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 a 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 x 3.8 x 6.0 cm. It was hypointense on TIW 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 TIW gadolinium images depicting complete excision of lesion.
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. Surgical management can be considered for patients with poor performance status and increased size of the tumor which indicates the need of a biopsy for diagnosis. In the present case the patient was not a good surgical candidate due to the high risk of herniation. Thus, treatment was initiated with high dose methotrexate and whole brain radiotherapy.

Surgery has been traditionally contraindicated in PCNSL due to its diffusely infiltrating properties 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. 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.

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

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

*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

Despite advancement in the treatment modalities the prognosis of PCNSL remains poor. Symptomatic therapy results in a median survival of 2-3 months. Steroids decrease the tumor volume by inducing apoptosis in the cells and prolongs the median survival to 4-5 months. The standard of care has been systemic chemotherapy with or without radiotherapy. Surgery has been traditionally contraindicated in PCNSL due to its diffusely infiltrative nature and has 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. 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. 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 increased size during the pregnancy in two cases and hemorrhage in two cases. 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.

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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Emergent management with favorable outcome of an unusual presentation of a primary central nervous system lymphoma in an immunocompetent patient

Sir,

This article is a report of a case with primary central nervous system (CNS) lymphoma, with impending brain herniation due to a huge mass, and surgical reduction was effective in obtaining neurological improvement.[1]

Usually the standard therapy for primary CNS malignant lymphoma, at present, is high-dose methotrexate therapy with or without radiotherapy, and surgery is mainly limited to tissue diagnosis, due to its diffusely infiltrative nature. However, in some cases, such as a huge tumor with impending herniation or CNS lymphoma cases with intratumoral hemorrhage, emergency surgery for mass reduction is required.

Intracerebral hemorrhage in primary CNS lymphoma is rare, but higher vascular endothelial growth factor (VEGF) immune reactivity in hemorrhagic cases is observed compared to non-hemorrhagic cases.[2‑5] Also, serum elevation of VEGF levels are predictors of poor prognosis. In the previously reported cases of intratumoral hemorrhage in primary CNS lymphomas, emergent craniotomy and removal of mass reduction was performed. Although impending herniation of primary CNS lymphomas is rare, a mass reduction surgery should sometimes be considered in such unique cases.

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