

Primary central nervous system lymphoma (PCNSL)

Primary [central nervous system lymphoma](#) (PCNSL) is a type of [central nervous system](#) restricted [non-Hodgkin lymphoma](#), whose histopathological diagnosis is majorly large [B cell lymphoma](#).

They are extranodal, of the diffuse large [B cell](#) type that is confined to the [brain](#), [eyes](#), [leptomeninges](#), or [spinal cord](#), in the absence of systemic lymphoma.

Classification

[Primary Central Nervous System Lymphoma Classification](#).

Epidemiology

Primary [cerebral lymphoma](#) represents 4%–7% of primary brain tumors, and its incidence has increased in the last 3 decades ¹⁾.

see [Primary central nervous system lymphoma epidemiology](#).

see [Primary central nervous system lymphoma in AIDS](#)

History

Bailey first described PCNSL as “perithelial sarcoma” of the CNS and Henry in 1974 recognized its lymphoid origin ²⁾.

Generally, PCNSL of DLBCL type is a highly aggressive tumor and although its prognosis is poor, about one-third of younger patients can hope for cure of the disease ³⁾.

Variants

see [Lymphomatosis cerebri](#).

It typically presents as multifocal T2 hyperintense lesions that both enhance after gadolinium administration and demonstrate mass effect. A case study in 1999 coined the term ‘lymphomatosis cerebri’ to describe an exceedingly rare variant of PCNSL characterised by diffuse parenchymal infiltration of lymphomatous cells.

Clinical features

Although exceedingly rare, primary central nervous system lymphoma can present as growing intracerebral hemorrhage due to repeated intratumoral hemorrhages. High expression of vascular endothelial growth factor and the mass effects of hemorrhage could be associated with the onset and growth of intracerebral hemorrhage. Early evaluation and meticulous observation are important to avoid progressive, life-threatening situations in such cases ⁴⁾.

Diagnosis

see [Primary central nervous system lymphoma diagnosis](#).

Differential diagnosis

[Primary central nervous system lymphoma differential diagnosis](#).

Treatment

see [Primary central nervous system lymphoma treatment](#).

Outcome

[Primary central nervous system lymphoma outcome](#).

Case series

see [Primary central nervous system lymphoma case series](#).

Randomised trials

Only three completed randomised trials are available for primary CNS lymphoma: one phase 3 and two phase 2 trials ^{5) 6) 7)}.

Case reports

Primary central nervous system lymphoma case reports.

1)

Surawicz TS, McCarthy BJ, Kupelian V, et al. Descriptive epidemiology of primary brain and CNS tumors: results from the Central Brain Tumor Registry of the United States, 1990–1994. *Neuro-oncol* 1999;1:14–25.

2)

Ney DE, Deangelis LM. *Non- Hodgkin Lymphomas*. Philadelphia: Lippincott Williams & Wilkins; 2010. Management of central nervous system lymphoma; pp. 527–539.

3)

Juergens A, Pels H, Rogowski S, Fliesbach K, Glasmacher A, Engert A, Reiser M. et al. Long-term survival with favorable cognitive outcome after chemotherapy in primary central nervous system lymphoma. *Ann Neurol*. 2010;67(2):182–189. doi: 10.1002/ana.21824.

4)

Matsumoto Y, Kashimura H, Aso K, Saura H, Osakabe M, Kurose A. Primary Central Nervous System Lymphoma Presenting as Growing Intracerebral Hemorrhage. *World Neurosurg*. 2018 Aug;116:155–158. doi: 10.1016/j.wneu.2018.05.107. Epub 2018 May 23. PMID: 29803066.

5)

Ferreri AJ, Reni M, Foppoli M, et al. High-dose cytarabine plus high-dose methotrexate versus high-dose methotrexate alone in patients with primary CNS lymphoma: a randomised phase 2 trial. *Lancet* 2009; 374: 1512–20.

6)

Thiel E, Korfel A, Martus P, et al. High-dose methotrexate with or without whole brain radiotherapy for primary CNS lymphoma (G-PCNSL-SG-1): a phase 3, randomised, non-inferiority trial. *Lancet Oncol* 2010; 11: 1036–47.

7)

Omuro AM, Chinot O, Taillandier L, et al. Methotrexate and temozolomide versus methotrexate, procarbazine, vincristine, and cytarabine for primary CNS lymphoma in the elderly population: an intergroup ANOCEF-GOELAMS randomised phase 2 trial. *Lancet Haematol* 2015; 2: e251–59.

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