

Case Report

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A Case Report of Primary Central Nervous T-Cell Lymphoma in an Immunologically Competent Patient

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1. Introduction

Primary central nervous system (CNS) lymphomas are relatively rare, with the most common subtype being diffuse large B-cell lymphoma. Primary CNS T-cell lymphomas (PCNSTL) represent less than 5% of CNS lymphomas [1-2].

2. Case Presentation

We present the case of a 19-year-old young man with no previous history, who was admitted to our facility in August 2020 for headaches, post-prandial vomiting associated with photophobia, phonophobia and bilateral decrease in visual acuity. The initial brain Magnetic resonance imaging revealed a large post-rolandic tumoral process measuring 48mm with subfalcine involvement. The exhaustive inflammatory and infectious screening was negative, including the cervico-thoraco-abdomino-pelvic (CTAP) scan and PET-scan. On ophthalmological examination, bilateral papillary atrophy was found. Given the severity of his symptoms, a brain

biopsy was performed, which revealed a T-lymphoproliferation (CD3+, CD4+, CD7+, CD8+, CD30- and anti-TDT-). The diagnosis of a primary, unspecified central nervous system T-lymphoma, CD30 negative with a cytotoxic phenotype was made. A bone marrow biopsy showed a hypocellular bone marrow without abnormal infiltration. A treatment with methotrexate, procarbazine, and vincristine was initiated. However, the patient's condition gradually deteriorated and he died two months after the start of treatment following septic shock.

3. Discussion

The diagnosis of primary CNS T-cell lymphoma (PCNSTL) is particularly difficult, and this condition is highly likely to be underdiagnosed due to the heterogeneous Magnetic resonance imaging findings of the lesions and the lack of specificity of the initial anatomic pathology analysis.

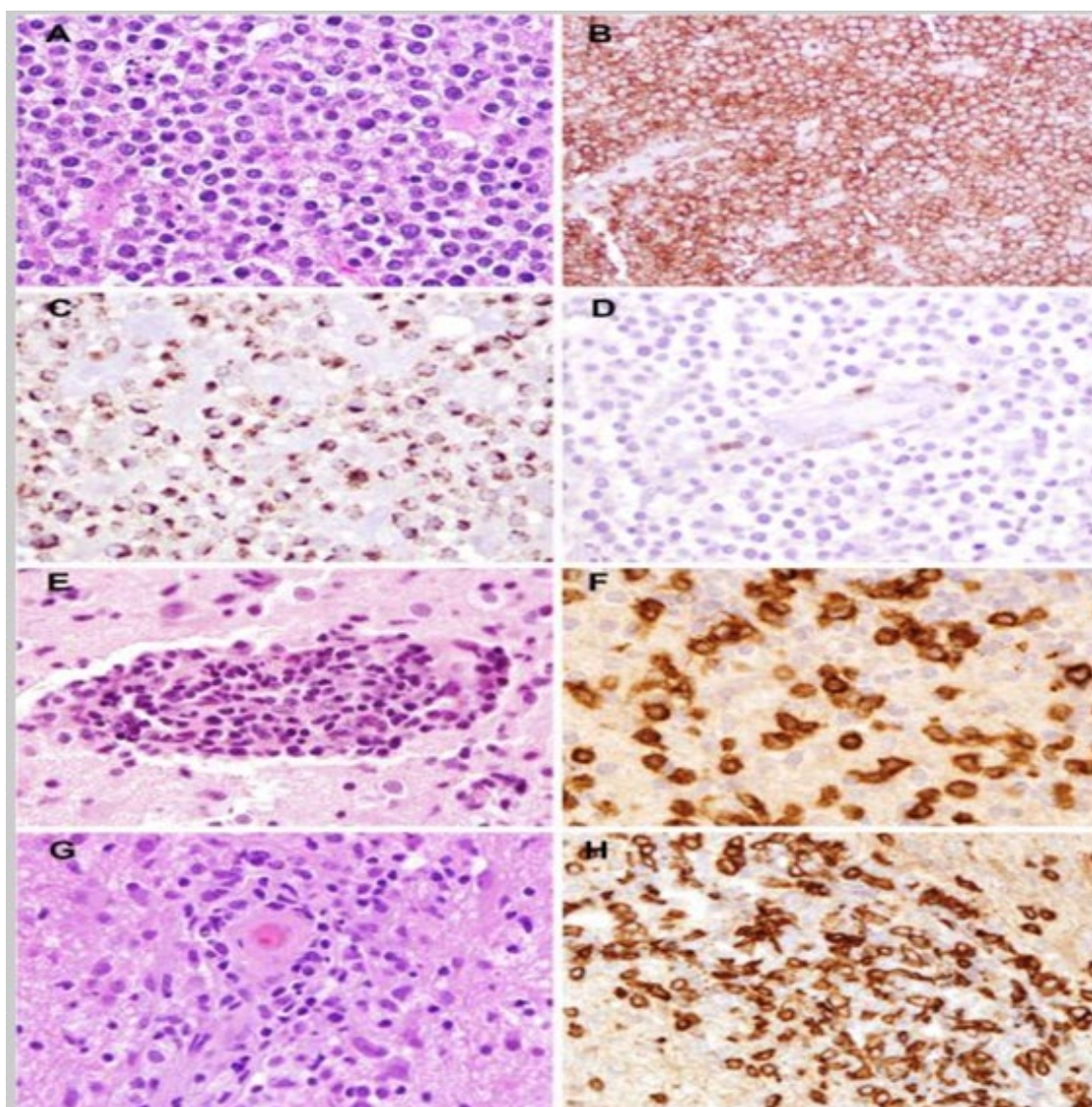


Figure: Phenotypic Aberrancies in Primary Central Nervous System T-cell Lymphoma (PCNSTL)

A) Monomorphic medium sized atypical lymphocytes with irregular nuclear contours, vesicular chromatin and occasional nucleoli. The neoplastic T-cells are CD8-positive (B), TIA-1-positive (C) and beta-F1 (D) negative. TCR gamma was also negative (not shown). (E-H) TCR gamma positive cases. The cells are mostly small-medium with irregular nuclear contours, admixed occasional larger cells and demonstrate prominent perivascular cuffing (E and G). The cells are strongly positive for TCR-gamma immunostain (F and H).

The characteristic symptoms of primary central nervous system lymphoma (PCNSL) are characterized by a slow progression and can include increased intracranial pressure, seizures, mental changes, and focal neurological deficits. The manifestation of these neurological symptoms is dependent upon the location of the lesions.

Primary central nervous system T-cell lymphoma (PCNSTL) can present in three distinct forms: peripheral T-cell lymphoma (PTCL), anaplastic large cell lymphoma (ALCL) ALK-negative, and ALCL ALK-positive. These subtypes exhibit similar clinical features, MRI findings, and prognoses as primary central nervous

system lymphoma (PCNSL) [3]. Despite the rarity of PCNSTL, treatment options remain unclear due to the limited sample size. However, a study by Nigo et al, showed that a combination of cranial radiotherapy and high-dose methotrexate (MTX) treatment, along with intrathecal methotrexate administration, increased the median disease-free and overall survival from 12-18 months to 30-40 months [4]. The International Primary CNS Lymphoma Collaborative Group conducted a study on 45 PCNSTL cases and found that the median survival time was 25 months, with the length of survival positively correlating with physical strength and the use of MTX [2].

The prognosis is poor, with an overall 5-year survival rate of 17% and a median progression-free survival of 22 months [2]. Several factors have been associated with a worse prognosis, such as advanced age, a deep tumoral process (cerebellum, brainstem, corpus callosum), high serum lactate dehydrogenase and high protein levels in the CSF [5,6].

4. Conclusion

Primary CNS T-cell lymphoma is a very rare condition, particularly in young adults. It is a rapidly progressing and poorly prognostic disease that requires prompt and early management [7].

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