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Case Report

A Rare Case of Isolated Intraventricular Primary Central Nervous System Lymphoma in an 85-Year-Old Man

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Abstract

Primary CNS lymphoma (PCNSL) is rare malignant B cell lymphoid tumor of brain which predominantly occurs in supratentorial region in periventricular location. Majority of PCNSL are of DLBCL type and idiopathic in etiology. Here we are reporting a case of primary CNS lymphoma, DLBCL involving extremely uncommon intraventricular location. Central neurocytoma, subependymal giant cell astrocytoma, choroid plexus tumors and meningiomas are the common diagnosis at this site. Aim of reporting this case is to bring awareness of unusual intraventricular location of primary CNS lymphoma which should be kept in mind before considering gross total excision of lesion.

Keywords: *Primary CNS lymphoma, Intraventricular lymphoma, DLBCL*

Introduction

Primary central nervous system lymphomas (PCNSL) constitute 2.4%–3% of all brain tumors and 4%–6% of all extranodal lymphomas.[1] Majority of them are supratentorial in location (60% of cases) and involve the frontal lobe, thalamus, basal ganglia, and periventricular brain parenchyma.[2] Isolated intraventricular primary CNS lymphoma is an extremely rare and only few cases have been reported in the English literature so far. We present a case of diffuse large B cell lymphoma (DLBCL) involving the lateral and third ventricles.

Case Report

An 85-year-old male presented with gradually progressive bilateral vision loss for 1 year. He also had altered sensorium, memory deficits, and decreased oral intake for 2 months. Magnetic resonance imaging (MRI) of the brain showed multiple solid lobulated enhancing masses of variable size involving the lateral and third ventricles and extending into the periventricular white matter; these lesions appeared mildly hypointense on T2-weighted images and did not show any diffusion restriction of diffusion-weighted images. No other parenchymal lesion was observed. Lymphoma was considered as one of the differential diagnosis. Positron emission tomography-computed tomography did not show any extra-extracranial fluorodeoxyglucose avid lesion [Figure 1].

Cerebrospinal fluid (CSF) was submitted to look for any atypical cells. The initial CSF sample collected from lumbar puncture was negative, however subsequent specimen from the intraventricular sample showed many immature lymphoid cells and was reported as positive for atypical lymphoid cells [Figure 2a]. The patient also had deranged thyroid functions, coagulation parameters, and liver function tests. The patient was planned for biopsy from the lateral ventricle for confirmation of diagnosis. The patient underwent left frontal craniotomy and biopsy was done. Paraffin-embedded sections of the specimens showed focally ependyma lined brain parenchyma diffusely infiltrated by sheets of intermediate to large size atypical lymphoid cells with brisk mitosis and numerous apoptotic bodies [Figure 2d]. Tumor cells were present with angiocentric accentuation. [Figure 2b, 2c] On immunohistochemistry, these atypical lymphoid cells were positive

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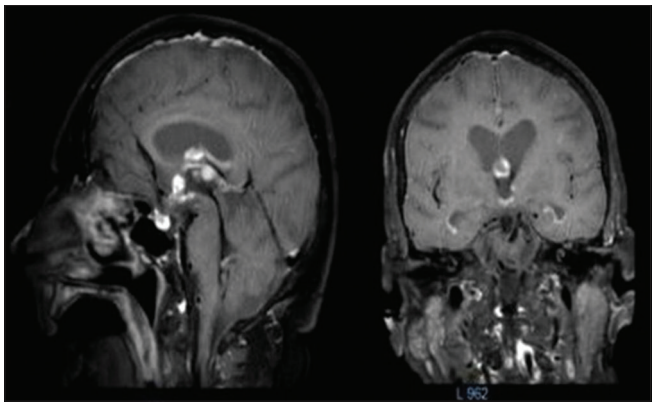

Figure 1: Magnetic resonance imaging of the brain showed multiple solid lobulated enhancing masses of variable size involving the lateral and third ventricles and extending into the periventricular white matter; these lesions appeared mildly hypointense on T2-weighted images and did not show any diffusion restriction of diffusion-weighted images. No other parenchymal lesion was observed. Positron emission tomography-computed tomography did not show any extra-extracranial fluorodeoxyglucose avid lesion

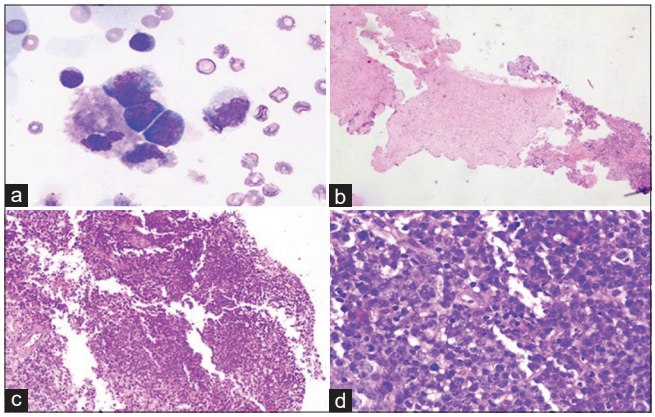

Figure 2: (a) Cytology of cerebrospinal fluid from ventricle shows atypical lymphoid cells. (b) Ependymal lined brain parenchyma infiltration by lymphoma cells. (c) Angiocentric pattern of distribution of lymphoid cells. (d) Intermediate to large size cells with brisk mitosis and apoptotic bodies

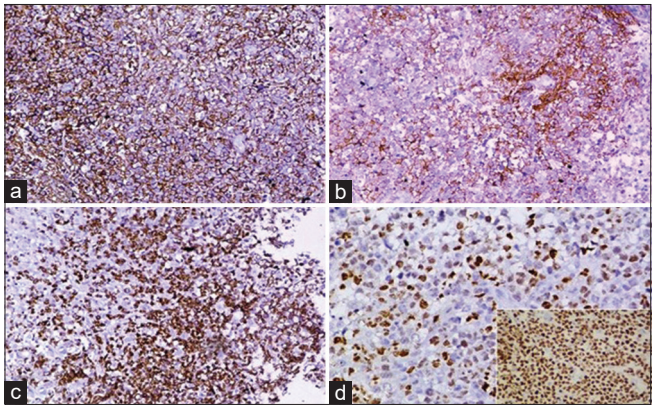

Figure 3: (a) Tumor cells are diffusely positive for CD 20 (×200) (b) tumor cells are positive for CD10 (c) tumor cells are positive for B cell lymphoma 2 (d) tumor cells are positive for B cell lymphoma 6 (inset showing proliferation index of 90%)

for CD 20, BCL 6, CD 10, and BCL-2. CD 3 highlighted few scattered background reactive population of T cells. [Figure 3] Based on classic histomorphological features and immunohistochemistry, a diagnosis of DLBCL germinal center type was rendered. In view of old age, the patient was offered combination of steroid and rituximab-based chemotherapy, followed by assessment for the administration of methotrexate and whole-brain radiotherapy. However, the patient was a foreign citizen and preferred to take treatment from his own country and left against medical advice.

Discussion

Primary CNS lymphomas constitute 2.4%–3% of all brain tumors and 4%–6% of all extranodal lymphomas.[2] Etiological factors in immunocompetent patients are unknown. Viruses like EBV, HHV6, HHV 8 do not play a role. But the etiology in immunocompetent individuals is unknown.[3] Stereotactic biopsy is the gold standard technique for confirmation of diagnosis and classification of lymphoma. But it must be done before administration of corticosteroids as they induce rapid destruction of tumor making diagnosis difficult. Most of the cases are supratentorial in location with frontal lobe and periventricular brain parenchyma being the most common sites. Posterior fossa and spinal cord are less frequently affected.[4] Pleocytosis is seen in 35%–60% of PCNSL cases and correlates well with meningeal dissemination.[5] Meningeal involvement can resemble meningitis or meningioma. On MRI, the lesions of lymphoma in CNS are hypointense on T1-weighted images and iso to hyperintense on T2-weighted images. These enhance densely on postcontrast imaging.[6] Generally, the lesions are sharp and well demarcated from the surrounding brain parenchyma with minimal associated peritumoral edema.[7] Diffuse borders and without even forming a distinct mass can be seen in some instances mimicking malignant gliomas. Histopathologically, the tumor cells invade the neural parenchyma in small clusters or single cells with angiocentric accentuation. Majority of PCNSL are DLBCL, postgerminal center type. CD 10 expression is seen in only 10% of cases of PCNSL, thus CD 10 positivity should prompt a search for systemic DLBCL.[8]

Conclusion

Intraventricular location for a PCNSL is an extremely uncommon site and raises the differential diagnosis such as central neurocytomas, subependymal giant cell astrocytomas choroid plexus tumors, and meningiomas. Awareness about the uncommon sites and supportive radiological features may limit the surgery only for obtaining biopsy for confirmation, followed by chemotherapy and radiotherapy.

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Conflicts of interest

There are no conflicts of interest.

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