risk disease (p < 0.05). Treatment with rituximab-containing regimen did not show OS benefit in our series. By multivariate analysis, only those with low-risk disease (IPI ≤ 1) showed significance (p = 0.019), suggesting that IPI score had prognostic value in evaluating patients with PTL.

Table 1

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Patient number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td></td>
</tr>
<tr>
<td>&gt; 60</td>
<td>16 (66.7)</td>
</tr>
<tr>
<td>≤ 60</td>
<td>8 (33.3)</td>
</tr>
<tr>
<td>Location</td>
<td></td>
</tr>
<tr>
<td>Left</td>
<td>11 (45.8)</td>
</tr>
<tr>
<td>Right</td>
<td>11 (45.8)</td>
</tr>
<tr>
<td>Bilateral</td>
<td>2 (8.3)</td>
</tr>
<tr>
<td>Histology</td>
<td></td>
</tr>
<tr>
<td>DLBCL</td>
<td>21 (87.5)</td>
</tr>
<tr>
<td>T-cell lymphoma</td>
<td>1 (4.2)</td>
</tr>
<tr>
<td>Burkitt’s lymphoma</td>
<td>2 (8.3)</td>
</tr>
<tr>
<td>Ann-Arbor stage</td>
<td></td>
</tr>
<tr>
<td>IE</td>
<td>10 (41.7)</td>
</tr>
<tr>
<td>II</td>
<td>7 (29.1)</td>
</tr>
<tr>
<td>III</td>
<td>1 (4.2)</td>
</tr>
<tr>
<td>IV</td>
<td>6 (25.0)</td>
</tr>
<tr>
<td>IPI</td>
<td></td>
</tr>
<tr>
<td>Low (0—1)</td>
<td>10 (41.7)</td>
</tr>
<tr>
<td>Low–intermediate (2)</td>
<td>5 (8.3)</td>
</tr>
<tr>
<td>High–intermediate (3)</td>
<td>6 (25.0)</td>
</tr>
<tr>
<td>High (4–5)</td>
<td>3 (12.5)</td>
</tr>
</tbody>
</table>

DLBCL — diffuse large B-cell lymphoma; IPI — International Prognostic Index.
Hodgkin’s and non-Hodgkin lymphomas, and is widely used by
diagnosis of bilateral testicular mass. In this report, we presented 24 patients with PTL treated in our
hospital after 1990. The mean patient age at diagnosis was
65.0 years, and the most pathological subtype is DLBCL. Typical
presentation is a unilateral or bilateral painless testicular swelling
and mass. The scrotal sonography of testicular lymphoma might
predict the prognosis of patients with aggressive non-Hodgkin
lymphoma. Jia et al. reported that, besides IPI, a primary testicular
tumor diameter larger than 7.5 cm was a poor prognostic factor for
PTL patients. In our series, the OS was different between low-risk
patients. The OS was also different between patients with a primary
testicular mass with increased blood flow. In our series, radical
orchiectomy was performed for both diagnostic and therapeutic
purposes, and only one patient received open testis biopsy for the
diagnosis of bilateral testicular mass.

Ann-Arbor staging is the staging system for lymphomas, both in
Hodgkin’s and non-Hodgkin lymphomas, and is widely used by
oncologists. Ahmad et al. reported that up to 90% of patients have
Stage I or II disease at diagnosis (60% and 30%, respectively), and
35% had bilateral testicular involvement. In our series, 41.6% of
patients was diagnosed Stage I disease and 29.2% was diagnosed
with Stage II disease. Bilateral testis involvement was only seen in
8.3% of patients. The OS of PTL is poor, especially in those with
extensive disease (Stage III/IV).

Like Ann-Arbor stage, IPI was also developed by oncologists to
predict the prognosis of patients with aggressive non-Hodgkin
lymphoma. Jia et al. reported that, besides IPI, a primary testicular
tumor diameter larger than 7.5 cm was a poor prognostic factor for
PTL patients. In our series, the OS was different between low-risk
group (IPI ≤ 1) and intermediate–high-risk group (IPI ≥ 2) pa-
tients. The OS was also different between patients with a primary
testis tumor diameter of smaller than 7.5 cm and those with tu-
mors larger than 7.5 cm. By multivariate analysis, only those with
IPI ≤ 1 had significantly better survival (p = 0.019) in our series,
indicating that IPI score still had its predictive value in evaluating
PTL patients.

The optimal management of PTL has not yet been established.
Several studies have demonstrated that orchiectomy alone should
not be considered as the sole treatment in patients with Stage I
disease. Early studies have reported that if these patients were
treated with surgery alone, most would experience relapse within
2 years, implying that in PTL, widespread microscopic disease is
present at diagnosis. Furthermore, relapse in the contralateral
testis occurred in 5–35% of PTL patients. The International
Extranodal Lymphoma Study Group had conducted a large

### Table 2
Clinical characteristics, treatment methods, and outcome of 24 patients with primary testicular lymphoma.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (y)</th>
<th>Site</th>
<th>Pathology</th>
<th>Ann-Arbor stage</th>
<th>IPI score</th>
<th>Initial management</th>
<th>Subsequent treatment</th>
<th>Follow-up (mo), status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>58</td>
<td>Left</td>
<td>DLBCL</td>
<td>IV</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>34, expired</td>
</tr>
<tr>
<td>2</td>
<td>59</td>
<td>Right</td>
<td>T-cell lymphoma</td>
<td>III</td>
<td>2</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>6, expired</td>
</tr>
<tr>
<td>3</td>
<td>80</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>2</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>93, expired</td>
</tr>
<tr>
<td>4</td>
<td>71</td>
<td>Right</td>
<td>DLBCL</td>
<td>II</td>
<td>2</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>89, expired</td>
</tr>
<tr>
<td>5</td>
<td>33</td>
<td>Right</td>
<td>DLBCL</td>
<td>IV</td>
<td>3</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>16, expired</td>
</tr>
<tr>
<td>6</td>
<td>27</td>
<td>Bilateral</td>
<td>DLBCL</td>
<td>IV</td>
<td>3</td>
<td>Biopsy</td>
<td>C/T</td>
<td>4, expired</td>
</tr>
<tr>
<td>7</td>
<td>67</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>65, expired</td>
</tr>
<tr>
<td>8</td>
<td>68</td>
<td>Left</td>
<td>Burkitt's lymphoma</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>182, alive</td>
</tr>
<tr>
<td>9</td>
<td>68</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>0</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>180, alive</td>
</tr>
<tr>
<td>10</td>
<td>70</td>
<td>Right</td>
<td>DLBCL</td>
<td>IV</td>
<td>4</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>9, expired</td>
</tr>
<tr>
<td>11</td>
<td>34</td>
<td>Bilateral</td>
<td>DLBCL</td>
<td>IE</td>
<td>2</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>23, expired</td>
</tr>
<tr>
<td>12</td>
<td>10</td>
<td>Right</td>
<td>DLBCL</td>
<td>II</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T + IT</td>
<td>158, alive</td>
</tr>
<tr>
<td>13</td>
<td>79</td>
<td>Right</td>
<td>DLBCL</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>Nil</td>
<td>42, expired</td>
</tr>
<tr>
<td>14</td>
<td>80</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>137, alive</td>
</tr>
<tr>
<td>15</td>
<td>72</td>
<td>Left</td>
<td>DLBCL</td>
<td>II</td>
<td>3</td>
<td>Radical orchiectomy</td>
<td>C/T with rituximab + R/T</td>
<td>21, expired</td>
</tr>
<tr>
<td>16</td>
<td>84</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T</td>
<td>120, alive</td>
</tr>
<tr>
<td>17</td>
<td>64</td>
<td>Left</td>
<td>Burkitt's lymphoma</td>
<td>IV</td>
<td>4</td>
<td>Radical orchiectomy</td>
<td>C/T + IT + R/T</td>
<td>5, expired</td>
</tr>
<tr>
<td>18</td>
<td>78</td>
<td>Right</td>
<td>DLBCL</td>
<td>IV</td>
<td>3</td>
<td>Radical orchiectomy</td>
<td>C/T with rituximab + IT</td>
<td>50, expired</td>
</tr>
<tr>
<td>19</td>
<td>57</td>
<td>Right</td>
<td>DLBCL</td>
<td>IE</td>
<td>0</td>
<td>Radical orchiectomy</td>
<td>C/T + rituximab + IT</td>
<td>70, expired</td>
</tr>
<tr>
<td>20</td>
<td>80</td>
<td>Right</td>
<td>DLBCL</td>
<td>II</td>
<td>4</td>
<td>Radical orchiectomy</td>
<td>C/T + rituximab + IT</td>
<td>4, expired</td>
</tr>
<tr>
<td>21</td>
<td>79</td>
<td>Right</td>
<td>DLBCL</td>
<td>II</td>
<td>3</td>
<td>Radical orchiectomy</td>
<td>C/T with rituximab</td>
<td>5, expired</td>
</tr>
<tr>
<td>22</td>
<td>73</td>
<td>Right</td>
<td>DLBCL</td>
<td>IE</td>
<td>2</td>
<td>Radical orchiectomy</td>
<td>Nil</td>
<td>3, expired</td>
</tr>
<tr>
<td>23</td>
<td>58</td>
<td>Left</td>
<td>DLBCL</td>
<td>IE</td>
<td>1</td>
<td>Radical orchiectomy</td>
<td>C/T with rituximab + IT</td>
<td>44, alive</td>
</tr>
<tr>
<td>24</td>
<td>81</td>
<td>Right</td>
<td>DLBCL</td>
<td>II</td>
<td>3</td>
<td>Radical orchiectomy</td>
<td>C/T with rituximab</td>
<td>7, expired</td>
</tr>
</tbody>
</table>

C/T – chemotherapy; DLBCL – diffuse large B-cell lymphoma; IPI – International Prognostic Index; IT – intrathecal chemotherapy; R/T – contralateral testis irradiation.

### Table 3
Results of univariate and multivariate analyses of prognostic factors of overall survival.

<table>
<thead>
<tr>
<th>Variables</th>
<th>n</th>
<th>Categorization</th>
<th>Univariable</th>
<th>Multivariable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (y)</td>
<td>16</td>
<td>60</td>
<td>1.413, 0.503–3.972</td>
<td>0.512</td>
</tr>
<tr>
<td>Tumor diameter (cm)</td>
<td>17</td>
<td>≤ 7.5</td>
<td>3.192, 1.140–8.934</td>
<td>0.027</td>
</tr>
<tr>
<td>LDH (U/L)</td>
<td>14</td>
<td>≤ 250</td>
<td>2.678, 1.013–10.778</td>
<td>0.047</td>
</tr>
<tr>
<td>Ann-Arbor stage</td>
<td>17</td>
<td>II/IV</td>
<td>3.968, 1.347–11.689</td>
<td>0.012</td>
</tr>
<tr>
<td>IPI score</td>
<td>10</td>
<td>≤ 1</td>
<td>6.671, 2.129–20.898</td>
<td>0.001</td>
</tr>
<tr>
<td>Rituximab</td>
<td>17</td>
<td>Not contained</td>
<td>1.900, 0.682–5.291</td>
<td>0.220</td>
</tr>
</tbody>
</table>

CI – confidence interval; HR – hazard ratio; IPI – International Prognostic Index; LDH – lactate dehydrogenase.

### 4. Discussion

In this report, we presented 24 patients with PTL treated in our
hospital after 1990. The mean patient age at diagnosis was
65.0 years, and the most pathological subtype is DLBCL. Typical
presentation is a unilateral or bilateral painless testicular swelling
or mass. The scrotal sonography of testicular lymphoma might
appear as a diffusely hypoechoic but hypereoptic testicle or a focal
testicular mass with increased blood flow. In our series, radical
orchiectomy was performed for both diagnostic and therapeutic
purposes, and only one patient received open testis biopsy for the
diagnosis of bilateral testicular mass.

Ann-Arbor staging is the staging system for lymphomas, both in
Hodgkin’s and non-Hodgkin lymphomas, and is widely used by
oncologists. Ahmad et al. reported that up to 90% of patients have
Stage I or II disease at diagnosis (60% and 30%, respectively), and
35% had bilateral testicular involvement. In our series, 41.6% of
patients was diagnosed Stage I disease and 29.2% was diagnosed
with Stage II disease. Bilateral testis involvement was only seen in
8.3% of patients. The OS of PTL is poor, especially in those with
extensive disease (Stage III/IV).
retrospective study and a prospective study for PTL treatment. These studies have the best evidence to date. The studies promoted combined treatment for PTL regardless of stages, including radical orchectomy, followed by anthracycline-based chemotherapy (CHOP) plus rituximab, intrathecal methotrexate CNS prophylaxis, and contralateral prophylactic testicular irradiation. Nonetheless, CNS prophylaxis warrants further investigation. The best strategy to prevent CNS relapse is still debatable, and a new prospective study by the International Extranodal Lymphoma Study Group is ongoing.

In our series, two patients did not receive any further treatment after radical orchectomy due to poor performance status and rapid disease progression resulting in mortality. The other 22 patients received further systemic chemotherapy. Only five patients received intrathecal prophylaxis. In our institute, intrathecal chemotherapy prophylaxis was still not routinely performed as the first-line treatment, since intrathecal chemotherapy has its limitations. First, in PTL patients with CNS relapse, the brain parenchyma is the most frequent relapse or metastasis site rather than the meninges. Hence, prophylactic intrathecal chemotherapy might not be effective. In our series, only one patient suffered from brain parenchyma relapse after complete remission. Three patients with partial remission had further CNS metastasis, including two in the brain parenchyma and one in the cauda equina. The relapse or metastasis sites were not the meninges. Second, PTL patients are usually elderly, who may have degenerated spine disease causing...
difficulty in performing lumbar puncture, and they may not tolerate aggressive CNS prophylaxis. In addition, lumbar puncture may cause complications, such as hemorrhage and infection, and may be fatal in severe conditions.

Rituximab, a chimeric monoclonal antibody against the protein CD20, was approved by the United States Food and Drug Administration in 1997 to treat B-cell non-Hodgkin lymphoma resistant to chemotherapy regimens. The combination of rituximab and CHOP chemotherapy was later proven to be superior to CHOP alone as treatment of DLBCL, and is currently used as the first-line treatment of DLBCL. However, although rituximab had improved the outcome of DLBCL, its impact on the outcome of PTL remained unclear. Gundrum et al. conducted a study using the Surveillance, Epidemiology, and End Results database to analyze the potential impact of the use of rituximab on PTL survival. The study concluded that the use of rituximab in clinical practice did not seem to improve the first 5-year outcome. In our series, seven patients received rituximab-containing chemotherapy, but these patients did not show better OS compared with those receiving traditional anthracycline-based chemotherapy.

In the current rituximab era, relapse is still the major failure pattern in patients with PTL. In recent years, some studies using gene expression assays have been published to investigate any unique gene expression in primary testicular DLBCL, which may serve as a potential target for immunotherapy or anti-cancer drug therapy. The distinctive gene expression profiling signature of primary testicular DLBCL was relevant to upregulation of some oncogenes and genes encoding components of B-cell receptor signaling and proliferation, and downregulation of some anti-oncogenes. Results of these protein or gene level analysis suggested that new agents may be promising for primary testicular DLBCL patients in the future.

This retrospective study has limitations. The study population is small and the treatment protocols are diverse, making it difficult to analyze relevant treatment prognostic factors.

In conclusion, the major pathology of PTL is DLBCL, and most patients are diagnosed in Ann Arbor Stage IE/II disease. The OS is poor, especially in those with extensive disease (Stage III/IV). In addition to radical orchiectomy for local control, systemic chemotherapy is still needed. In this study, the CNS relapse site is the brain parenchyma, so prophylactic intrathecal chemotherapy may not be beneficial to PTL patients. IPI is prognostic in evaluating PTL patients and the OS is better in low-risk group patients (IPI ≤ 1). The use of rituximab in the treatment regimen does not seem to improve the survival in our series.

Conflicts of interest

The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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References