Recurrent Laryngeal Neurofibroma Not Associated with Neurofibromatosis Type I: Case Report & Literature Review

Jeffrey M. Phillips, MD; Xin Gu, MD; and Cherie-Ann Nathan, MD, FACS

1 Department of Otolaryngology-Head & Neck Surgery, 2 Feist-Weiller Cancer Center, 3 Department of Pathology

ABSTRACT

Neurofibromas of the larynx are rare entities that may be found in association with Neurofibromatosis type 1 (NF-1), an autosomal dominantly-inherited systemic disorder with neurocutaneous, skeletal, and endocrine manifestations. Recurrent laryngeal neurofibromas are known to arise from a variety of upper aerodigestive tract symptoms, including dysphonia, stridor, dyspnea, and globus sensation, depending on the size and location of the mass. The supraglottic larynx is most commonly affected, with the arytenoids and aryepiglottic folds the most frequent subsite. Despite its first description in 1925, there have been only a few reports of solitary laryngeal neurofibromas in the literature and no reports of neurofibromas arising from the recurrent laryngeal nerve. Early diagnosis and surgical management is critical in the treatment of patients with laryngeal neurofibromas, as larger lesions may obstruct the airway and be difficult to excise endoscopically in a minimally-invasive approach. Imaging with contrast-enhanced CT is critical in determining the extent of the tumor and is critical in the treatment of progressive dysphonia, dysphagia, and globus sensation, depending on the size and location of the mass. The supraglottic larynx is most commonly affected, with the arytenoids and aryepiglottic folds the most frequent subsite. Despite its first description in 1925, there have been only a few reports of solitary laryngeal neurofibromas in the literature and no reports of neurofibromas arising from the recurrent laryngeal nerve.

METHODS AND MATERIALS

This is a case report of a 36-year-old Caucasian male who was referred to our cancer center in November 2011 for complaints of progressive dysphonia, dysphagia, and globus sensation. Medical history, examination and flexible laryngoscopy was reviewed, revealing right vocal cord paralysis and a submucosal mass deviating the right arytenoid medially. CT imaging was obtained and reviewed. The patient underwent transoral endoscopic excision of the submucosal mass causing deviation of the right arytenoid. CT imaging revealed a well-defined, non-enhancing mass at the level of the posterior glottis. Transoral endoscopic surgical excision of the mass was performed.

RESULTS

A healthy, 36-year-old Caucasian male was referred to our cancer center due to progressively worsening dysphonia and dysphagia over ten months. He reported dysphagia to liquids and globus sensation. Past medical and surgical histories were unremarkable and the patient was a non-smoker. Examination was unrevealing for neck masses, palpable lymph nodes, or any back-lying or mental defects. Flexible laryngoscopy revealed a 2 cm submucosal mass deviating the right arytenoid and posterior glottis to the left, with paresis of the right true vocal cord.

CONCLUSIONS

Neurofibromas of the larynx are commonly associated with Neurofibromatosis, type 1, an inherited disorder with variable expressivity of neurocutaneous, skeletal, and other manifestations. There have been few reports of solitary neurofibromas, not associated with this disorder. These tumors exhibit slow growth and typically arise from the supraglottic larynx at the arytenoids or aryepiglottic fossae. These areas are rich in terminal nerve plexuses and anastomoses between the superior and recurrent laryngeal nerves. Patients often present with progressive dysphonia, dysphagia, globus, or dyspnea depending on the size and location of the tumor. Flexible laryngoscopy reveals a prominent submucosal mass, often deviating the airway. Imaging with contrast-enhanced CT is critical in assessing the extent of tumor and in surgical planning. Neurofibromas often appear hypodense, however may enhance moderately after contrast circulation. Enhancement and tumor heterogeneity may be variable on MRI, however this modality is useful in delineating the tumor’s relationship to surrounding soft tissue structures.

REFERENCES