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Laryngeal Schwannoma: Emphasis on Treatment with Surgical Instrument Excision

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Abstract **Background:** Laryngeal schwannomas are rare benign tumors of schwann cell origin and commonly present in the supraglottic area. Patients present with non-specific symptoms such as dysphagia and dysphonia with the occasional patient presenting with more severe symptoms such as stridor. **Case Presentation:** A 65-year-old female presented to the clinic with symptoms of hoarseness and globus sensation for three months. Fiberoptic flexible nasolaryngoscopy revealed a mass at the right arytenoid, appearing consistent with a mucosally covered cyst-like lesion. Computed Tomography (CT) of the neck showed a lobular hypoenhancing mass at the level of the aryepiglottic fold. A schwannoma was diagnosed via histopathology, positive for S-100. **Conclusion:** Tailored surgical excision via an intraoral approach successfully removed the laryngeal schwannoma with no recurrence at one-year follow up. This case highlights the clinical presentation, diagnosis, surgical treatment with outcome of this rare laryngeal tumor.

Keywords: *laryngeal schwannoma, neurogenic tumor, larynx*

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1. Introduction

Schwannomas are a type of benign neurogenic tumor originating from schwann cells. Around 45% of neurogenic tumors present in the parapharyngeal space [1]. However, schwannomas presenting in the larynx are extremely rare, and account for less than 1.5% of all benign laryngeal tumors [2]. Laryngeal schwannomas are indolent growths that primarily appear in the supraglottic area. Symptoms include dysphagia, dysphonia, odynophagia and stridor due to mass effect. Histopathology is definitive for diagnosis, which includes a positive S-100 immunohistochemical stain [3]. Surgical excision is typically required for treatment.

2. Case Presentation

A 65-year-old female with a past medical history of post nasal drip, gastroesophageal reflex, hypertension, psoriasis, and diabetes mellitus presents to the Otolaryngologist for evaluation of dysphonia and a cough. The patient reports symptoms of voice change and post nasal drip for the past 3 months with an associated cough. She also states she has been taking Azelastine as prescribed by her PCP, with no relief of symptoms. Patient is an ex-smoker over 20 years and drinks alcohol on occasion.

On physical examination, mild nasal mucosa edema was noted but no physical abnormalities of the neck were observed. Additionally, flexible nasopharyngoscopy was preformed noting a mass arising from what appeared to be her right arytenoid, making it difficult to completely visualize her vocal cords. The mass did not appear ulcerated or irregular in appearance, but rather as well-defined cystic appearing mass. CT scan of the neck was performed, showing a low attenuating mass at the level of the aryepiglottic fold posteriorly and right of the midline. It measured 2.4 x 1.9 x 1.3 cm. No lymphadenopathy was noted.

Microlaryngoscopy was performed with the goal of obtaining a biopsy for pathologic evaluation. The procedure was performed under a general anesthesia with a 5.0 endotracheal tube inserted with the assistance of the Glideslope without difficulty. The Dedo laryngoscope was inserted, allowing for visualization under the microscope as shown in Figure 1. The mass protruding from the right arytenoid process with a ball and valve effect. On biopsy of the lesion, a small amount was obtained to evaluate on frozen section. The initial frozen section diagnosis suggested the mass was consistent with a schwannoma.

With the second biopsy attempt, the entire encapsulated mass approximately 3 cm, pealed out from the arytenoid mucosa and was resected in its entirety as shown in Figure 2. The complete specimen was sent for formal pathologic evaluation.

The patient followed up in the clinic one week later and reported feeling well with mild throat discomfort. She

denied having any hoarseness of her voice or difficulty swallowing. The globus sensation also improved. She was counselled regarding the distinct possibly of reoccurrence. The patient elected to proceed with close follow up with repeat flexible laryngoscopy as opposed to further surgical resection. At last follow up, one year after resection, she remains symptom free and no evidence of recurrence has been noted on flexible laryngoscopy.

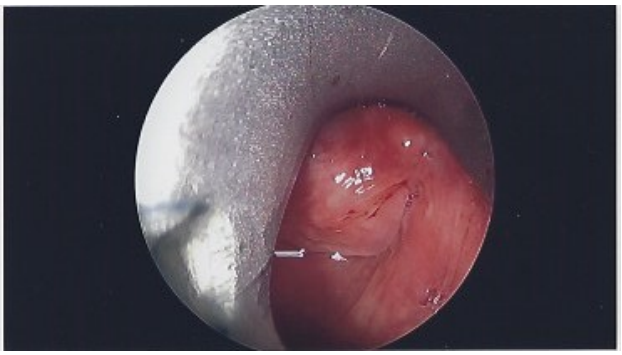

Figure 1. View of the schwannoma visualized via laryngoscope

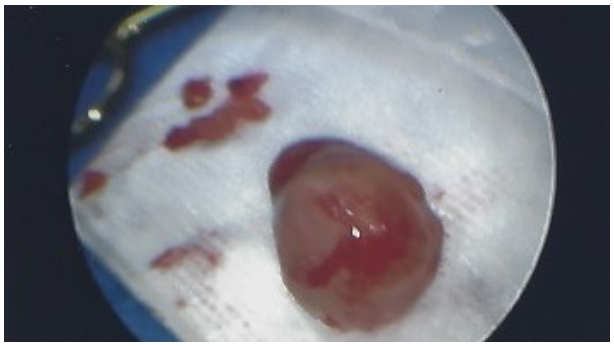

Figure 2. Excised laryngeal schwannoma in its entirety

3. Discussion

Schwannomas are one of two benign neurogenic tumors that rarely present in the larynx, the other being a neurofibroma [4]. Since being first documented in 1925 by Shouchnek et al, less than 200 cases have been reported in literature [5]. It is important to differentiate the two neurogenic tumors, as the surgical outcome varies. Schwannomas are encapsulated tumors arising from the perineural schwann cells, growing extrinsically in relation to the parent nerve; able to be easily excised while sparing the nerve of origin [6]. Neurofibromas in turn originate from perineural fibrocytes, which are within the nerve fascicles, and grow intermingled with the parent nerve; making surgical removal of this tumor from the originating nerve theoretically impossible [7]. In addition, neurofibromas may be associated with neurofibromatosis, a genetic condition affecting multiple organ systems, such as the CNS, eyes and skin.

Of reported schwannomas, approximately 25-35% present in the head and neck region, but only 0.1-1.5% have been reported as tumors of the larynx [1]. The incidence of laryngeal schwannomas is higher in women during the 4^{th} to 5^{th} decades of life [8]. However, no age group is immune, as in this case the patient presented in her 6^{th} decade of life.

In concordance with this case, the most common site of laryngeal schwannomas presents in the supraglottic region: aryepiglottic folds, arytenoids, and false vocal cords [9]. The nerve of origin for laryngeal schwannomas is presumed to be the internal branch of the superior laryngeal nerve [10]. Given the slow growing nature of the tumor, presenting symptoms are related to gradual mass effect over the period of months to years. Symptoms include sore throat, odynophagia, dysphonia, globus sensation, dyspnea and stridor. The differential diagnosis includes: papilloma, hemangioma, traumatic neuroma, leiomyoma and malignant peripheral nerve sheath tumor.

Diagnostic workup of a laryngeal schwannoma commonly includes Computed Tomography (CT), Magnetic Resonance Imaging (MRI), and fiberoptic laryngoscopy. CT scans show a well-defined hypodense mass without local destruction [6]. On MRI, T1-weighted images show variable intensity, which is enhanced with gadolinium while T2-weighted images of schwannomas present with high intensity [11]. The characteristic finding on laryngoscopy is a submucosal mass commonly seen on the aryepiglottic fold, which may result in an obstructed view of the laryngeal inlet or reduced mobility of the vocal fold [11].

Histology of the lesion is required for definitive diagnosis of a schwannoma. The option of fine needle aspiration (FNA) or biopsy, such as in this case, can be performed. However, reports show FNA is less favorable since often inconclusive. The criteria for histological diagnosis was established by Enzinger and Weiss, which include two areas: Antoni A areas - spindle cells with a myxoid stroma with microcystic/cystic areas, and Antoni B areas - compacted spindle cells with nuclei arranged in a palisading manner. Schwannomas must also stain positive for S-100 protein on immunohistochemistry. Schwannomas are also negative for CD34 [7].

Surgical excision is typically the definitive treatment of laryngeal schwannomas. Various surgical approaches can be performed depending on the size of the mass. These approaches include transoral approach with the use of laser or steel instruments for smaller tumors, or open approaches including lateral or median thyrotomy or lateral pharyngtomy for larger tumors [12,13]. Prognosis is good but recurrence can occur rapidly with incomplete surgical excision. Rapid regrowth after excision has been described but did not occur in this case.

4. Conclusion

Laryngeal schwannomas are rare benign tumors which present with a gradual course. It can cause symptoms such as globus sensation, hoarseness and stridor. On physical exam these tumors can be mistaken for cysts or laryngoceles. Histopathology is required for diagnosis. Surgical excision is the definitive treatment, and should be tailored to the qualities of the tumor and surgeon's comfort.

In this case, a favorable surgical outcome for the removal of laryngeal schwannoma with adequate endolaryngeal exposure, at one year follow up is presented. Specifically, the intraoral approach with the use of a cold steel instrument for excision.

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