

Emergent management with favorable outcome of an unusual presentation of a primary CNS lymphoma in an immunocompetent patient

Sir,
 Primary CNS lymphomas (PCNSL) are extra-nodal, malignant tumors arising within the brain, eyes, leptomeninges and spinal cord in the absence of systemic involvement at the time of diagnosis.^[1,2] PCNSL is seen in both immunocompromised and immunocompetent patients.^[1,2] PCNSL differs from its systemic counterpart by having a poorer prognosis and

demonstrating an angio-centric growth pattern without follicles.^[2] In immunocompetent patients most of the PCNSL (90-95%) are B cell lymphomas.^[2]

PCNSL occur in decreasing order of frequency in the cerebral hemisphere (38%), basal ganglia and thalamus (16%) corpus callosum (14%), ventricular region (12%), and the cerebellum (9%), demonstrating a proclivity toward the midline peri-ventricular structures.^[1,3] The commonest presenting features are neuropsychiatric.^[1,3] Although features of raised intracranial pressure may be present in as many as 33% (mainly due to hydrocephalus), impending herniation during the presentation is extremely rare and only three such cases have been reported in the literature [Table 1].^[1-5]

A 48-year-old male patient presented to the emergency room in an altered sensorium of 1 h duration with preceding progressive hemiparesis of six months duration. The neurological examination revealed a deeply comatose patient with a Glasgow Coma score of 7/15 [E1V1M5]. The patient had a dilated right pupil with left hemiparesis of grade 2. MRI of the brain [Figure 1] showed a well-defined lobulated parasagittal lesion.

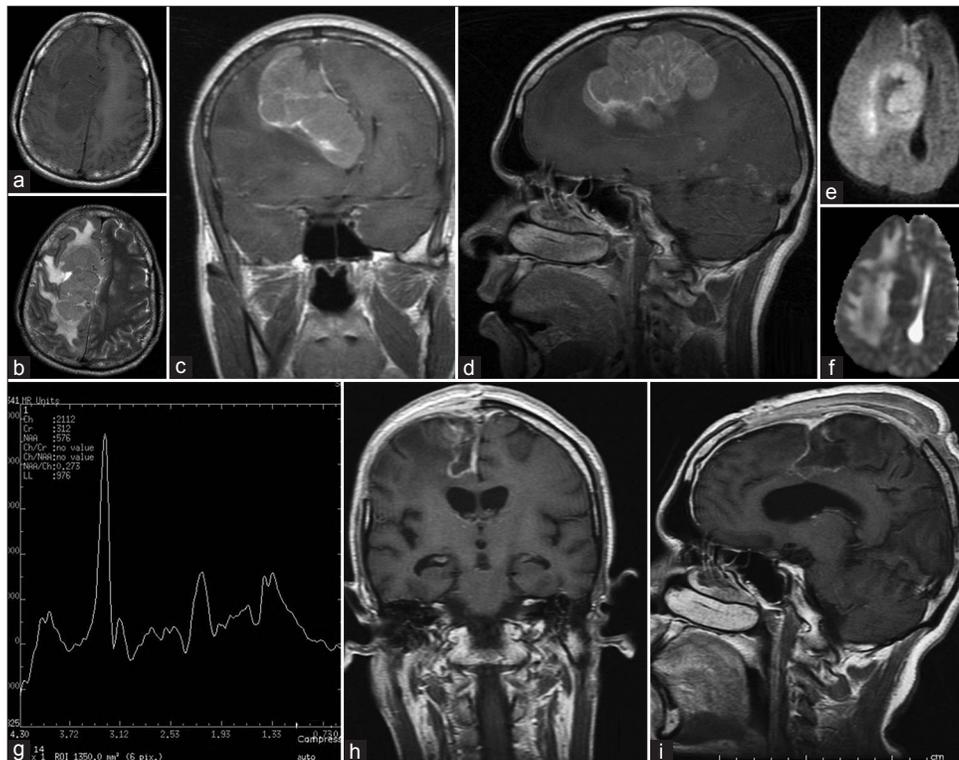


Figure 1: MRI of the brain revealing a right parasagittal lesion measuring 7.4 × 3.8 × 6.0 cm. It was hypointense on T1W images (a) Hyperintense on T2W images (b) With mass effect and midline shift. Figure 1c and d depict the contrast enhancement on T1W coronal and sagittal images. There was restriction on DWI (e) And hypointensity on ADC. (f) The MR spectroscopy revealed an elevated choline to creatinine ratio and loss of NAA on MRS (g) With surrounding peri-lesional edema with subfalcine/uncal herniation. H and i are post-operative T1W gadolinium images depicting complete excision of lesion

Table 1: Review of literature of PCNSL operated as an emergency

Year	Author	Age/sex	Presentation	Acute herniation signs	Location	Cause of acute presentation	Procedure	Follow up
2008	Kim IY <i>et al.</i> ^[5]	49/F	Deterioration in sensorium, hemiparesis, raised ICP	Present	Left frontal lobe	Hemorrhage	Decompression	10 months
2012	Ilker A, <i>et al.</i> ^[4]	33/F	Headache, drowsiness	N.A	Frontal – septum pellucidum	Increase in size and mass effect during pregnancy	Tumor decompression delayed 2 weeks*	Death
2013	Cheatle JT <i>et al.</i> ^[3]	31/F	Headache/ataxia	N.A	Septum pellucidum	Increase in size and mass effect during pregnancy^	Decompression	3 years
2013	Matsuyama J <i>et al.</i> ^[6]	64/M	Comatose	Present	Right frontal	Hemorrhage	Endoscopic evacuation	Death
2013	Present case	48/M	Headache raised ICP	Present	R posterior frontal	Acute herniation	Emergency craniotomy and decompression	3 months

*Tumor decompression delayed until fetal lung maturity was attained, ^ Biopsied earlier- inconclusive, R: Right, ICP: Intracranial pressure, N.A: Not available, PCNSL: Primary CNS lymphomas

The patient underwent emergency craniotomy and decompression of the lesion. Intra-operatively the tumor had a poor plane with the brain in some areas. Histopathology clinched the diagnosis of a B cell non-Hodgkin's lymphoma [Figure 2]. Post-operatively the patient improved in sensorium as did his motor power to grade 4. Evaluation for systemic lymphoma was negative. The patient underwent whole brain radiotherapy of 60 Gy and received high dose of methotrexate chemotherapy.

Despite advancement in the treatment modalities the prognosis of PCNSL remains poor.^[3] Symptomatic therapy results in a median survival of 2-3 months.^[1,2] Steroids decrease the tumor volume by inducing apoptosis in the cells and prolongs the median survival to 4-5 months.^[2] The standard of care has been systemic chemotherapy with or without radiotherapy.^[1-5] Surgery has been traditionally contraindicated in PCNSL due to its diffusely infiltrative nature^[1,2] and has mainly been limited to tissue diagnosis especially in the cases of atypical radiology and partial decompression has no proven benefit over a stereotactic biopsy and has sometimes been associated with a poorer prognosis.^[1,2] There has been one recent review by Weller *et al.* which challenges this traditional view favoring cytoreductive surgery at least for a subset of patients with PCNSL^[7] We have done a review of the literature highlighting the very few instances in which a PCNSL should be operated [Table 1]. In this group of patients, the presentation was due to increased size during the pregnancy in two cases and hemorrhage in two cases.^[3-6] There have been no cases reported in which the patient had impending herniation due to the size of the tumor, as in the present case.

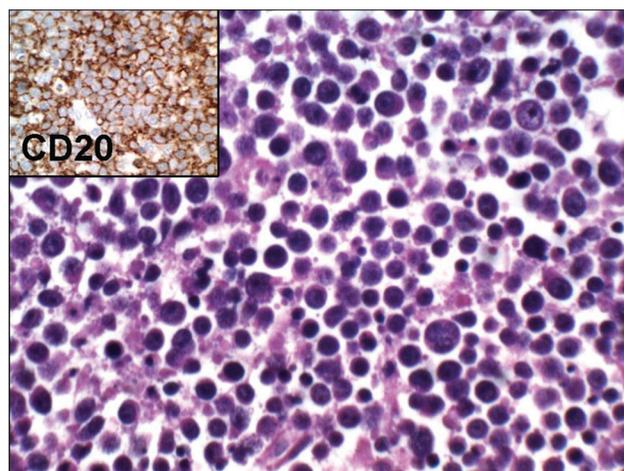


Figure 2: Paraffin section of non-Hodgkin lymphoma showing monomorphic round to oval cells with hyperchromatic nuclei, mitotic activity and (Inset) immunopositivity for CD20. (H and E x400) [Inset: Avidin Biotin Complex Immunoperoxidase x400]

Despite the criticism associated with surgery in PCNSL, it has a significant and special role in the case of rare emergency presentation and may have a favorable outcome such as in the present case.

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