Bilateral Thalamic Glioma: Case Report and Review

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ABSTRACT

Bilateral thalamic glioma is extremely rare and the incidence cannot be adequately expressed. We present the case of a 72 years old male suffering from the rapid deterioration of cognitive function to moderately severe dementia in a short period of time. Magnetic resonance studies demonstrated a bilateral thalamic glioma with a minimal focal gadolinium uptake in the left thalamus. Biopsy was performed and pathology report was of anaplastic astrocytoma, WHO grade III. Radiotherapy was proposed but was rejected by the patient’s relatives. The patient deceased 57 days later. We performed an extensive review of the literature and by updating the previous described series we can state that to the best of our knowledge this is the 60th case described in the literature and the second eldest patient presented. Patients suffering from this disease present a poor prognosis, the longest survival described being of 3 years in patients diagnosed with grade II bilateral thalamic glioma. Adjuvant therapy in form of radiotherapy to the thalami is most commonly used but the benefits are unclear. The natural progression of WHO grade III bilateral thalamic glioma left untreated, as can be seen from our case, has an even poorer outcome.

KEYWORDS: Astrocytoma, Bilateral thalamic glioma, Biopsy

INTRODUCTION

Primary thalamic gliomas are rare and have been presented in recent reports with an incidence of 0.84–5.2% among all intracranial tumors (2,4,15,18).

Bilateral thalamic gliomas (BLTG) constitute a unique intracranial tumoral pathology typically manifested by varying degrees of personality disorder. The rate of BLTG is much lower than that of unilateral thalamic glioma and cannot be objectively expressed in any way at present (4–7,13).

Imaging in BLTG is typical and reveals large tumors with symmetrical and strictly limited involvement of bilateral thalami, without any apparent connecting tumoral tissue (2).

The prognosis of BLTG is poor and a rapid fatal evolution has been observed (14) with the longest period of survival after the diagnosis reported as 3 years in patients with grade II BLTG (2,10).

We present the case of a BLTG manifesting with fast progressive cognitive dysfunction leading to dementia in an elderly patient.

CASE REPORT

A 72-year-old right handed male, treated only for essential arterial hypertension stage I, presented 3 months prior to admittance, with gradual deterioration of the cognitive functions and intermittently aggressive behavior towards close relatives. The symptoms progressed gradually, with the patient not being able to perform basic everyday tasks and presenting periods of extreme agitation.

A cerebral computed tomography (CT) scan was performed and showed bilateral thalamus swelling and protrusion into the third ventricle without hydrocephalus or enhancement after the administration of intravenous contrast agent. A magnetic resonance imaging (MRI) was performed showing...
homogenous bilateral thalamic lesions of hypointense appearance on T1-weighted sequences with one minimal unilateral left focal uptake after administration of gadolinium and homogenous hyperintense appearance on T2 and fluid attenuated inversion recovery (FLAIR) sequences (Figure 1A-C).

The patient presented to our department with a Glasgow Coma Scale (GCS) score of 14 points (ocular 3 points, verbal 5 points, motor 6 points), Functional Assessment Staging Test (FAST) value of grade 6a – moderately severe dementia, somnolence alternating with periods of extreme agitation, presenting signs of sensory ataxia, and without motor deficit. A left thalamic stereotaxic biopsy with the target set on the focal enhancement was proposed.

Due to poor cooperation, the patient was sedated and intubated before the placement of the CRW Radionics head frame (Radionics, Plainsboro, New Jersey, USA). Contiguous 2 mm slices were performed on a 1 Tesla machine. Data was fed into the StereoPlan Software (Version 2.0, Plainsboro, New Jersey; Radionics, 2003) and a left thalamic biopsy through a left parietal burrhole was planned and performed. Using a side-cutting 2.0 mm brain biopsy needle and negative pressure, 5 samples were obtained using a target offset of 2 mm. Frozen sections were not available.

The histopathological aspect revealed highly dense cellular and vascular tissue with cells showing a gemistocytic appearance, stroma with hemorrhagic foci, and perivascular microgliosis (Figure 2A-C). The final pathological diagnosis was anaplastic astrocytoma, WHO grade III.

An oncology consultation was requested. Radiotherapy to the bilateral thalami was proposed but was rejected by relatives requesting only palliative care. The patient was discharged after 7 days in the same clinical condition as that on admission and died 57 days later.

**Figure 1:** MRI imaging. **A)** Axial T2-weighted MR shows hyperintensity and bilateral symmetrical diffuse enlargement of thalami. **B)** Axial FLAIR sequence shows bilateral hyperintensity. **C)** T1-weighted gadolinium–enhanced image with the arrow showing the minimal focal uptake in the left thalamus.

**Figure 2:** Histopathological aspects: **A)** Densely cellular and vascular nervous tissue with numerous microglial and gemistocytic elements. Vascular structures present a hyperplastic endothelium (HE stain, 20x); **B)** Immunohistochemical staining for neural filaments reveals a dense structure of the stroma formed by NF-positive neural prolongations (NF stain, 40x); **C)** Immunohistochemical staining reveals the prevalence of GFAP (glial fibrillary acidic protein) positive cells, with fine prolongations densely arranged in between. Besides the irregular aspect of the glial elements, other dysplastic forms can be seen such as voluminous, vacuolar and nucleated nuclei and vascular hyperplasia (GFAP stain, 40x).
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Table I: Case Reports of Patients Aged over 65 Years Diagnosed with BLT

<table>
<thead>
<tr>
<th>Author, year</th>
<th>No. of cases</th>
<th>Age range (years)</th>
<th>Glioma grade (WHO)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ruel et al. 1990 (19)</td>
<td>1</td>
<td>70</td>
<td>IV</td>
</tr>
<tr>
<td>Lagares et al. 2004 (11)</td>
<td>1</td>
<td>67</td>
<td>II</td>
</tr>
<tr>
<td>Kouyatis et al. 2004 (10)</td>
<td>1</td>
<td>65</td>
<td>II</td>
</tr>
<tr>
<td>Habek et al. 2007 (7)</td>
<td>1</td>
<td>68</td>
<td>III</td>
</tr>
<tr>
<td>Frosini et al. 2008 (6)</td>
<td>1</td>
<td>66</td>
<td>II</td>
</tr>
<tr>
<td>Nakazato et al. 2013 (16)</td>
<td>1</td>
<td>80</td>
<td>II</td>
</tr>
<tr>
<td>Our case 2014</td>
<td>1</td>
<td>72</td>
<td>III</td>
</tr>
</tbody>
</table>

DISCUSSIONS

The literature is scant in reports and sometimes confusing in the number of cases presented (1,18,20). By performing an extensive review of the literature and updating the already presented series (2,8,21), we can state that a total of 59 cases have been published to the best of our knowledge (excluding the present case).

The age range of the patients is also very wide, varying between 3 months (4) and 80 years (16), with our case (age 72) being the second eldest case reported. There are too few cases reported to correctly assess the prevalence of BLTG for a certain age but we have found that there are only 6 cases reported in patients over 65, possibly implying an earlier onset of this disorder (Table I).

The presentation of BLTG is extremely varied due to the varied involvement of the thalamic tracts, nuclei and pathways of this region. Unfortunately, the early stages of the disease are silent, with the patients having mild signs and symptoms even with large tumors (14). Symptoms mainly comprise mental deterioration, personality changes, and apathy, rather than focal neurological signs (17,21). This is not the case of pediatric patients who rarely present with personality disorders even in the case of large tumors (4,8).

Hydrocephalus is usually absent or mild, and it is not the cause of the raised intracranial pressure, which is due to the direct mass effect of the tumor (4). We believe that the rapid progressive dementia in our patient was secondary to the involvement of the dorsomedial thalamic nuclei and their connections with the temporal and frontal lobes, as described by Hirano and Kouyialis (8,10).

BLTG presents with characteristic imaging features. CT scans show symmetrical isodensity lesions and do not provide eloquent information on the mass effect (18), MRI reveals a hypointense to isointense lesion in T1-weighted images and a homogeneous hyperintense lesion in T2 and FLAIR sequences. Gadolinium enhancement is not present in grade II BLGT, but minimal focal uptake has been described in grade III BLGT (12). Such a minimal focal uptake was also visible unilaterally in the left thalamus in our case (Figure 1C).

Surgical management is restricted due to the eloquence of the area, and the bilateral diffuse involvement of the thalamus. There is one anecdotal report of bilateral resection performed by the transcortical-transventricular approach (1) but no information is provided by the authors regarding gross tumor removal and the postoperative status of the patient. Rajput et al. (18) present one case of interhemispheric transcallosal approach, with approximately 30% tumor removal with high morbidity and poor post-operative results. Other authors report biopsy being performed via stereotaxy (4-8,13,17,20,21), neuronavigation guidance (3), or transventricular endoscopic approaches (2,14). One case presenting with unilateral tremor due to bilateral thalamic glioma was treated with unilateral biopsy and lesioning of the ventralis intermedius nucleus and the ventralis oralis anterior – ventralis oralis posterior complex, with good tremor control up to 1 month postoperatively (9).

The histology of BLTG is variable with a predilection being described for grade II (14). The occurrence of BLTG grade III and IV at ages over 65 years is extremely rare with only two other cases having been reported (Table I).

The role of adjuvant therapies such as radiotherapy, brachytherapy or chemotherapy remains unclear (2,4,14,18). Our patient has not received adjuvant treatment in any way and we can speculate that the natural progression of type III BLTG is extremely aggressive as our case died 6 months after the onset of symptoms.

CONCLUSION

Bilateral thalamic gliomas are extremely rare and constitute severe tumoral pathology. The origin is most probably different than unilateral thalamic gliomas (18). Even large tumors can present only with mild cognitive dysfunctions and not with focal signs at first. Surgical and adjuvant therapy do not tend to improve the survival of these patients. As presented in our case report, WHO grade III BLTG in patients over 60 years of age is even more uncommon and has a poorer outcome.

REFERENCES


Balasa A. et al: Bilateral Thalamic Glioma


