ABSTRACT

BACKGROUND: Paragangliomas of the larynx are exceedingly rare tumors, with less than 80 reported cases in the literature. These submucosal vascular lesions arise from neural crest cells and are effectively treated with surgical resection. A role for external beam radiation in this disease has not yet been explored. We present the successful management of four cases of laryngeal paragangliomas treated at a large tertiary-care cancer center over a 35 year period and review the diagnostic and therapeutic management of this disease.

DESIGN: 124 cases of head and neck paragangliomas treated at a single institution from 1970 to 2005 were retrospectively studied. Patients with laryngeal paragangliomas were identified, and a comprehensive clinico-pathological review was undertaken.

RESULTS: We identified 4 patients with tumors arising in the larynx at the following subsites: supraglottis (2), glottis (1), and subglottis (1). Three patients were treated successfully with surgery alone; two with total laryngectomy and one with a supraglottic laryngectomy. One patient with a glottic paraganglioma was treated with definitive radiation alone and had no tumor progression after five years of follow-up. At last follow-up, no patients developed local recurrence.

CONCLUSIONS: Surgical resection has been advocated in the past for these lesions and still remains the gold standard for treatment. Furthermore, most patients will require a total laryngectomy for oncological control.

CLINICAL SIGNIFICANCE: 4 cases of laryngeal paragangliomas are added to the literature base. Though predominately surgical management has been used in the past, radiation is now a treatment consideration for these tumors. Pathological findings and clinical management of epiglottic and subglottic paragangliomas are compared to the more common ventricular paragangliomas. Finally, with only one other case of metastatic paraganglioma in the literature, a treatment plan for this rare presentation is explored.

STUDY DESIGN AND SETTING

A retrospective review of the last 35 years was IRB approved. 124 patients with paragangliomas were identified by ICD-9 codes. 5 cases of pharyngolaryngeal paragangliomas were identified. 1 patient with a hypopharyngeal paraganglioma was excluded.

Case #1 (Fig. 1, 2)

- An 85 year old woman presented with a right supraglottic lesion
- The mass extended to the epiglottis
- The tumor was diagnosed as spindle cell CA
- One year evaluation showed progression
- Biopsy revealed paraganglioma
- Patient preferred external beam radiation
- On 5 year follow-up, tumor was stable

Case #2 (Fig. 3, 4, 5, 6, 7)

- A 39-year-old black female presented with a glottic lesion
- Tracheostomy was done for airway protection due to tumor bulk
- The lesion extended from the posterior aspect of the left arytenoid to the epiglottis superiorly and to the subglottis inferiorly
- Treated with embolization and an extended supraglottic laryngectomy
- On last follow-up she had no evidence of disease

Case #3

- A 67 y/o woman presented with a submucosal glottic tumor
- The mass extended to the vocal folds at the anterior commissure
- A total laryngectomy was performed
- Cartilage destruction evident on histology
- The patient died later from other causes

Case #4

- A 50 year old man presented with a recurrent mass of the left supraglottis
- Previously treated with a supraglottic laryngectomy
- A surgical salvage total laryngectomy was done
- 8 months after surgery, the patient developed lung and brain metastasis
- He was treated with systemic chemotherapy
- The patient died 16 months after diagnosis

CONCLUSION

- Immunohistochemistry is the standard of care for diagnosis
- Surgical resection is the standard of care for treatment
- Due to unique circumstances, radiotherapy was shown to be a valid option for treatment
- The second case of distant metastatic disease from a laryngeal paraganglioma is presented
- The first regionally metastatic laryngeal paraganglioma is added to the literature

<table>
<thead>
<tr>
<th>Sx vs Rad</th>
<th>Type of Sx</th>
<th>Functioning</th>
<th>Multicentric</th>
<th>Misdiagnosis</th>
<th>Familial Recurrence</th>
<th>Death</th>
<th>Metastasis</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Sx</td>
<td>SGL/TL</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>SG</td>
</tr>
<tr>
<td>2 Sx</td>
<td>TL</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>RM/DM</td>
</tr>
<tr>
<td>3 Rad</td>
<td>None</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Subglottis</td>
</tr>
<tr>
<td>4 Sx</td>
<td>ESGL</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Left SG/G</td>
</tr>
</tbody>
</table>