Letters to the Editor

Synchronous Pediatric Supratentorial Glioblastoma Multiforme with Noncontiguous Infratentorial Pilocytic Astrocytoma: A Rare Event

SIR,

Multicentric gliomas of the central nervous system are rare lesions with an estimated incidence being 2.4%–4.9% of all gliomas. Even more uncommon are the ones which arise in the supra- and infra-tentorial compartments, especially of different histopathological subtypes. We present the first pediatric case of a multicentric bi-compartmental glioma of different histopathological subtypes.

A 12-year-old girl presented with chief complaints of generalized headache with vomiting and blurring of vision at peak of headache; features suggestive of raised intracranial pressure for a total duration of 3 months. Historically, she also reported to have difficulty in maintaining balance while walking for 1-month duration. She had previously been asymptomatic and had no positive history for other medical or surgical illnesses. On neurological examination, the patient was conscious oriented, and the higher mental functions were normal. The motor and cerebellar examination revealed weakness of the right upper and lower limb with grade 4/5, gross truncal ataxia with bilateral horizontal gaze-evoked nystagmus. Fundus examination showed bilateral papilledema. The patient was positive for nystagmus. Fundus examination showed bilateral papilledema. Magnetic resonance (MR) imaging of the brain showed a diffuse lesion in the left temporal lobe with significant mass effect on ipsilateral white matter and lateral ventricle with midline shift of 1.3 cm and evidence of uncal herniation. The lesion was hypointense on T1, hyperintense on T2, enhancing brightly, and heterogeneously on contrast administration with central nonenhancing part suggesting necrosis. The size of this lesion was approximately 6.8 cm × 4.5 cm × 3.9 cm [Figure 1a-e].

GBMs are the most common intraparenchymal primary brain tumor, representing approximately 30% of all brain tumors [Table 1]. The multiple gliomas can be classified as “multifocal” if there is an apparent route of dissemination (i.e., a white matter tract). In contrast, if there is no obvious pathway for spread,
it is termed multicentric. Multifocal glioma consists of tumors separated by white matter tracts within the same hemisphere, whereas multcentric glioma consists of noncontiguous tumors in opposite hemispheres or separated by the tentorium. Multicentric gliomas involving the supratentorial and infratentorial regions are very rare.\(^2,3\)

In an autopsy series, incidence of multifocal glioma was found to be 27.8% of cases, and the ratio of multiple to multicentric gliomas was 10.6:1.37. Review of the pertinent documented literature revealed that only 12 other cases of concomitant supratentorial and infratentorial glial tumors have been documented. Historically, multifocal and multicentric GBMs have been associated with a worse prognosis than solitary GBM, with median patient survival estimates of 6-8 months after different treatment modalities.\(^2\) Tumor dissemination at the time of diagnosis can also serve as a prognostic marker. One study stratified patients into three groups based on solitary or multifocal tumors and whether there was subependymal or subarachnoid dissemination at the time of diagnosis.\(^5\)

According to certain theories, multicentricity arises from two events.\(^2\) The first stage is neoplastic transformation, in which a wide field becomes more susceptible to neoplastic growth. The second stage is tumor proliferation at two or more activated sites that can occur simultaneously in response to various stimuli including biochemical, hormonal, and viral triggers. More contemporary theories have looked at molecular associations. For example, there is a reported association between p53 mutations and multifocal GBM that correlates the pattern of p53 mutation to tumor migration and augmented growth.\(^6\)

In a study of the growth factor receptor c-Met in GBM, one group found that 42.9% of tumors that overexpressed c-Met displayed invasive and multifocal features on initial MR imaging, whereas

<table>
<thead>
<tr>
<th>Authors</th>
<th>Age (years), gender</th>
<th>Location of Tumor</th>
<th>Histopathology</th>
<th>Treatment</th>
<th>Survival (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present study</td>
<td>12, female</td>
<td>Left temporal lobe Vermis</td>
<td>GBM, Pilocytic astrocytoma</td>
<td>Near total resection for both lesions</td>
<td>Receiving adjuvant treatment</td>
</tr>
<tr>
<td>Sophia S. Shkur et al., 2013</td>
<td>72, male</td>
<td>Right posterior periventricular region Right cerebellar hemisphere</td>
<td>GBM, GBM</td>
<td>Subtotal resection and temozolomide with WBRT</td>
<td>3</td>
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<tr>
<td>Salunke et al., 2010</td>
<td>50, male</td>
<td>Right insula Cerebellar vermis</td>
<td>Grade II astrocytoma GBM GBM</td>
<td>Subtotal resection with WBRT</td>
<td>18</td>
</tr>
<tr>
<td>Kotwica and Papierz, 1992</td>
<td>53, female</td>
<td>Left temporal lobe Cerebellar vermis</td>
<td>GBM GBM</td>
<td>Gross total resection with WBRT</td>
<td>10</td>
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<tr>
<td>Salvati et al., 1991</td>
<td>47, male</td>
<td>Right frontal lobe Left cerebellar hemisphere</td>
<td>GBM Grade III astrocytoma</td>
<td>Biopsy</td>
<td>2</td>
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<tr>
<td>Salvati et al., 1991</td>
<td>42, male</td>
<td>Right temporal lobe Pons</td>
<td>GBM</td>
<td>Gross total resection with WBRT</td>
<td>Postoperative mortality</td>
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<td>Salvati et al., 1991</td>
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<td>Left frontoparietal lobe Right cerebellar hemisphere</td>
<td>GBM GBM</td>
<td>Biopsy, chemotherapy, and WBRT</td>
<td>7</td>
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<td>Kudo et al., 1990</td>
<td>74, male</td>
<td>Right occipital lobe Right cerebellar hemisphere Right thalamus</td>
<td>GBM GBM</td>
<td>Subtotal resection</td>
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<td>Bussone et al., 1979</td>
<td>49, male</td>
<td>Right CP angle Corpus callosum Cerebellar vermis</td>
<td>GBM GBM</td>
<td>None</td>
<td>4</td>
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<td>Takeda et al., 1976</td>
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<td>Right frontal brain stem</td>
<td>GBM GBM</td>
<td>Unknown</td>
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<td>GBM GBM</td>
<td>Unknown</td>
<td>3</td>
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</tbody>
</table>

GBM: Glioblastoma, CP: Cerebellopontine, WBRT: Whole-brain radiation therapy
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There are no conflicts of interest.

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