

Primary central nervous system lymphoma epidemiology

Primary central nervous system lymphoma (PCNSL) is a relatively common central nervous system tumor.

It is rare in children with immunocompromise as an important risk factor.

The majority of Primary central nervous system lymphomas (PCNSL) (95%) are considered diffuse large B cell lymphomas (DLBCLs).

Because of the low incidence of primary central nervous system lymphoma (PCNSL) in non-HIV individuals and because of the lack of specific clinical manifestations and auxiliary examinations, the disease is easily missed or misdiagnosed.

Primary CNS lymphomas are estimated to account for up to 1% of all lymphomas, 4–6% of all extranodal lymphomas, and about 3% of all CNS tumors ^{1) 2) 3) 4) 5) 6)}.

The incidence of primary central nervous system lymphoma (PCNSL) has increased in the last two decades and the clinical research regarding the treatment for PCNSL patients has also increased. After a continuous rise in the incidence of primary CNS lymphoma during the 1980s and 1990s, epidemiological data in high-income countries show a decrease in incidence, particularly among young patients with AIDS ⁷⁾.

By contrast, the incidence of primary CNS lymphoma continues to rise in elderly patients, who represent most patients in immunocompetent populations ^{8) 9) 10)}.

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