Primary central nervous system lymphoma (PCNSL) is rare in adults, and no standardized treatment options have been defined. Systemic chemotherapy with a high-dose methotrexate backbone is the mainstay of induction. Approaches to consolidation have consisted of whole-brain radiotherapy or high-dose chemotherapy with autologous stem cell transplantation (HDC-ASCT).\(^1\)

PCNSL is exceedingly rare in children, with approximately 100 cases described over the last 20 years.\(^2,3\) As with adults, diffuse large B-cell lymphoma remains the most frequent histological subtype; however, in children, a greater proportion is represented by other subtypes, such as anaplastic large cell lymphoma (ALCL) and Burkitt lymphoma. Overall, fewer than 30 cases of pediatric primary central nervous system (CNS) ALCL have been described in the literature. Although there have been heterogeneous approaches to treatment, a significant number of children with primary CNS ALCL have received radiotherapy as part of first-line therapy.\(^4,5\) However, there is concern about the use of whole-brain radiotherapy in the upfront setting in children because of the well-recognized sequelae of radiotherapy on the developing brain.

We have read with interest the study by DeFilipp et al.\(^6\) who describe favorable outcomes for adults with PCNSL in first complete remission who underwent HDC-ASCT with a CNS-directed thiotepa, busulfan, and cyclophosphamide (TBC) conditioning regimen. Our experience supports the feasibility of such an approach in children. A 4-year-old immunocompetent boy presented with right-sided facial droop, headaches, and right-sided upper limb hemiparesis. Brain magnetic resonance imaging revealed a 30-mm \(\times\) 32-mm \(\times\) 34-mm left basal ganglia mass, which was confirmed to be primary CNS ALCL after full-staging investigations and stereotactic biopsy. Induction therapy consisted of 4 courses of methotrexate (3.5 g/m\(^2\) on day 1) and cytarabine (2 g/m\(^2\) twice daily on days 2 and 3), with each course administered every 3 weeks in accordance with the results of a randomized phase 2 trial of the International Extranodal Lymphoma Study Group.\(^7\) Complete radiological remission was achieved after the second cycle of therapy. To avoid the use of whole-brain irradiation, which has frequently been administered to children with primary CNS ALCL, HDC-ASCT was administered as consolidation therapy. The same TBC conditioning regimen described by DeFilipp et al was delivered, except that the cumulative daily busulfan dose was prescribed as a single daily intravenous infusion; pharmacokinetic dose adjustments were made after the first dose to achieve an area under the curve of 4800 \(\mu\)mol min/L. Treatment was well tolerated without any significant complications, and the child remains in complete clinical and radiological remission 18 months after the completion of therapy. In comparison with a baseline assessment, complete neuropsychological evaluation 1 year after the completion of therapy demonstrated improvements in general cognitive functioning, adaptive behavior, and quality of life, with psychosocial functioning remaining constant. This indicates the potential for preserving neurocognitive function with this treatment approach.

In conclusion, our findings support the feasibility of HDC-ASCT with TBC conditioning, as reported by DeFilipp et al.\(^6\) as radiation-sparing consolidation therapy for primary CNS ALCL in children. As with all rare childhood tumors, a single global, unified approach is ultimately required to determine the best strategy for managing pediatric PCNSL.\(^8\)

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REFERENCES


