Adult At/Rt: Predilection for the Pituitary?

Jacqueline M. Regan1, Mahtab Tehrani1, Fausto J. Rodriguez2 and Joseph C. Watson3*

1Inova Fairfax Hospital, 3300 Gallows Road, Department of Pathology, Falls Church, VA, 22042, USA
2Assistant Professor, Department of Pathology, Division of Neuropathology, Johns Hopkins Hospital, Sheikh Zayed Tower, Room M2101, 1800 Orleans Street, Baltimore, MD 21231, USA
3Associate Professor, Department of Neurosurgery, VCU School of Medicine, Cerebrum MD, 8230 Boone Blvd, USA

Received Date: April 22, 2015, Accepted Date: May 19, 2015, Published Date: May 27, 2015.

*Corresponding author: Joseph C. Watson, Associate Professor, Department of Neurosurgery, VCU School of Medicine, Cerebrum MD, 8230 Boone Blvd, USA, Tel: (703) 748-1000; E-mail: jwatson4@vcu.edu

Abstract

Atypical teratoid/rhabdoid tumors (AT/RT) presenting in adults are particularly rare. Interestingly there have been seven cases of AT/RT in the sellar region of adults reported thus far. In this report, we present another case that reinforces a particular pattern common to these tumors. A 45-year-old woman presented with headaches and double vision. She was found to have left sixth nerve palsy. Transsphenoidal resection for presumed apoplexy of a pituitary macroadenoma was performed but the pathology revealed an atypical teratoid/rhabdoid tumor. Adult AT/RT’s in the sellar-region are extremely rare but they do mimic macroadenomas. They may also present more commonly in women with oculomotor palsies.

Keywords: Adult atypical teratoid/rhabdoid tumor; Sixth nerve palsy; Oculomotor nerve palsy; Cranial nerve palsy; Pituitary adenoma; Pituitary tumor

Introduction

Atypical teratoid/rhabdoid tumor is an aggressive highly malignant neoplasm of the central nervous system. The tumor is well known for its heterogenous microscopic pattern, demonstrating primitive neuroectodermal, mesenchymal, epithelial and glial components and rhabdoid cells. Immunophenotypic heterogeneity is also a characteristic finding and the most striking feature is loss of INI1.

Though AT/RT’s are primarily a tumor of young children, there are extremely rare adult cases. Of the 41 previously reported adult AT/RT’s, 7 (19.0%) have been in or around the sella turcica (Table 1). All seven previously reported sellar region adult AT/RT cases occurred in females of various ages (range = 20–61, mean = 43 years) (Table 2). There is a strong female predominance (100%) with atypical teratoid/rhabdoid tumors in the sella. Our patient presented herein follows the pattern of female predominance and oculomotor palsy in adult sellar region atypical teratoid/rhabdoid tumors.

Case Report

Presentation and Examination

A 45-year-old previously healthy right-handed woman presented with complaints of a subacute onset severe headache for nine days associated with nausea and vomiting. She developed side-by-side double vision over this period of time that was not improving. She had no prior constitutional symptoms or weight loss and no family history of tumors. On examination, she did not appear Cushingoid or acromegalic. Her extraocular movements were not full to confrontation, as was the remainder of her neurological examination. A sella MRI revealed a pituitary tumor most consistent with a macroadenoma on the left side of the sella with extension into the ipsilateral cavernous sinus (Figure 1).

Operation

A transsphenoidal approach to the tumor was performed. The sella was found to be slightly enlarged, predominantly on the left. The tumor seemed to emanate from the posterior portion of the sella was found to be slightly enlarged, predominantly on the left. A sella MRI revealed a pituitary tumor most consistent with a macroadenoma on the left side of the sella with extension into the ipsilateral cavernous sinus (Figure 1).

Table 1: Summary of all AT/RT reports.

<table>
<thead>
<tr>
<th>Location</th>
<th>Absolute frequency</th>
<th>Age range</th>
<th>Gender (M:F)</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cortical</td>
<td>24 (57.1%)</td>
<td>18-50</td>
<td>17:7</td>
<td>[4–19]</td>
</tr>
<tr>
<td>Sellar region</td>
<td>8 (19.0%)</td>
<td>20-61</td>
<td>0:8</td>
<td>[20], [3], [14], [21], [2], current</td>
</tr>
<tr>
<td>Pineal region</td>
<td>3(7.1%)</td>
<td>27-33</td>
<td>1:2</td>
<td>[1,2,22,23]</td>
</tr>
<tr>
<td>Thalamus</td>
<td>1 (2.4%)</td>
<td>35</td>
<td>1:0</td>
<td>[24]</td>
</tr>
<tr>
<td>Cerebellum</td>
<td>4 (9.5%)</td>
<td>19-45</td>
<td>3:1</td>
<td>[21, 25–27]</td>
</tr>
<tr>
<td>Spinal Cord</td>
<td>2 (4.8%)</td>
<td>21-43</td>
<td>0:2</td>
<td>[6,28]</td>
</tr>
<tr>
<td>Total</td>
<td>42</td>
<td>18-61</td>
<td>22:20</td>
<td></td>
</tr>
</tbody>
</table>

Table 2: Summary of all sellar-region AT/RT reports.

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>Presentation</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>32</td>
<td>F</td>
<td>Headache</td>
<td>[3]</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Nausea</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Sudden onset of visual disturbance</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>20</td>
<td>F</td>
<td>Loss of vision in the right eye due to tumor</td>
<td>[21]</td>
</tr>
<tr>
<td>3</td>
<td>31</td>
<td>F</td>
<td></td>
<td>[21]</td>
</tr>
<tr>
<td>4</td>
<td>56</td>
<td>F</td>
<td>Right abducens and oculomotor nerve palsy</td>
<td>[20]</td>
</tr>
<tr>
<td>5</td>
<td>46</td>
<td>F</td>
<td>Headsaches</td>
<td>[14]</td>
</tr>
<tr>
<td>6</td>
<td>61</td>
<td>F</td>
<td>Left abducens palsy</td>
<td>[2]</td>
</tr>
<tr>
<td>7</td>
<td>57</td>
<td>F</td>
<td>Headache</td>
<td>Current</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Double vision</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Right oculomotor nerve palsy</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>45</td>
<td>F</td>
<td>Headache</td>
<td>Current</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Double vision</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Left abducens palsy</td>
<td></td>
</tr>
</tbody>
</table>
anterior lobe, as there was some normal gland over the tumor. A subcapsular dissection was performed to remove the sella portion of the tumor and spare the gland. There were some areas of hemorrhage within this specimen, consistent with a presumed apoplectic event. Approximately half of the tumor mass was located lateral to the gland in the left cavernous sinus. We removed the medial wall of the left cavernous sinus and resected the tumor for a gross total resection, despite its relation to the carotid. The tumor itself was more firm than a typical adenoma, but otherwise was not grossly different than what we had expected to see. Cavernous sinus bleeding was controlled with gentle packing. There was no cerebral spinal fluid leak.

**Post-operation Follow-up**

Postoperatively, she had preserved pituitary function but at one month post-surgery, she still has a dense left CN VI palsy and her tumor has recurred in the cavernous sinus and tentorium (Figure 2). The metastatic workup with a bone scan and chest, abdomen, and pelvis CT was negative. She received stereotactic radiosurgery to the cavernous sinus and parasellar region. Her cranial nerve deficits progressed and her mental status declined. She died in hospice care 6 months after surgery. As described in Shonka et. al. [1] the range of survival after diagnosis for adult AT/RT’s is extremely wide [1] though most patients succumb to the disease within a year.

**Pathologic Findings**

Microscopically, the tumor cells were arranged in sheets. The neoplastic cells showed high degree of anaplasia with a high nuclear to cytoplasmic ratio, scant eosinophilic cytoplasm, vesicular enlarged nuclei and prominent nucleoli. Focally the tumor cells had cytoplasmic vacuoles. Mitoses were numerous. By immunohistochemical stains the tumor cells were positive for cytokeratin (CK8/18), CD56, p53 and focally for epithelial membrane antigen (EMA), pancytokeratin, smooth muscle actin (SMA) and S100. The neoplastic cells were negative for synaptophysin, chromogranin, HMB45, Mart-1, cytokeratin 7, cytokeratin 20, GCDFP15, mammoglobulin, desmin. Germ cell and lymphoma markers were negative. Pituitary hormones were positive in the surrounding benign pituitary tissue but negative in the tumor cells. INI1 staining was absent in the tumor cells but present in the benign background lymphocytes and vessels (positive internal control).

**Discussion**

AT/RT’s in adults are rare indeed. Considering our case, and those previously reported, a clinical pattern is emerging. There appears to be a predilection for the sella; now 8 of 42 (19%) reported adult AT/RT patients had sella tumors. Of these sellar region patients, 50% have presented with cranial nerve palsies (Table 2), with an abducens’ palsy being the single most common type, likely due to its relative vulnerability within the cavernous sinus. Headache was also a presenting complaint in these patients.
with sellar AT/RT's although obviously non-specific, it suggests that these tumors either hemorrhage, as our case did, or are rapidly growing.

With the high rate of adult AT/RT's (19%) in the sella, what is intriguing further still is that all of these sellar region AT/RT's have been in females. Though it cannot be concluded that sellar region AT/RT's are sex-specific, the female prevalence is notable. Typically, pediatric and adult AT/RT's have a male predominance (male to female ration in children 2:1, in adults 1.1:1) [2.3](Table 1). The sellar region significantly skews the gender ratio of adult AT/RT's towards the females (8 of the reported 20 female cases bore sella tumors).

Another characteristic of adult AT/RT's in the sella is the presentation with optic nerve, abducens, or oculomotor palsies and headache. In this way it is similar to that of the presentation of a typical apoplektisic pituitary adenoma. This similarity in presentation makes it indistinguishable from pituitary apoplexy. With review of the current case and analysis of previous reports a significant pattern has arisen in the 42 reported cases of AT/RT; a seeming predilection for the pituitary, a complete female predominance in the sellar region, and common presentation of cranial nerve palsies in sellar region AT/RT patients.

Conclusion

Based on our case and a review of the published reports, adult AT/RT's in the sella present in females in a manner that is clinically indistinguishable from apoplexy. Clinicians should be aware of this possibility in the differential of apoplexy, which (excluding post-partum hemorrhage) is otherwise dominated by hemorrhagic or infarcted adenomas.

References

*Corresponding Author:* Joseph C. Watson, Associate Professor, Department of Neurosurgery, VCU School of Medicine, Cerebrum MD, 8230 Boone Blvd, USA, Tel: (703) 748-1000; E-mail: jwatson4@vcu.edu

Received Date: April 22, 2015, Accepted Date: May 19, 2015, Published Date: May 27, 2015.

Copyright: © 2015 Jacqueline M. Regan, et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.