Abstract

Objective: Surgical resection, embolization, radiation therapy, and stereotactic radiosurgery have been used to treat glomus jugulare tumors (GJT). However, the optimal treatment of these tumors is still unclear. The authors report their data on treatment of GJTs with gamma knife radiosurgery (GKS).

Design: Retrospective review and pooled analysis.

Methods: Fifteen patients (9 female, 6 male) were treated with GKS at a single tertiary care institution for GJTs over a 14-year period. Criteria for selection included GKS followed by at least one post-treatment radiographic image and volumetric analysis was performed. A 15% change in tumor volume was required to be considered real. Pooled analysis was performed to compare with other series.

Results: The mean total radiological follow-up was 43.2 months. The mean dose to the tumor margin was 14.6 Gy. The mean tumor size at presentation was 7.3 cc and 6.3 cc at last follow-up. After treatment, 7 tumors decreased (46.7%), 5 remained unchanged (33.3%), and 3 (20%) grew on imaging. Treatment failures received a mean marginal dose of 13.2 Gy compared with 15.1 Gy for treatment successes (p = 0.08). Overall tumor control rate after GKS in the existing literature with inclusion of the present study is 90.5%.

Conclusions: GKS is an effective treatment option for patients with GJTs, including those with prior surgical resection. Marginal radiation doses greater than 13 Gy may be optimal for tumor control. Longer follow-up periods will better define the benefits and risks of stereotactic radiosurgery in treating patients with GJT.

Level of Evidence: 2B

Introduction

Glomus jugulare tumors (GJT) arise from paraganglionic tissue of the jugular bulb.

Incidence: ~1:1 million

Benign and slow growing

Signs and symptoms, commonly including headache, pulsatile tinnitus, aural fullness, dizziness, and lower cranial nerve palsies

Treatment: surgical resection, embolization, radiation therapy, and, more recently, stereotactic radiosurgery (SRS).

SRS is thought to deliver a high dose of radiation to a limited area with a sharp dose decrease at the margin causing cellular and vascular damage and eventually tumor necrosis.

SRS with Gamma Knife has shown promise in treating well-circumscribed, vascular neoplasms such as glomus jugulare tumors.

Methods

University of Virginia (UVA) data collection

An Institutional Review Board-approved retrospective review was performed of 15 patients treated from 1992 through 2006 at the University of Virginia for a diagnosis of glomus jugulare tumor.

Follow-up

Radiologic follow-up consisting of at least one neuroradiologic image was available for all 15 identified patients.

Our definition of change in tumor size was any 15% deviation in volume.

Stable tumors and those that shrunk were considered treatment successes.

Those that increased in size were considered failures

Literature search

Systematic review of PubMed

11 unique studies identified in the existing literature, with the addition of our present series formed the basis of our pooled analysis.

Definitions of tumor growth varied.

Results

UVA patient characteristics

15 patients treated with GKS for GJT between 1992 and 2006

Table 1 summarizes the demographics

Tumor control and treatment parameters

Mean radiographic follow-up = 43.2 months (range 5 to 168 months)

7 tumors (46.6%) decreased in volume, 5 remained unchanged (33.3%), and 3 (20.0%) increased.

80.0% tumor control as defined by this study.

Mean (+/- standard error of the mean) GKS margin dose of radiation administered was 14.6 +/- 1.6 Gy.

Paired t-test analysis demonstrated no statistical difference (p>0.05) between the treatment success group and the treatment failure group for:

• number of isocenters

• margin dose

• maximum treatment dose

Pooled data

11 unique studies in literature (Table 2).

Cohorts ranged from 5 to 66 patients, yielding 229 patients.

All studies characterized radiologic successes as those tumors that remained stable or decreased in size.

Tumor size decreased in 80, remained unchanged in 111, increased in 6 patients, and 12 lost to follow-up.

The overall pooled rate of successful treatment with GKS, with inclusion of the patients in the present series, is 90.5% (95% CI 85.7 – 94.4%).

Discussion

• Literature present control rates after GKS ranging from 76 – 100%

• Only surgery offers the possibility of complete tumor removal, but also carries the possibility of significant morbidity.

• Radiation is not thought to directly kill tumor cells, but rather to create fibrosis and destruction of feeding vessels leading to tumor necrosis.

• The fibrosis complicates future surgical procedures.

• Not all tumors can be treated with radiosurgery due to large tumor size or location.

• Complications and morbidities associated with GKS are rare.

• Based on a paired t-test, we did not find a statistical relationship but a trend between marginal dose and failure rates was evident in our patient population (p = 0.08).

Conclusions

GKS is a reasonably effective treatment option for patients with glomus jugulare tumors both as a primary and secondary treatment. In the published literature, even after attrition is accounted for, 90.5% of individuals had successful outcomes at the time of last radiologic follow-up.

The ideal treatment dose has not yet been established, but data suggests that a tumor margin doses greater than 13 Gy may be optimal for achieving tumor control.

Acknowledgments

The authors acknowledge Ladislaus Steiner, MD for sharing his treatment data for many of those patients. The efforts and collaboration of the neuroradiologists is also tantamount to being able to carry out these studies.

References


Table 1: Characteristics of the patients treated at the University of Virginia for glomus jugulare tumors with Gamma Knife radiosurgery.

<table>
<thead>
<tr>
<th>Author</th>
<th>No. pts</th>
<th>Mean Follow-up (mo)</th>
<th>Mean Marginal dose (Gy)</th>
<th>% success</th>
<th>95% CI</th>
<th>Definition of failure/relapse</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bitaraf</td>
<td>16</td>
<td>18.5</td>
<td>18</td>
<td>87.5</td>
<td>60.4-97.8%</td>
<td>any vol change</td>
</tr>
<tr>
<td>Eustachio</td>
<td>19</td>
<td>86.4</td>
<td>Not reported</td>
<td>94.7</td>
<td>71.9-99.7%</td>
<td>2mm change</td>
</tr>
<tr>
<td>Ganz</td>
<td>14</td>
<td>28.6</td>
<td>13.6</td>
<td>100</td>
<td>73.2-100%</td>
<td>Not reported</td>
</tr>
<tr>
<td>Gerosa</td>
<td>20</td>
<td>50.9</td>
<td>17.3</td>
<td>95</td>
<td>73.1-99.7%</td>
<td>20% change</td>
</tr>
<tr>
<td>Jordan</td>
<td>8</td>
<td>27</td>
<td>16.3</td>
<td>87.5</td>
<td>46.7-98.3%</td>
<td>Not reported</td>
</tr>
<tr>
<td>Liscak</td>
<td>52</td>
<td>24</td>
<td>NR</td>
<td>90.4</td>
<td>78.2-96.4%</td>
<td>Not reported</td>
</tr>
<tr>
<td>Polslock</td>
<td>42</td>
<td>44</td>
<td>14.9</td>
<td>90.5</td>
<td>76.5-96.9%</td>
<td>2mm change</td>
</tr>
<tr>
<td>Sanger</td>
<td>13</td>
<td>50.4</td>
<td>12.5</td>
<td>100</td>
<td>71.7-100%</td>
<td>Change in greatest dimension</td>
</tr>
<tr>
<td>Sheehan</td>
<td>8</td>
<td>32</td>
<td>15</td>
<td>87.5</td>
<td>46.7-93.9%</td>
<td>any vol change</td>
</tr>
<tr>
<td>Varma</td>
<td>17</td>
<td>48</td>
<td>15</td>
<td>76.5</td>
<td>49.8-92.2%</td>
<td>Not reported</td>
</tr>
<tr>
<td>Willen</td>
<td>5</td>
<td>19</td>
<td>15</td>
<td>100</td>
<td>46.3-100%</td>
<td>Not reported</td>
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</table>

Table 2: Review of the literature of patients treated with Gamma Knife radiosurgery for glomus jugulare tumors.

<table>
<thead>
<tr>
<th>TOTAL No. pts</th>
<th>Age at treatment (yrs)</th>
<th>Sex</th>
<th>Presenting Symptoms</th>
<th>Fisch tumor class</th>
<th>Total patients (U of VA)</th>
</tr>
</thead>
<tbody>
<tr>
<td>229</td>
<td>30-84</td>
<td>Male</td>
<td>Hearing loss</td>
<td>C1-3</td>
<td>15</td>
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<tr>
<td></td>
<td>60</td>
<td>Female</td>
<td>Pulsatile tinnitus</td>
<td>D1-2</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Unsteadiness</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Cranial n palsies</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Axial MRI of a patient near the time of GKS (left) and 76 months after GKS treatment (right) illustrating a 62.5% decrease (0.8 cc to 0.3 cc) in tumor volume. The arrows indicate the glomus jugulare tumor in this patient.

Axial MRI of a patient near the time of GKS (left) and 76 months after GKS treatment (right) illustrating a 62.5% decrease (0.8 cc to 0.3 cc) in tumor volume. The arrows indicate the glomus jugulare tumor in this patient.

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