Primary Central Nervous System Lymphoma in an Immunocompetent Young Adult Patient: A Rare Case

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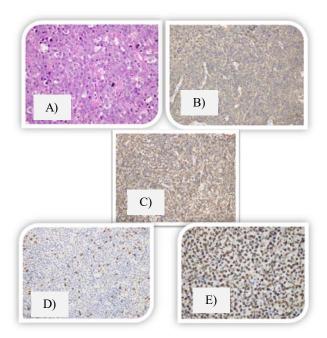


Figure 1. Pathological Anatomy. A) Hematoxylin & Eosin A PCNSL Biopsy Basal Ganglia. B). CD 20 positive. C). CD 45 positive. D). CD 3 negative. E). KI 67+90-95% high grade.

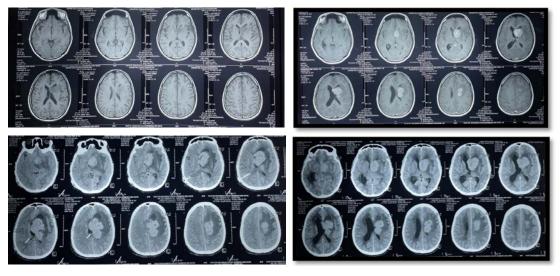


Figure 2. Brain MRI Patients Before Chemotherapy.



Figure 3. Brain MRI Patients After 2 Cycle Chemotherapy.

Primary CNS Lymphoma (PCNSL) is a rare form aggressive extra nodal non-Hodgkin Lymphoma (NHL) that comprising 1-2% of the primary brain tumors that develops in the brain, spinal cord, eye or leptomeningeal area without evidence of systemic involvement.¹ The overall incidence of PCNSL with immunocompetent patients is only 0.47/100,000 year in PCNSL. Approximately 10-20% of patients had ocular involvement and around one third had multifocal neurological disease. Overall long-term survival rate was only 20-40%, this is because the management of PCNSL is limited by ability of the drug due to cross the blood brain barrier (BBB). We present a B-cell central nervous system lymphoma in an immunocompetent patient who responded to chemotherapy.²

A 35-year-old man presented to our hospital with sudden change in mental status since 4 hours before admission. He was experiencing headache and blurred of vision within 3 months and have episode seizure. On Examination, Glasgow Coma Scale (GCS) E2M1V aphasia. The patient was found to have right hemiparesis with bilateral papilloedema. Visual acuity of both eyes was no light perception (NLP). The other physical exam was normal. Laboratory tests Hb 10.7 g/dl, LDH 446 U/L, and D-dimer 3.21 ug/ml. Rubella IgG 76.9, CMV Ig G 245.6 and, HSV IgG and IgM negative, HIV test nonreactive, Toxoplasma IgG and Toxoplasma IgM negative, HbsAg and HCV test negative. Brain MRI and MRI Stereoscopy: Lobulated mass size 7.08 cm x 4.75 cm at the left caudate nucleus and left lateral periventricular area. Choline/NAA ratio: 5-9, Choline/Creatine ratio 6-11 suggestive of malignancy, possibly lymphoma. Whole spine MRI showed bulging intervertebral disc at the level of C4-C5. Chest and abdomen CT-Scan result was normal. Bone survey result was normal, Electroencephalogram (EEG) suggested epileptiform discharges of left temporal area. Cerebrospinal Fluid: Showed gliosis and suspected malignancy. The patient underwent

ventriculoperitoneal shunt (VP shunt) due to obstructive hydrocephalus and craniotomy. Pathology biopsy and immunohistochemistry (IHC) of basal ganglia revealed a Diffuse Large B Cell Lymphoma (NHL) Non-Germinal Center, CD 20 +, Ki 67 95% (High Grade), CD 45 +, CD 3 -, BCL6 +, Mum 1+. For this patient, we started induction therapy with RMP Regimens (Rituximab 375 mg/m², day 1, 15 and 29, High Dose Methotrexate (HDMTX) 3000 mg/m² day 2, 16 and 30, and Procarbazine 60 mg/m² day 3-12) Due to the unavailability of procarbazine in town, we decided to change it with Dacarbazine 375 mg/m^2 days 3, 17 and 31), Dexamethasone 5 mg/6 hours, and low dose whole brain radiotherapy (WBRT).

PCNSL is a rare form aggressive extra nodal NHL, especially in Immunocompetent patient. In this particular case of patients High Dose Methotrexate Chemotherapy has achieved high respond especially for this patient that showed GCS E4M5V6 and recovery neurological deficit after 2 cycle chemotherapy.

REFERENCES

- 1. Portnow LJ, Baehring J. Central nervous system cancers. NCCN Clinical Practice Guidelines in Oncology. Version 2.2021.
- Rubenstein JL, Gupta NK, Mannis GN, LaMarre AK, Treseler P. How I treat CNS lymphomas. Blood. 2013;122:2318–30. doi: 10.1182/ blood-2013-06-453084