Journal of Neuroinfectious Diseases
ISSN: 2314-7326

Stoma, J Neuroinfect Dis 2024, 15:2

Review Article Open Access

Primary Lymphoma of the Central Nervous System as a Neurological Issue

Bożena Stoma*, Trojanowski Szczepanek, Ewa Sokołowska

Department of Neurosurgery and Paediatric Neurosurgery, Medical University in Lublin, Poland

Abstract

Primary central nervous system lymphoma (PCNSL) is a rare form of extranodal non-Hodgkin lymphoma that arises within the brain, spinal cord, leptomeninges, or eyes without evidence of systemic involvement at the time of diagnosis. Its rarity and unique presentation make PCNSL a challenging diagnostic and therapeutic dilemma in the field of neurology. This article aims to review the current understanding of PCNSL, including its epidemiology, pathogenesis, clinical presentation, diagnostic modalities, treatment options, and prognosis. Additionally, we discuss emerging research directions and potential novel therapeutic approaches to improve outcomes for patients with this neurological issue.

Keywords: Primary central nervous system lymphoma, PCNSL, neurology, diagnosis, treatment, prognosis

Introduction

Primary central nervous system lymphoma (PCNSL) is an uncommon subtype of non-Hodgkin lymphoma (NHL) characterized by malignant lymphoid proliferation within the brain, spinal cord, leptomeninges, or eyes without systemic involvement at presentation. Although PCNSL accounts for less than 3% of all primary brain tumors, its incidence has been rising, particularly among immunocompromised individuals, such as those with human immunodeficiency virus (HIV) infection or organ transplant recipients. The unique clinical and radiological features of PCNSL pose diagnostic challenges for neurologists, oncologists, and other healthcare providers involved in the care of affected patients. In this article, we provide a comprehensive review of PCNSL as a neurological issue, focusing on its epidemiology, pathogenesis, clinical manifestations, diagnostic approaches, treatment strategies, and prognosis [1,2].

Epidemiology: PCNSL comprises approximately 4% of all primary central nervous system (CNS) tumors and represents 4-6% of all extranodal NHL cases. The annual incidence of PCNSL has been estimated at 0.47 per 100,000 person-years, with a slight male predominance. Although PCNSL can occur at any age, it is more common in older adults, with a median age at diagnosis ranging from 55 to 65 years. Notably, PCNSL has a higher incidence among immunocompromised individuals, including those with HIV/AIDS, organ transplant recipients, and patients receiving immunosuppressive therapy. The introduction of highly active antiretroviral therapy (HAART) has led to a decline in the incidence of PCNSL among HIV-infected individuals; however, it remains a significant cause of morbidity and mortality in this population [3].

Pathogenesis: The exact etiology of PCNSL remains incompletely understood; however, several factors have been implicated in its pathogenesis. Immunosuppression, either due to HIV infection, organ transplantation, or immunosuppressive therapy, is a well-established risk factor for the development of PCNSL. Additionally, there is evidence to suggest a role for oncogenic viruses, such as Epstein-Barr virus (EBV), in the pathogenesis of PCNSL, particularly in immunocompromised individuals. Genetic alterations, including gains of chromosome 9 and losses of chromosome 6q and 6p, have also been identified in PCNSL tumors, although their precise contribution to tumorigenesis remains to be elucidated [4].

Clinical Presentation: The clinical presentation of PCNSL can vary widely depending on the location and extent of CNS involvement. Common symptoms include headache, cognitive impairment, focal neurological deficits, seizures, and visual disturbances [5]. In immunocompromised patients, PCNSL may present with atypical features, such as multifocal lesions, subacute onset, or rapid neurological deterioration. Ocular involvement, manifesting as vitreous opacities, uveitis, or retinal lesions, occurs in approximately 20-25% of PCNSL cases and may precede the diagnosis of CNS involvement.

Diagnostic Evaluation: The diagnosis of PCNSL requires a multimodal approach, including neuroimaging, cerebrospinal fluid (CSF) analysis, and histopathological examination of brain tissue. Magnetic resonance imaging (MRI) is the preferred imaging modality for evaluating suspected PCNSL, demonstrating characteristic findings of enhancing, T2-hyperintense lesions that are often periventricular or perivascular in distribution. Contrast-enhanced MRI can also aid in the differentiation of PCNSL from other CNS lesions, such as glioblastoma multiforme or metastatic disease. CSF analysis may reveal elevated protein levels, lymphocytic pleocytosis, and the presence of malignant lymphocytes on cytological examination [6]. However, definitive diagnosis requires histopathological confirmation, typically obtained via stereotactic brain biopsy or surgical resection.

Treatment Strategies: The optimal management of PCNSL remains controversial due to its rarity and heterogeneous clinical presentation. Historically, high-dose methotrexate-based chemotherapy regimens, often in combination with whole-brain radiation therapy (WBRT), have been the cornerstone of treatment for PCNSL. However, concerns regarding neurotoxicity and long-term cognitive impairment associated with WBRT have led to a shift towards

***Corresponding author:** Bożena Stoma, Department of Neurosurgery and Paediatric Neurosurgery, Medical University in Lublin, Poland, E-mail: bozenastoma@umlub.pl

Received: 01-Mar-2024, Manuscript No: JNID-24-130968; **Editor assigned:** 04-Mar-2024, Pre-QC No: JNID-24-130968 (PQ); **Reviewed:** 18-Mar-2024, QC No: JNID-24-130968; **Revised:** 25-Mar-2024, Manuscript No: JNID-24-130968 (R); **Published:** 29-Mar-2024, DOI: 10.4172/2314-7326.1000498

Citation: Stoma B (2024) Primary Lymphoma of the Central Nervous System as a Neurological Issue. J Neuroinfect Dis 15: 498.

chemotherapy-based approaches, including high-dose chemotherapy with autologous stem cell transplantation (ASCT) or novel targeted agents, such as monoclonal antibodies against CD20 (rituximab) or immunomodulatory drugs (lenalidomide). The use of intrathecal chemotherapy or CNS-penetrant small molecule inhibitors, such as ibrutinib or temozolomide, may also play a role in the management of PCNSL, particularly in patients with refractory or relapsed disease.

Prognosis: The prognosis of PCNSL varies depending on several factors, including age, performance status, extent of disease, and response to treatment. Overall survival rates have improved in recent years with the introduction of high-dose methotrexate-based chemotherapy regimens and consolidation therapy with ASCT; however, long-term survival remains poor, particularly in elderly patients or those with refractory or relapsed disease. The presence of certain prognostic factors, such as elevated lactate dehydrogenase (LDH) levels, poor performance status, or involvement of deep brain structures, is associated with worse outcomes [7]. Early diagnosis and prompt initiation of treatment are critical for optimizing outcomes in patients with PCNSL.

Future Directions: Advances in molecular profiling and targeted therapies offer promising avenues for improving the management of PCNSL and overcoming treatment resistance. Targeted agents directed against specific molecular pathways implicated in the pathogenesis of PCNSL, such as the PI3K/AKT/mTOR pathway or B-cell receptor signaling cascade, are currently under investigation in clinical trials. Additionally, the development of novel imaging techniques, such as positron emission tomography (PET) and advanced MRI sequences may enhance the early detection and monitoring of PCNSL lesions. Collaborative efforts among neurologists, oncologists, and basic scientists are essential for advancing our understanding of PCNSL and developing more effective therapeutic strategies for this challenging neurological issue [8-10].

Conclusion

Primary central nervous system lymphoma represents a unique neurological issue characterized by malignant lymphoid proliferation within the brain, spinal cord, leptomeninges, or eyes. In conclusion, primary lymphoma of the central nervous system represents a complex neurological issue that requires a comprehensive understanding of its clinical manifestations, diagnostic approaches, and therapeutic interventions. By advancing our knowledge and collaborative efforts in this field, we can strive to enhance the management and prognosis of PCNSL, ultimately improving the lives of patients affected by this challenging condition.

Acknowledgments

None

Conflict of Interest

None

References

1. Silver MH, Newell K, Brady C, Hedley-White ET, Perls TT (2002) Distinguishing between neurodegenerative disease and disease-free aging: correlating neuropsychological evaluations and neuropathological studies in centenarians. Psychosom Med 64: 493–501.
2. Stek ML, Gussekloo J, Beekman ATF, Van Tilburg W, Westendorp RGJ (2004) Prevalence, correlates and recognition of depression in the oldest old: the Leiden 85-plus study. J Affect Disord 78: 193–200.
3. von Heideken Wågert P, Rönnmark B, Rosendahl E, Lundin-Olsson L, M C Gustavsson J, et al. (2005) Morale in the oldest old: the Umeå 85+ study. Age Ageing 34: 249–255.
4. Miles TP, Bernard MA (1992) Morbidity, disability, and health status of black American elderly: a new look at the oldest-old. J Am Geriatr Soc 40: 1047–1054.
5. Gueresi P, Troiano L, Minicuci N, Bonafé M, Pini G, et al. (2003) The MALVA (MAntova LongeVA) study: an investigation on people 98 years of age and over in a province of Northern Italy. Exp Gerontol 38: 1189–1197.
6. Nybo H, Petersen HC, Gaist D, Jeune B, Andersen K, et al. (2003) Predictors of mortality in 2,249 nonagenarians—the Danish 1905-Cohort Survey. J Am Geriatr Soc 51: 1365–1373.
7. Von Strauss E, Fratiglioni L, Viitanen M, Forsell Y, Winblad B (2000) Morbidity and comorbidity in relation to functional status: a community-based study of the oldest old (90+ years). J Am Geriatr Soc 48: 1462–1469.
8. Andersen HR, Jeune B, Nybo H, Nielsen JB, Andersen-Ranberg K, et al. (1998) Low activity of superoxide dismutase and high activity of glutathione reductase in erythrocytes from centenarians. Age Ageing 27: 643–648.
9. Ankri J, Poupard M (2003) Prevalence and incidence of dementia among the very old. Review of the literature. Rev Epidemiol Sante Publique 51: 349–360.
10. Wilkinson TJ, Sainsbury R (1998) The association between mortality, morbidity and age in New Zealand's oldest old. Int J Aging Hum Dev 46: 333–343.m