Primary central nervous system lymphomas (PCNSL) are rare and aggressive type of non-Hodgkin lymphomas which form about 2-3% of all CNS tumors. We undertook a retrospective study of PCNSL at our Institute over a period of 6 years, wherein the clinical and immunomorphological features of PCNSL were studied. There were 27 cases of PCNSL, males were more commonly affected with an age range of 12-68 years. Almost all were B cell lymphomas and diffuse large B cell lymphoma (DLBCL) was the commonest type, with most showing centroblastic morphology. Further subtyping into germinal centre B cell (GCB) and non-germinal centre B cell (NGCB) groups by IHC was possible in 18 cases using Hans algorithm, which showed higher number of NGCB cases. There were one case each of anaplastic large cell lymphoma and follicular lymphoma.

**ABSTRACT**

Primary central nervous system lymphomas (PCNSL) are rare and aggressive type of non-Hodgkin lymphomas which form about 2-3% of all CNS tumors. We undertook a retrospective study of PCNSL at our Institute over a period of 6 years, wherein the clinical and immunomorphological features of PCNSL were studied. There were 27 cases of PCNSL, males were more commonly affected with an age range of 12-68 years. Almost all were B cell lymphomas and diffuse large B cell lymphoma (DLBCL) was the commonest type, with most showing centroblastic morphology. Further subtyping into germinal centre B cell (GCB) and non-germinal centre B cell (NGCB) groups by IHC was possible in 18 cases using Hans algorithm, which showed higher number of NGCB cases. There were one case each of anaplastic large cell lymphoma and follicular lymphoma.

**KEYWORDS**

Primary CNS lymphomas; DLBCL; ALCL; Follicular lymphoma
Primary CNS lymphomas are rare, aggressive neoplasms with distinctive clinicopathological features and with a poor outcome in most of the cases. We evaluated the demographic, histomorphologic and immunophenotypic profile of PCNSL at a regional cancer centre in South India and compared the trend occurring in other parts of India and other countries.

Demographic features: The mean age at presentation in our study was 40.9 yrs which was similar to many Indian studies [3,6,9,11,12,16,18]. However in the study by Mahadevan et al. [7] and some western studies the mean age of occurrence was slightly older [8,11,12]. There was male preponderance in our study which is similar to most Indian studies, however a few western studies have shown a female preponderance [8,11,12]. All the studies including ours showed supratentorial region as the commonest site of involvement by the neoplasm. All the patents in this study were immunocompetent and negative for serologic tests for HIV 1 and 2, which was the case in most other studies except a few studies which had a few HIV positive cases [17].

One case of ALCL in our study occurred in a 14 year old immunocompetent boy in the dura mater of the frontal region without any evidence of the disease elsewhere. Very few primary dural follicular lymphomas reported in the literature, which showed similar features with female preponderance [20].

Histopathology: Most of the cases of DLBCL in our study were of the centroblastic type on morphology, which correlated with other studies with where morphologic subtyping was done [29,30]. The case of ALCL in our study resembled any other ALCL at nodal or extranodal site and was similar to the case series reports by George et al. [17]. CNS follicular lymphoma in our study was low grade (grade 2 of 3), which was similar to the study by Beral et al. [11].

Immunohistochemistry: DLBCLs can be subtyped into GCB and NGCB types using IHC markers, which has prognostic significance using any of the algorithms available of which the Hans algorithm [19] is the widely used and makes use of the IHC markers, CD 10, BCL6 and using any of the algorithms available of which the Hans algorithm is the most common subtype forming 77.8% (14 cases). Most other Indian and international studies have also shown that NGCB subtype is the commonest type of DLBCL in the CNS though the proportion of NGCB varied from 64.7% to 96.3% in different studies which are summarised in Table 1. The relative proportion of GCB subtype is higher in Asian studies as compared to the western studies [7,10,11,12,16,18].

Table 1: Comparison of GCB vs NGCB in various studies with the present study.

<table>
<thead>
<tr>
<th>Study group/year</th>
<th>Country / Study duration</th>
<th>Total no. of cases</th>
<th>GCB</th>
<th>NGCB</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sharma et al. [2016]</td>
<td>India / 10yrs</td>
<td>64</td>
<td>13 (20.3%)</td>
<td>51 (79.7%)</td>
</tr>
<tr>
<td>Patel et al. [2015]</td>
<td>India / 12yrs</td>
<td>51</td>
<td>18 (35.3%)</td>
<td>33 (64.7%)</td>
</tr>
<tr>
<td>Mahadevan et al. [2015]</td>
<td>India / 12yrs</td>
<td>24</td>
<td>22 (91.6%)</td>
<td>2 (8.4%)</td>
</tr>
<tr>
<td>Aki et al. [2013]</td>
<td>Turkey / 15yrs</td>
<td>39</td>
<td>6 (15.4%)</td>
<td>33 (84.6%)</td>
</tr>
<tr>
<td>Hattab et al. [2010]</td>
<td>USA / 7yrs</td>
<td>51</td>
<td>5 (10%)</td>
<td>46 (90%)</td>
</tr>
<tr>
<td>Roux et al. [2010]</td>
<td>France / 12yrs</td>
<td>29</td>
<td>13 (25.7%)</td>
<td>16 (74.3%)</td>
</tr>
<tr>
<td>Preuser et al. [2019]</td>
<td>Austria / 39yrs</td>
<td>75</td>
<td>3 (4%)</td>
<td>72 (96%)</td>
</tr>
<tr>
<td>Bhagavath et al. [2008]</td>
<td>USA / 20yrs</td>
<td>21</td>
<td>2 (5.5%)</td>
<td>19 (90.5%)</td>
</tr>
<tr>
<td>Broot et al. [2006]</td>
<td>France / 8yrs</td>
<td>82</td>
<td>3 (3.7%)</td>
<td>79 (96.3%)</td>
</tr>
<tr>
<td>Lin et al. [2016]</td>
<td>Taiwan / 12yrs</td>
<td>51</td>
<td>11 (21.6%)</td>
<td>40 (78.4%)</td>
</tr>
<tr>
<td>Present study</td>
<td>India / 6yrs</td>
<td>18</td>
<td>4 (22.2%)</td>
<td>14 (77.8%)</td>
</tr>
</tbody>
</table>

ALK positive ALCLs of the CNS, typically express CD 30 and ALK1 on IHC, and occur in patients younger than 18 years, and carry a good prognosis like ALCL at any other location [15]. The case of ALCL in our study is also disease free after completing chemotherapy and is on regular follow up.

Follicular lymphomas of the CNS are very rare and IHC is useful in differentiating them from other low grade lymphomas and meningiomas, particularly when they are dura based [17].

Conclusions: In this 6 year retrospective study of 27 cases of PCNSL, presenting at a regional cancer centre in South India, all the patients were immunocompetent with DLBCL being the commonest type of primary CNS lymphoma, and NGCB type was the most common among DLBCL. The proportion of GCB type to NGCB type was higher in our study which is similar to other Asian studies. There was one case each of ALCL and FL with typical morphoimmunological features.

References


