Adult Supratentorial Extraventricular Anaplastic Ependymoma: Therapeutic Approach and Clinical Review

Abstract

We report a 69-year-old patient with left paresthesia and hemiparesis. Magnetic resonance imaging revealed a right frontoparietal cystic tumor. A subtotal surgical resection was performed, and an Ommaya reservoir was left in place. The pathological diagnosis was supratentorial extraventricular anaplastic ependymoma. Radiation therapy was administered, and Ommaya reservoir drainages were performed. Four months after, her clinical status deteriorated after a reservoir drainage and image revealed an acute hemorrhage. An additional resection was carried out, and chemotherapy was undergone. One month later the tumor relapsed and the patient died 18 months after initial diagnosis. Some poor prognostic factors have been suggested in the literature: Young age, incomplete tumor resection – eloquent area location, histological anaplasia, supratentorial, and extraventricular locations. Ommaya reservoirs may be used in cystic lesions as a temporary measure only. Surgery is the mainstay of therapy with adjuvant radiotherapy and/or chemotherapy.

Keywords: Chemotherapy, Ommaya reservoir, radiotherapy, supratentorial extraventricular anaplastic ependymomas, surgery, treatment

Introduction

Ependymomas are primary tumors of the central nervous system arising from the ependymal cells lining the ventricular system and central canal of the spinal cord. They account for about 2% of all intracranial tumors; most commonly affect the pediatric population, and in the adult represent 3–5% of the glial tumors.[1–4] Although the majority of ependymomas develops infratentorially, they may also arise at the supratentorial level and rarely with no connection with the ventricular system.[2,5,6] Three hypothesis have been proposed:[7] Tumors develop from intraparenchymal or subarachnoid ependymal cysts originating from germinal matrix migration disorders; or they generate from neuroectodermal neoplasms that differentiate into the ependymal lineage; or, finally, they result from neoplastic growth of an ectopic ependymal cell, as a consequence of a migrational error.

Although the standard approach for anaplastic ependymomas is maximal surgical resection followed by postoperative focal irradiation in children,[8] it is not clear whether pediatric population management should be used in the adult supratentorial extraventricular counterpart.[4]

Case Report

A 69-year-old woman presented with a left upper limb paresthesia and weakness to the emergency room. Neurological examination disclosed grade 4 paresis of the left upper limb and left hemihypesthesia. The magnetic resonance imaging (MRI) showed a right frontoparietal lesion with cystic and solid components [Figure 1]. A right parietal craniotomy was performed with resection of the mural nodule. Considering the eloquent area and the thin cyst wall with no anatomical cleavage plan, no gross total resection was possible. An Ommaya reservoir was inserted. Postoperative MRI showed mild enhancement of the cystic wall and decreased mass effect [Figure 2]. The pathological diagnosis was supratentorial extraventricular anaplastic ependymoma (SEAE) [Figure 3].

External conformational radiation therapy was administered in 30 daily fractions up to a total of 60 Gy.

The patient experienced some symptomatic deteriorations related to cystic fluid...
content controlled with Ommaya reservoir percutaneous drainage. After one episode, the patient did not improve, and an imagiological evaluation was performed sustaining signs of acute hemorrhage within the lesion cavity after drainage [Figure 4]. Clinical evaluation showed symptomatic worsening with a left hemiparesis.

The patient was reoperated but refused an aggressive surgical resection due to the high risk of hemiplegia. The cystic content was drained, and subtotal resection of the mural nodule and cystic walls were accomplished, preserving motor function. Postoperative MRI showed a linear contrast-enhanced zone and a 12.9 mm nodule remaining [Figure 5]. Histological examination still discloses some areas of typical ependymoma although most of the parenchyma tumor now presented features of an undifferentiated, high-grade glioma.

The patient partially improved being autonomous at hospital discharged. She was started on chemotherapy with temozolomide which she did not tolerate due to pancytopenia. Her general condition deteriorated, she became partially dependent in daily living activities and died 18 months after the initial diagnosis, 9 months after reoperation.

Discussion

To the best of our knowledge, only 17 cases of adult intra-axial SEAE have been previously reported in PubMed [Table 1].[4,5,9-16] The age of diagnostic was 18–70 years (mean = 39.4 ± 18.5 years). Our patient was 69 years old, an unusual age for this diagnosis as she is in the upper tail of age distribution. No meaningful differences according to gender were noted. Regarding the topographic tumor distribution, the frontal and parietal locations were the most frequent, being responsible for 67% (12/18) of the cases, and correlated with the presenting symptoms. The imagiological appearance of a subcortical cyst with a mural nodule is common in low-grade gliomas.[17] However, supratentorial anaplastic ependymomas most frequently appear as a heterogeneous enhancing solid or cystic/solid lesion[18] which is supported by this report. Despite some cases present hemorrhage and calcifications, none was present in this case. Surgery resection was performed in all the cases with data concerning treatment; 75% (12/16) of patients underwent radiation therapy; and 31% (5/16) adjuvant chemotherapy.

A reoperation was also performed due to a hemorrhagic event. In only 2 cases,[13,16] reoperation is reported in the literature at the time of first recurrence. During the follow-up, 44% (8/18) of the tumors did not recur, 33% (6/18) had at least one episode of recurrence but for 22% (4/18) cases no data concerning recurrence exists. The minimum survival was 14 months with 44% (8/18) patients surviving for more than 4 years. In this case, disease progression was elicited 9 months after surgery, and the patient survived for 18 months.

In patients with gliomas, Ommaya reservoir provides a clinical improvement when used for recurrent aspiration in patients with cystic tumors.[11] It may be used as a temporary measure if complementary treatment or re-intervention is being administered or planned. It can also be used for
Intracystic brachytherapy which might have been an option if no hemorrhagic event had happened. Although the clinical impact of such approach is unknown, the literature supports some benefit when used in a compassionate add-on treatment.\(^{[19]}\)

The literature considers as poor prognostic factors: Young age, incomplete tumor resection, histological anaplasia, supratentorial, and extraventricular location.\(^{[6,11]}\) By definition, SEAE comprises the last three, leaving age and extension of surgical resection as important prognostic factors in these patients. We consider tumor location in eloquent areas is an important limiting factor for a complete surgical resection which is the main modifiable negative prognostic factor in SEAE. Complete resection should be attempted in all cases, and reintervention should be considered on a case basis when first surgery limiting factors are overcome.

To our best knowledge, this is the first report where an Ommaya reservoir was used for symptom control while waiting for adjuvant treatment to control cystic fluid production after incomplete resection in an eloquent area.

**Conclusion**

Adult SEAE is a rare entity raising controversy regarding its best approach. Some poor prognostic features are identified: Young age and incomplete tumor resection. Tumor location in the eloquent area is also a factor of poor outcome as it usually precludes complete resection. Ommaya reservoirs may be used in cystic lesions as a temporary measure. Gross total resection is the preferred approach followed by adjuvant treatment with radiotherapy (no consensual place for chemotherapy).

**Patient consent**

The next of kin has consented to the submission of the case report for submission to the journal.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.
Table 1: Clinical review of published SEAE

<table>
<thead>
<tr>
<th>Authors; year of publication</th>
<th>Gender, age (years)</th>
<th>Symptoms</th>
<th>Localization, size, features</th>
<th>Treatment</th>
<th>Follow-up; survival period</th>
</tr>
</thead>
<tbody>
<tr>
<td>Takeshima et al., 2002[16]</td>
<td>Female, 70</td>
<td>Consciousness disturbance and head trauma; progressive dementia</td>
<td>Right frontal lobe, intratumoral hemorrhage</td>
<td>Macroscopic total resection; second operation due to hemorrhage; bedridden</td>
<td>No recurrence for 14 months</td>
</tr>
<tr>
<td>Moritani et al., 2003[13]</td>
<td>Female, 50</td>
<td>Severe headache</td>
<td>Right temporal lobe, 20 mm</td>
<td>Macroscopic total resection; chemotherapy</td>
<td>Recurrence 7 and 13 months after first surgery with reoperation and chemotherapy and irradiation after the first recurrence. Survival for 20 months</td>
</tr>
<tr>
<td>Kojima et al., 2003[12]</td>
<td>Female, 56</td>
<td>Transient mild vertigo, seizure of the right extremities, mild dysarthria, agraphia, headache</td>
<td>Left temporal lobe, hematoma, calcification</td>
<td>Subtotal resection; focal radiation therapy</td>
<td></td>
</tr>
<tr>
<td>Shuangshoti et al., 2005[15]</td>
<td>Male, 33</td>
<td>Acute headache, vomiting, transient motor aphasia</td>
<td>Left parietal</td>
<td>Surgery; radiation; chemotherapy</td>
<td>No recurrence; survival for 18 years</td>
</tr>
<tr>
<td>Miyazawa et al., 2007[5]</td>
<td>Female, 18</td>
<td>Simple partial motor seizures for 5 months</td>
<td>Right frontal lobe; 4 cm</td>
<td>Gross total resection; radiation</td>
<td>Recurrence; death 14 months after treatment</td>
</tr>
<tr>
<td>Romero et al., 2012[14]</td>
<td>Male, 23</td>
<td>Seizures, hemiparesis, and aphasia</td>
<td>Right frontal lobe; 2,4 cm</td>
<td>Gross total resection; radiation</td>
<td>No recurrence; uneventful for 29 months</td>
</tr>
<tr>
<td>Davis et al., 2011[9]</td>
<td>Female, 22</td>
<td>Headache and dysarthria</td>
<td>Frontotemporal</td>
<td>Macroscopic total resection; radiotherapy</td>
<td>Metastatic systemic disease 1 year after treatment; 5 years follow-up</td>
</tr>
<tr>
<td>Elsharkawy et al., 2013[10]</td>
<td>Male, 25</td>
<td>Seizure, finger paresthesia</td>
<td>Frontoparietal lesion</td>
<td>Complete macroscopic removal; radiotherapy</td>
<td>Follow-up of 6 months with no recurrence</td>
</tr>
<tr>
<td>Iwamoto et al., 2014[11]</td>
<td>Male, 61</td>
<td>Severe acute headache for 3 days, consciousness disturbance, coma</td>
<td>Right temporal lobe, 40 mm, repeated intratumoral hemorrhage</td>
<td>Macroscopic total resection; focal radiation therapy (60 Gy) and chemotherapy (temozolomide)</td>
<td>Recurrence and dissemination to lower thoracic spinal cord</td>
</tr>
</tbody>
</table>

SEAE – Supratentorial extraventricular anaplastic ependymoma; IAR – ACNU-Radiation therapy; ACNU – 1-(4-amino-2-methyl-5-pyrimidinyl)-methyl-3-(2-cholroethyl)-3-nitrosourea hydrochloride

References


