

Laryngeal Schwannoma: A Case Report and Literature Review

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Abstract

Case Report

Background: Schwannoma is a benign nerve tumor whose laryngeal location is very rare, diagnosis is relatively easy as it presented as benign tumor. However, recurrence after surgery happens quite often. **Case Presentation:** We report the case of a 50-year-old male patient with chronic dysphonia, nasofibroscope examination showed a pedunculated mass of the left arytenoid suggesting a benign tumor. The patient underwent CO2 laser resection of the tumor under general anaesthesia on direct laryngoscopy. Histopathology examination revealed a schwannoma. Two years later, the patient is symptom-free with no evidence of recurrence on laryngeal endoscopy. **Conclusion:** The purpose of the present case report is to highlight this rare entity and to emphasize technical considerations in order to allow a perfect removal of the disease and to avoid recurrence.

Keywords: Schwannoma – larynx – surgery – CO2 laser.

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BACKGROUND

Forty-five percent of neurogenic tumors develop in the head and neck region [1]. They include solitary schwannoma, neurofibroma, paraganglioma, and granular cell tumors [2]. Schwannomas arise from perineural Schwann cells and the epineurium surrounding the peripheral nerve. Laryngeal schwannomas are very rare; they count for 0.1% to 1.5% of all benign tumors of the larynx. Eighty percent are located in the aryepiglottic fold and twenty percent occur in the false or true vocal cords. The majority of laryngeal schwannomas are submucosal [3]. The surgical approach depends on the tumor volume, whether the tumor is pedunculated or not, and the quality of endoscopic exposure of the tumor. Recurrence after surgery is mainly related to residual disease which is more seen with non-pedunculated tumors [1]. We report a case of laryngeal schwannoma presented as polypoid growth of the vocal process of the arytenoid cartilage and discuss the epidemiology, clinical manifestations, and surgical technique.

CASE REPORT

A 50 years old male patient with no significant history presented to the ENT outpatient clinic with a

one-year history of chronic dysphonia with no other associated symptoms such as dyspnea, choking, or dysphagia. Endoscopic exploration of the upper aerodigestive tract disclosed a submucosal polypoid mass of 08mm rising from the vocal process of the left arytenoid. Vocal folds movement was preserved apart from uncompleted closure of the glottis due to the mass volume, which explains the patient's dysphonia. Additionally, we noticed that supraglottic folds collapse related to vocal forcing. Those features were consistent with a benign laryngeal tumor (Figure 1). Since the mass was of a little volume and was well-circumscribed no further investigations were necessary. For instance, we did not perform a cervical CT scan. Our patient underwent an endoscopic microsurgical approach for the removal of the mass under general anesthesia using laser CO2 (Figure 2). The use of the laser implies several considerations. Settings used for the CO2 laser are selected with keeping tissue cooling in mind; we use super-pulsed modes for this purpose. Depending on the lesion size and location, CO2 laser settings can range from 3 to 10 Watts with a 3 to 5 milliseconds delay to avoid thermal damage to target tissues, as well as surrounding tissues and subsequent fibrosis that could impact mucosal vibration during phonation. The

anaesthesiologist uses a laser-resistant endotracheal tube and maintains oxygen concentration in the anaesthetic gas below 30%. The use of laser implies strict precautions to prevent airway fire which include smoke evacuation during the procedure and placement of saline-soaked cottonoids above the endotracheal tube cuff. Other precautions to avoid patient injury are the placement of saline-soaked surgical towels over the patient's face, as well as saline-soaked gauze eye pads placed to protect the patient's eyes. Operating room personnel should wear laser-safe goggles, as well as the appropriate filters placed on any operating microscope lens to protect the surgeon's eyes, as well. Post operatively, the patient received antibiotics, paracetamol for one week, anti-reflux medication for two months and humidification for two weeks. Also, voice training was recommended to resolve pre-operative supraglottic movement related to vocal forcing. Histopathology examination concluded to a schwannoma of the left arytenoid. (Figure 3) After two years follow-up, there was a clear improvement in voice quality with no evidence of tumor recurrence.

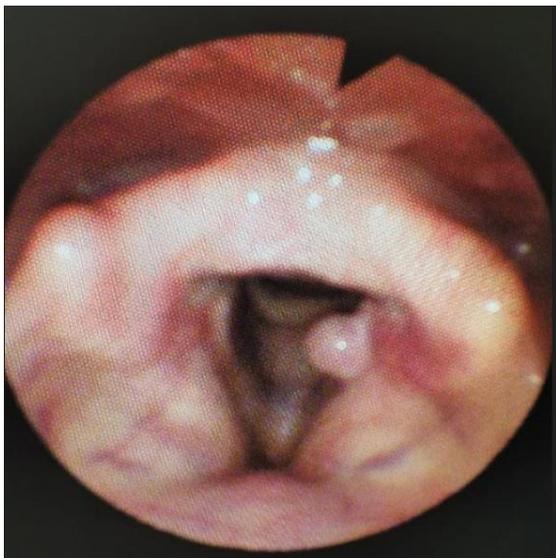


Figure 1: Endoscopic view showing a non-pedunculated mass rising from the vocal process of the left arytenoid



Figure 2: CO2 laser microsurgery settings

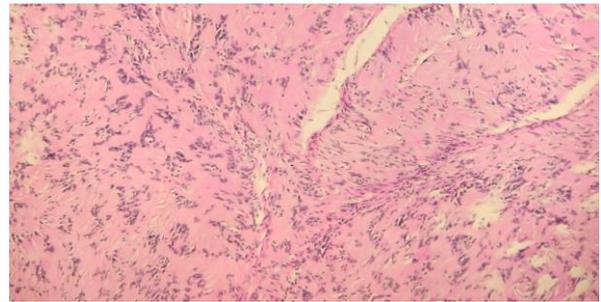


Figure 3: Histopathology examination of the tumor showing tumor proliferation made of spindle cells with nuclear palisades (HESx100).

DISCUSSION

Schwannoma or Neurilemmomas [1], are benign encapsulated tumors originating from neoplastic Schwann cells of the neural sheaths. Laryngeal localization of schwannoma is exceptional. Schwannoma arising from the vagus nerve (X) is mainly located in the plexiform ganglion in the retrostyloid space. Laryngeal schwannoma originates from the superior laryngeal nerve, after its passage through the thyrohyoid membrane, more rarely from the recurrent laryngeal nerve. This explains its frequent localization in the aryepiglottic fold [1].

However, some studies note a slight female predominance [3-6]. Schwannomas are not known to be associated with toxic habits or lifestyle risk factors. The only known risks are radiation exposure and genetic predisposition [1, 7].

The main symptom is dysphonia. It can be alone or associated with other symptoms such as stridor, foreign body sensation especially in pedunculated forms, dysphagia, odynophagia, and/or dyspnea in the case of obstructive tumor [4, 5]. Large tumors can be palpated in the neck, by lateral extension through the thyroepiglottic space [2].

The endoscopic aspect is of a firm submucosal mass, sometimes pedunculated, regular, or bumpy, sometimes mechanical ulcerations can occur. Schwannomas of the larynx are rarely voluminous. Indeed, the smallest tumors cause symptoms and thus lead to the diagnosis [3]. However, a large pedunculated tumor can be life-threatening causing acute respiratory distress due to a valve mechanism [1]. Regarding tumor location, most cases described in the literature are individual cases. Supraglottic localization is the most frequent, especially in the false vocal cord, aryepiglottic fold, and the arytenoid [3, 4, 6, 8], they can also develop in the glottis and subglottis in extremely rare cases [9-11].

Imaging techniques allow the orientation toward a benign tumor and disclose some differential diagnoses such as cysts and laryngoceles, lipomas, cartilaginous and vascular tumors [8]. On CT scan,

most authors describe a well- defined ovoid mass, hypodense compared to the muscle density with a heterogeneous contrast enhancement [2, 4]. On MRI, the lesion is isointense in T1-weighted sequences with a strong heterogeneous enhancement after gadolinium injection. In T2, the lesion is hyperintense [4]. However, even if the tumor does not exhibit malignancy features, squamous cell carcinoma, lymphoma, melanoma, and other malignancies should be kept in the differential diagnosis [1].

The diagnosis can only be made histologically. Direct laryngoscopy with biopsy or incisional biopsy of the lesion is usually performed first for large tumors. However, an incisional biopsy may be difficult due to the solid capsule of the tumor and holds several drawbacks such as false- negative due to insufficient tissue material, scaring of the incisional biopsy site which can make subsequent surgery more difficult, complications such as damaging highly functional structures adjacent to the tumor site. Patients that underwent incisional biopsy were exposed to a higher risk of vocal fold immobility [1]. Furthermore, with incisional biopsy patients are subject to at least 2 surgical procedures. A one-step complete surgical excision of the tumor or excisional biopsy is well-regarded, especially in the case of small, well-limited, and pedunculated tumors [2, 4]. It allows simultaneous diagnosis and treatment of the lesion.

Surgery is the gold standard management tool. Submucosal and extracapsular resection of the schwannoma is the only way to avoid recurrence. The surgical approach depends on the site and the size of the tumor, but more importantly on the fact that the tumor is pedunculated or not. For instance, pedunculated forms can be managed endoscopically regardless of the size of the tumor. Also, no residual disease was observed in this subgroup [1].

Whereas, management of non-pedunculated forms depends on a variety of parameters, such as the tumor size, location, and the endoscopic exposure of the tumor site. An external approach by median or lateral thyrotomy or by lateral pharyngotomy might be used in case of large non-pedunculated tumor or unfavorable endoscopic exposure. A transitory tracheostomy is then realized. The residual disease is present in 17% of the cases in this group [1, 7, 9]. Moreover, open approaches carry more risk of post-operative vocal fold immobility or hypomobility and worse vocal outcome.

Transoral endoscopic microsurgery with or without laser is the best approach for small, glottic, pedunculated tumors with good laryngeal exposure. It allows for better preservation of the laryngeal mucosa, thus vocal outcome is better. Also, it is a less invasive technique with no cervical scaring, no tracheostomy, and less hospitalization duration. CO2 laser offers more precision, less trauma for the mucosa, reduction of the

healing time, and less infection risk and recurrence [9, 12].

Cases of recurrence have been reported in the literature, especially in cases of incomplete excision [4, 5]. Note that schwannoma is radio-resistant [2, 10].

Histological examination of schwannoma is characterized by the triad established by Enger and Weiss [10], including the presence of a capsule, Antoni A and/or Antoni B stroma, and positive staining for S-100. In type A, the Schwann cells are spindle-shaped, with oval nuclei arranged in palisades. In type B, the cells are dispersed in a mucoid stroma and may show cellular atypia. Malignant degeneration of schwannoma is extremely rare, unlike neurofibroma [4, 10].

In their meta-analysis, Tulli *et al.*, observed that follow-up intervals ranged from two to 204 months, and relapsing disease occurred in the first postoperative 3 months. Therefore, they recommended performing fibrolaryngoscopy every 3 months during the first year post-operatively, then annually for at least 2 years. They claim that further investigation with MRI is useful in the case of laryngeal asymmetry [1].

CONCLUSION

Schwannomas of the larynx are extremely rare. They are benign nerve tumors usually located in the supraglottic larynx. The most frequent revealing symptom is dysphonia. Curative treatment consists of complete extracapsular surgical excision. Most intralaryngeal, especially glottic, small size, and pedunculated schwannomas can be resected via transoral approach using or not CO2 laser with the resolution of symptoms in the majority of cases. In case of a long history of dysphonia, the patient may develop vocal forcing behavior. Therefore, dysphonia may not be completely resolved after tumor excision and the patient may require adjuvant voice training.

DECLARATIONS

Ethics Approval and Consent to Participate

Not applicable

Consent for Publication

An informed consent for publication purpose was obtained from the patient

Availability of Data and Material

The datasets generated and/or analyzed during the current study are not publicly available due to patient's data confidentiality but are available from the corresponding author on reasonable request

Competing Interests

Not applicable

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Author's Contributions

No was involved in diagnosis, surgery procedure and manuscript drafting, AT was involved in literature review and drafting of the manuscript, NH was involved in pathology study and reviewed the manuscript, NB was involved in surgery procedure and reviewed the manuscript, MNA reviewed the manuscript for insightful remarks. All authors read and approved the final manuscript.

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