CASE REPORT

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DIAGNOSIS AND TREATMENT OF SUPRAGLOTTIC LARYNGEAL PARAGANGLIOMA: REPORT OF A CASE

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Abstract: Background. Paragangliomas of the larynx are unusual tumors that are seen as a vascular submucosal mass. These usually are seen in the supraglottic larynx but have also been found in the subglottis. This is the only laryngeal neuroendocrine tumor with a female predilection. It is important that paragangliomas be differentiated from other neuroendocrine tumors of the larynx, including atypical carcinoid, because of differing treatment modalities.

Methods. We present the clinical, radiologic, and pathologic findings of a supraglottic laryngeal paraganglioma seen in a 50-year-old woman with a 6-month history of slowly progressive hoarseness.

Results. The tumor was successfully approached by means of a midline laryngofissure with mucosal preservation. The patient remains disease free 24 months after surgery.

Conclusions. Preoperative CT and angiography are useful in making the diagnosis of paraganglioma before surgical intervention. Complete excision through an external mucosa-sparing approach is the treatment of choice. Distinguishing laryngeal paraganglioma from other neuroendocrine tumors can be difficult. Immunohistochemistry is an important tool for the correct pathologic diagnosis. © 2004 Wiley Periodicals, Inc. Head Neck 26: 94–98, 2004

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Laryngeal paragangliomas are uncommon neuroendocrine neoplasms of neural origin that are thought to arise from the superior or inferior laryngeal paraganglia.1 This is in contrast to neuroendocrine tumors of epithelial origin (carcinoid tumor, atypical carcinoids, and small cell neuroendocrine carcinomas). More than 90% of laryngeal paragangliomas occur in the supraglottic larynx.1 Unlike other neuroendocrine tumors of the larynx, only laryngeal paragangliomas show a female preponderance (male–female ratio of 1:3).2 The distinction between paragangliomas and neuroendocrine carcinomas of the larynx can be a diagnostic challenge. Although this group of neoplasms shares similarities, the biologic behavior, management, and prognosis differ. Therefore, diagnostic accuracy is paramount.3

Surgical removal of paragangliomas of the larynx has been described using multiple approaches, including supraglottic laryngectomy and lateral pharyngotomy.4–7 Because this is a benign submucosal lesion, a conservative resection is preferred.

We present the clinical, radiologic, and pathologic findings of a supraglottic laryngeal para-
ganglioma. The surgical approach to this tumor by means of a midline laryngofissure is discussed.

CASE REPORT
A 50-year-old woman was referred for a 6-month history of slowly progressive dysphonia. The patient was a nonsmoker and nondrinker. Although the patient denied any breathing or swallowing difficulty, she did complain of a feeling of fullness in her throat. Endoscopic examination revealed a submucosal mass involving the right side of the larynx, extending from the level of the aryepiglottic fold to the right true vocal cord (Figure 1). Both vocal cords were normally mobile. The surface of the mass contained a network of dilated blood vessels (Figure 2). The subglottis was normal, as was the base of the tongue and pharynx. No stridor or airway obstruction was noted. No neck lymphadenopathy was appreciated.

CT with contrast demonstrated an enhancing right supraglottic mass consistent with a vascular mass (Figure 3). MRI with gadolinium showed a 3-cm, well-circumscribed, homogeneously enhanc-
ing mass extending from the right vallecula to the level of the right true vocal cord (Figure 4). On the basis of the radiologic findings, the differential diagnosis included paraganglioma, hemangioma, and schwannoma.

Because of the possibility of a paraganglioma, the patient had evaluation of serum and urine catecholamine levels, which were normal. An elective tracheotomy was performed under local anesthesia to secure the patient’s airway before undergoing an arteriogram and embolization. Arteriography demonstrated that the right superior thyroid artery supplied greater than 90% of the blood supply to the laryngeal mass. Arteriovenous shunting was observed within the tumor. Embolization of the tumor was performed without complication.

The patient underwent midline laryngofissure and submucosal resection of the tumor. After opening the larynx, a mucosal incision was made over the mass in the supraglottic mucosa in an anterior to posterior direction, paralleling the false vocal cord. The mass was encapsulated and could be safely dissected from the overlying mucosa and surrounding musculature of the larynx. The inferior extent of the tumor reached the ventricle and did not involve the vocal ligament, which was preserved (Figure 5). Intraoperative blood loss was minimal. The patient had an uneventful postoperative course, and her tracheotomy tube was removed within 2 weeks of the resection. Her voice quality is excellent, and she has not had a recurrence of the lesion at 2 years follow-up (Figure 6).

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Pathologic Findings. Gross examination of the surgical specimen revealed a firm, 3- × 2-cm ovoid, well-circumscribed mass with a thin fibrous capsule (Figure 7). Microscopic examination showed polygonal epithelioid cells arranged in discrete nests (Zellballen pattern) (Figure 8). The cells contain abundant cytoplasm and hyperchromatic nuclei, which at times were pleomorphic. The nests were surrounded by scant stroma containing a delicate vascular network. The tumor cells had strong immunoreactivity with chromogranin but not cytokeratin (Figure 9). The chief cells of paragangliomas are immunoreactive for chromogranin and usually negative for cytokeratin stains. In addition, S-100 immunostains highlighted the elongated sustentacular cells surrounding the chief cells. These findings confirmed the diagnosis of laryngeal paraganglioma.

DISCUSSION
Paragangliomas arising in the larynx are rare. Sixty-five cases of laryngeal paraganglioma have been reported, with a mean age of 44 years (range, 14–80 years). In one review, reported mean duration of symptoms was 26 months. Dysphonia is the most commonly reported symptom, followed by stridor, dysphagia, and dyspnea. Unlike other neuroendocrine tumors of the larynx, there is a definite female preponderance. In the larynx, paragangliomas are seen as a submucosal mass. Of the reported cases, 90% occurred in the supraglottic larynx, but they might arise in the glottic and subglottic region. Rarely has a laryngeal paraganglioma been functionally active (2.9%), and there are no reported cases of paraneoplastic syndromes. A recurrence rate of 17% has been reported in a series of 30 cases with long-term follow-up. Paragangliomas can infrequently have focal necrosis, mitoses, and vascular invasion. These features are not synonymous with malignant behavior. Only one accepted case of metastatic laryngeal paraganglioma has been reported in the literature.

Immunohistochemistry is the most useful method in distinguishing neuroendocrine tumors of the larynx. There can be an overlap of histologic features of paraganglioma, atypical carcinoid, and melanoma. Paragangliomas are composed of chief cells and sustentacular cells. The chief cells demonstrate immunoreactivity to neuroendocrine markers, including chromogranin, synaptophysin, and neuron-specific enolase and are usually negative for epithelial markers including cytokeratin. Atypical carcinoids of the larynx show immunoreactivity for both neuroendocrine markers and epithelial markers. The sustentacular cells do show immunoreactivity with S-100, but the chief cells are negative for melanoma markers S-100, HMB45, and melan-A, distinguishing this from melanoma.

Surgical removal of paragangliomas should allow complete removal while minimizing functional deficits. For laryngeal paragangliomas, the endoscopic approach provides limited exposure and access to the lesion. This is not a good approach for vascular tumors, because hemorrhage might obscure visualization of tissue planes. Supraglottic laryngectomy has been reported as a treatment for supraglottic paraganglioma, but because the tumor usually does not invade the mucosa, this degree of mucosal resection is not necessary. An external approach and submucosal dissection is preferable, because it minimizes trauma to the mucosa, thereby limiting postoperative complications associated with supraglottic laryngectomy such as dysphagia and webbing. The midline laryngofissure with submucosal dissection was used successfully in this case. A mucosal incision was necessary in this case to allow dissection of the underlying tumor, but complete mucosal preservation was accomplished. This resulted in no postoperative dysphagia and excellent voice preservation. A temporary tracheotomy is necessary to secure the airway during the operative and postoperative period. A lateral thyrotomy approach has also been described for removal of supraglottic submucosal tumors, including paraganglioma. This can provide access for submucosal resection but with more limited access. A lateral thyrotomy approach can be per-
formed without a tracheotomy for smaller masses, but for resection of vascular masses, a tracheotomy should be performed.

CONCLUSIONS

Paragangliomas of the larynx are unusual tumors that are seen as a submucosal mass. These tumors must be differentiated from other neuroendocrine tumors of the larynx, including atypical carcinoid, because of differing treatment modalities. Preoperative CT and angiography can aid in the diagnosis of parapharyngeal tumors. Conservative but complete excision through an external approach with mucosal preservation is the treatment of choice.

REFERENCES


