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# ALK-Positive Primary Central Nervous System Anaplastic Large T-Cell Lymphoma: A Unique Case Presentation

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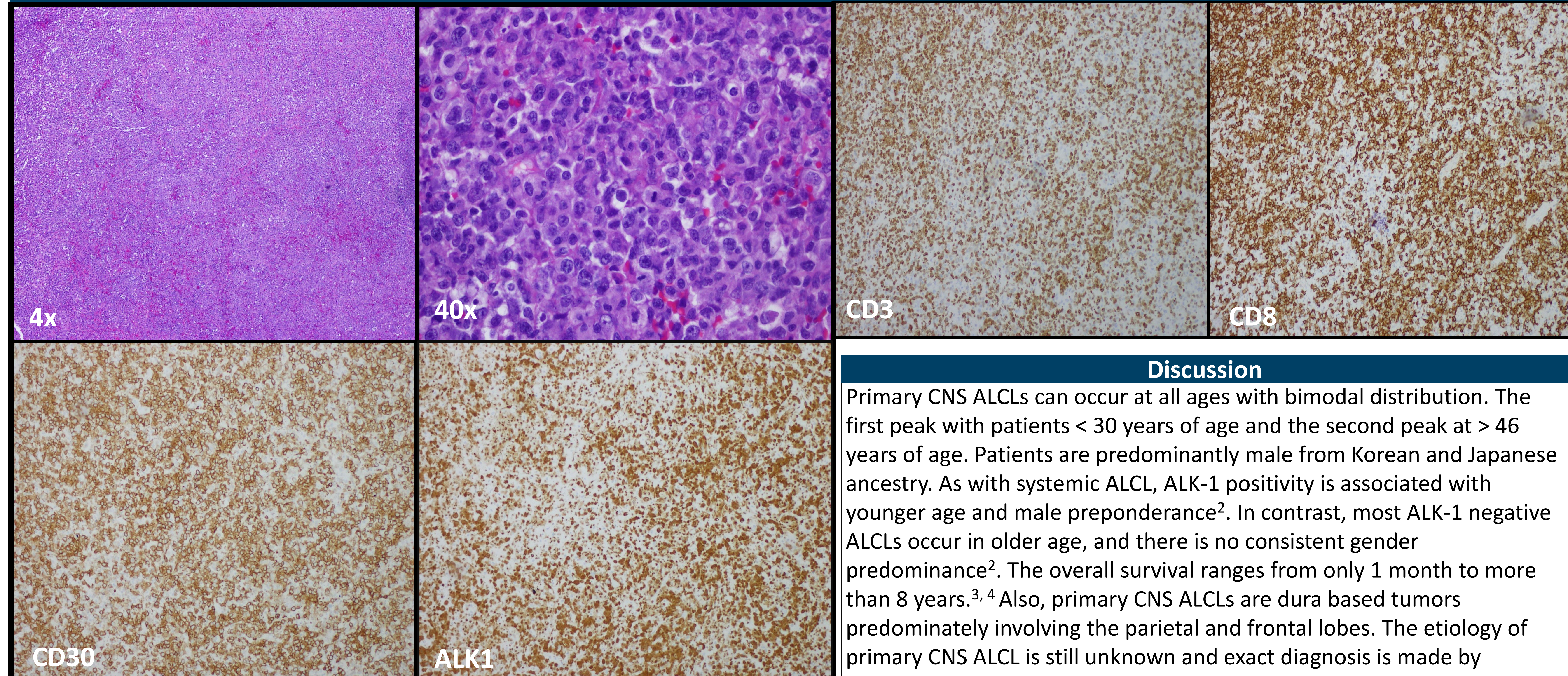
**Introduction**

Primary central nervous system lymphoma (PCNSL) is known to be primarily of B-cell lineage, with <5% of cases being of T-cell lineage. In recent years there has been a rise in PCNSL because of HIV/AIDS. Interestingly, for unknown reasons the incidence of PCNSL also appears to be increasing in immunocompetent individuals. Anaplastic large cell lymphoma (ALCL) is an uncommon type of peripheral T-cell lymphoma characterized by painless lymphadenopathy and usually presenting in immunocompetent young adults and children with female preponderance. ALCL is broadly sub-classified into anaplastic lymphoma kinase (ALK)-positive and ALK-negative subtypes. ALK-positive tumors tend to have a better response to therapy and better overall survival. CNS involvement by ALCL T-cell lymphoma is extremely rare<sup>1</sup>. Approximately 30 cases are reported in the literature, mostly in male patients of Korean and Japanese ancestry and involving the parietal and frontal lobes. Because of the rarity of this lymphoma, the pathogenesis, prognostic factors and treatment strategies have not been well studied. We report a unique case of ALK+ ALCL in an Asian female and predominantly involving the occipital lobe.

**Patient**

The patient is an immunocompetent 18 year old Asian female with a recent history of viral meningitis with right VI nerve palsy which resolved on its own. The patient presented with a 3 week history of non-progressive left sided vision changes and complaints of left sided dull headache not associated with the vision changes and unaccompanied by neck stiffness or fevers.

**Histology and Immunohistochemical stains**



MRI was performed and showed marked thickening and heterogeneous enhancement of the medial left occipital gyri with associated mass effect and vasogenic edema. The patient underwent stereotactic biopsy of the mass.

**Histology**

Histologic sections of the mass showed neural tissue with perivascular cuffing and a diffuse parenchymal and leptomeningeal infiltrate of discohesive, pleomorphic predominantly large cells with abundant cytoplasm, round to irregular nuclei, dispersed chromatin and prominent nucleoli. Vascular thrombosis and multifocal areas of necrosis and intratumoral hemorrhage were also seen. There was prominent reactive astrocytosis in the parenchyma adjacent to infiltrated areas.

**Immunohistochemistry and Fluorescent In-Situ Hybridization Studies**

The neoplastic cells were positive for CD3, CD7, CD8, ALK1, CD25, EMA, Granzyme B and CD30 on stains; and negative for CD20, CD19, CD56, CD57, TDT, CD1a, EBV (EBER), and CD15. There was equivocal staining for CD4 by some of the neoplastic cells. CD68 stained the background histiocytes. Ki67 proliferation index was 60%-80%. GFAP highlighted the prominent reactive astrocytosis. Additionally, interphase FISH hybridization studies were positive for ALK associated translocation t(2; 5).

**Discussion**

Primary CNS ALCLs can occur at all ages with bimodal distribution. The first peak with patients < 30 years of age and the second peak at > 46 years of age. Patients are predominantly male from Korean and Japanese ancestry. As with systemic ALCL, ALK-1 positivity is associated with younger age and male preponderance<sup>2</sup>. In contrast, most ALK-1 negative ALCLs occur in older age, and there is no consistent gender predominance<sup>2</sup>. The overall survival ranges from only 1 month to more than 8 years.<sup>3,4</sup> Also, primary CNS ALCLs are dura based tumors predominately involving the parietal and frontal lobes. The etiology of primary CNS ALCL is still unknown and exact diagnosis is made by histopathological examination and by immunohistochemistry. In addition to necrosis, gliosis and histiocytic infiltration; helpful features in diagnosing these lesions are prominent perivascular infiltration and perivascular cuffing which are common among primary CNS B-cell and T-cell lymphomas. Since, Primary CNS ALCL is very rare, management of this localized lymphoma is not well established. Currently the treatment regimen is similar to B-Cell PCNSL with high dose methotrexate chemotherapy and/or whole brain radiotherapy. The case presented here is unique, as this is the first case report which describes ALK+ ALCL in an Asian female with involvement of the occipital lobe and leptomeninges. We believe this case will be a unique addition to the cases previously reported in the literature of this extremely rare lymphoma.

**References**

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