

A Rare Case of Laryngeal Schwannoma – with Review of Literature

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ABSTRACT

Benign tumours constitute 5% or less among all laryngeal tumours. Schwannomas present rarely in the larynx, and constitutes about 0.1 to 1.5% of benign tumours. The majority occurs in supraglottic region with order of preference in the aryepiglottic fold (80%), followed by

the arytenoids, ventricular folds, vocal cord and rarely in subglottis. In our case, it was arising from posterior commissure which is a very rare presentation. The case report illustrates the clinical diagnosis, radiological confirmation up to the surgical treatment and follows-up.

Keywords: Benign tumour, Laryngeal mass, Neurogenic tumour, Supraglottis

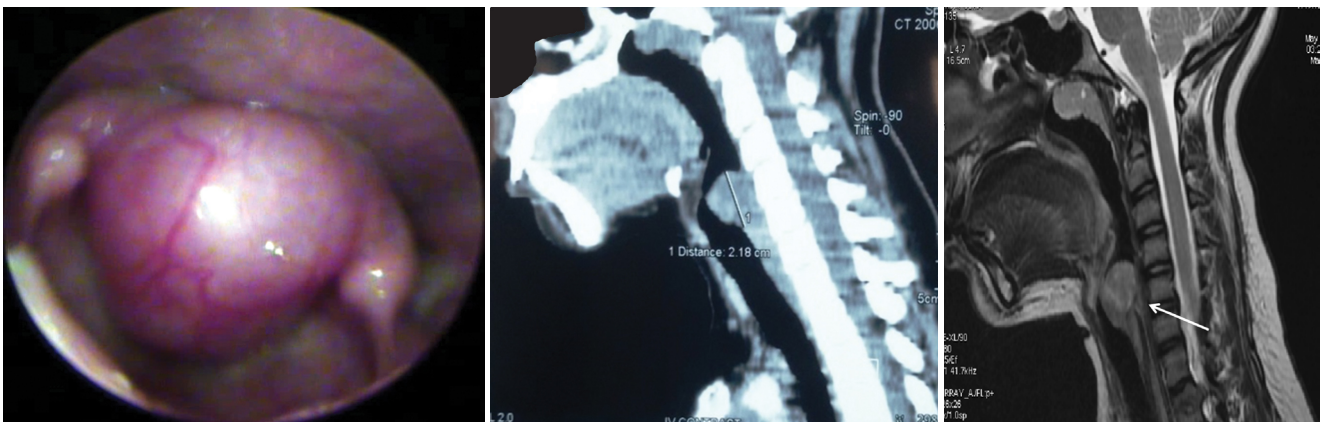
CASE REPORT

A 15-year-old girl presented with dysphonia and dyspnoea on exertion, over a 5-year period, increased effort in speaking and night snoring for one month. There were no associated systemic symptoms. On examination, she was Grade 2 dyspnoeic and not in stridor. General physical examinations were unremarkable.

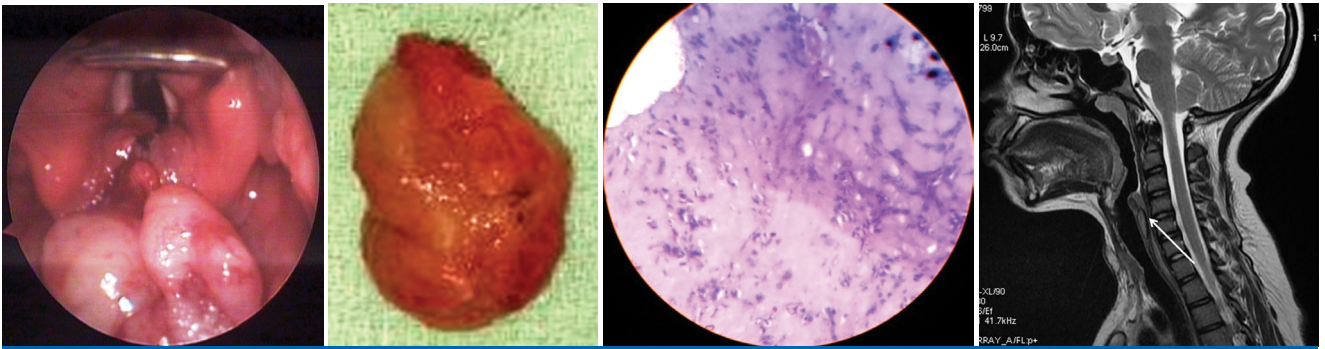
Video laryngoscopy revealed a smooth globular soft tissue swelling with superficial vascularization in the supraglottic region in between arytenoids probably arising from the posterior commissure obscuring the view of the vocal cords [Table/Fig-1].

Patient was advised for CT [Table/Fig-2] and it showed a well circumscribed low attenuation midline supraglottic minimal enhancing mass measuring 2.2 x 2.1 x 1.9 cm in maximum dimensions in between arytenoids with no signs of bone invasion. For further characterisation, MRI of the neck was done and it showed a well defined homogenous T1 hypointense and T2 hyperintense lesion in the supraglottis causing narrowing of the lumen at the level of C4 and C5 vertebral level [Table/Fig-3]. The attenuation of this mass was slightly lesser than that of fluid, suggesting a soft tissue lesion.

Microlaryngeal surgery and excision of mass was planned. A tracheostomy was performed to secure the airway.



[Table/Fig-1]: Globular smooth soft tissue swelling seen in the supraglottic region in between the arytenoids obscuring the view of the vocal cords. **[Table/Fig-2]:** Sagittal reformatted CT image shows soft tissue density mass in the posterior laryngeal wall. **[Table/Fig-3]:** Sagittal T2W Magnetic Resonance (MR) scan shows well defined hyperintense lesion in the supraglottis causing narrowing of the laryngeal lumen at the level of C4 & C5. (images from left to right)



[Table/Fig-4]: Per operative picture showing excision of the tumour. **[Table/Fig-5]:** Gross specimen showing firm yellowish grey encapsulated mass. **[Table/Fig-6]:** Histopathology image shows hypercellular Antoni A palisading spindle shaped cells with formation of verrucay bodies and hypocellular Antoni B areas with collagenised bundle of fibrous tissue and few congested blood vessels. **[Table/Fig-7]:** Post-operative follow-up MRI Sagittal T2 sequence shows no evidence of recurrence or residual lesion. (images from left to right)

Kleinsasser suspension laryngoscope introduced and mass visualised. Aspiration was tried but no aspirate was obtained. After infiltration, submucosal dissection was done along the capsule of the solid tumour, since further manipulation of such large tumour was difficult with suspension laryngoscope, we switched over to Macintosh laryngoscope. Tumour was excised in toto [Table/Fig-4], vocal cords were visualised and found to be normal. Extreme care was taken to preserve as much normal laryngeal mucosa as possible in order to prevent future cicatrization or scarring and for a favourable clinical outcome of voice in our patient. Subglottis was visualised and no mass or swelling found. The gross specimen [Table/Fig-5] was firm in consistency and yellowish grey in color with capsule.

Histopathological examination [Table/Fig-6] was showing biphasic pattern of Antoni A palisading spindle shaped cells with Verrocay bodies formation and Antoni B areas with collagenised bundle of fibrous tissue confirming the diagnosis of a laryngeal schwannoma.

Patient was followed-up two weeks later and had good voice preservation after successful decannulation. Post operative MRI [Table/Fig-7] showed no evidence of mass lesion. Patient consent was obtained for scientific publication.

DISCUSSION

Schwannomas are benign, encapsulated, slow-growing, submucosal tumours that arise from the Schwann cells of peripheral, cranial, or autonomic nerves [1]. About 25 to 45% of all schwannomas appear in the head and neck region, with the majority in the parapharyngeal spaces. Schwannomas being one of the rare benign laryngeal tumours, presenting commonly in the 4th and 5th decade of life [2], especially in women. Majority occurs in submucosa of supraglottic area. Commonly encountered sites are the aryepiglottic folds. Other rare locations are arytenoids, ventricular folds, vocal cords and subglottis. In our case, the lesion was arising from the posterior commissure. The schwannoma is said to be

arising from the distal nerve fibres of the internal branch of the superior laryngeal nerve [3].

Patients characteristically present with dysphonia, dysphagia and globus sensation. As the tumour expands, it may cause dyspnoea and stridor. Our patient presented with dysphonia and dyspnoea. The diagnostic work-up includes laryngoscopy, imaging studies, and histological biopsy. Laryngoscope shows a globular submucosal lesion. CT and MRI are useful in defining the nature and extent of the lesion, with MRI offering superior soft tissue delineation and best imaging tool for suggesting the diagnosis in the appropriate clinical context. On CT scans, schwannomas appear as well-defined, hypodense masses [4], without adjacent tissue invasion. On MRI schwannomas appear as well defined isointense to slightly hypointense lesion in T1-weighted sequences, hyperintense in T2-weighted sequences and shows enhancement with contrast.

Laryngeal ultrasonography as a diagnostic method is rarely used, provides additional information on the vascular pattern, lesion texture and the absence of invasion of the thyroid cartilage, the posterior limits of the lesion may not be visualized [1].

The differential diagnosis includes laryngeal cyst, internal laryngoceles, adenomas, mucoceles, chondromas, lipomas and malignant tumours [1].

Histology remains the gold standard. Following are the Enger and Weiss histological criteria: 1. The presence of a stromal Antoni A (compacted, nucleated bipolar cells in a palisade form) and/or Antoni B (loosely arranged spindle cells within a myxoid matrix) histological pattern. 2. The presence of a capsule. 3. Stain positive for S-100 [5].

It is important to differentiate a schwannoma from a neurofibroma because, neurofibromas have a higher chance of recurrence and malignant transformation of approximately 10%. The risk of malignant transformation of laryngeal schwannoma is very rare [6], Surgical removal of a tumour in toto is possible in schwannomas as it originates from the nerve

sheath, and impossible in neurofibromas. Schwannomas originating from perineural Schwann cells grow extrinsically to the nerve fascicles and may expand along any somatic or sympathetic nerve in the body. In case of neurofibromas they originate from perineural fibrocytes and involves nerve fibers and neural sheath cells [7].

The definitive treatment is surgery and the approach depends on the location and size of the lesion. Endoscopic approach is preferred in smaller lesions with or without a laser. In our case even though the lesion is about 2 cm in size, we approached endoscopically and removed in toto. Larger tumours are treated by lateral pharyngotomy laryngofissure or lateral thyrotomy [8]. Transoral robotic surgery-aided excision of schwannomas is recently reported [9]. Protection of laryngeal mucosa and preservation of laryngeal function warrants better surgical outcome [10,11]

CONCLUSION

Even though neurogenic tumours of larynx are rare, it should be recognised, as the surgical treatment gives cure. The primary management involves securing the airway. To prevent recurrence, complete surgical resection is the treatment of choice.

REFERENCES

- [1] Souza LR, De Nicola H, Yamasaki R, Pedrosa JE, Brasil OD, Yamashita H. Laryngeal schwannoma: a case report with emphasis on sonographic findings. *Radiologia Brasileira*. 2014;47(3):191-93.
- [2] Ramakrishnan Y, Issing WJ. Laryngeal schwannoma: case report and literature review. *ISRN Otolaryngology*. Volume 2011 (2011), Article ID 540643, 3 pages.
- [3] Nanson EM. Neurilemoma of the larynx: a case study. *Head & neck surgery*. 1978;1(1):69-74.
- [4] Plantet MM, Hagay C, De Maulmont C, Mahé E, Banal A, Gentile A, et al. Laryngeal schwannomas. *European Journal of Radiology*. 1995;21(1):61-66.
- [5] Brandner S. The genetics and molecular biology of neural tumours. *Br J Cancer*. 2010;102(4):784-85.
- [6] De Lozier HL. Intrinsic malignant schwannoma of the larynx a case report. *Annals of Otolaryngology, Rhinology & Laryngology*. 1982;91(3):336-38.
- [7] Rosen FS, Pou AM, Quinn FB. Obstructive supraglottic schwannoma: a case report and review of the literature. *The Laryngoscope*. 2002;112(6):997-1002.
- [8] Takumida M, Taira T, Suzuki M, Yajin K, Harada Y. Neurilemmoma of the larynx: (A case report). *The Journal of Laryngology & Otolaryngology*. 1986;100(07):847-50.
- [9] Kayhan FT, Kaya KH, Ylmazbayhan ED. Transoral robotic approach for schwannoma of the larynx. *Journal of Craniofacial Surgery*. 2011;22(3):1000-02.
- [10] Ingels K, Vermeersch H, Verhoye C, De Potter C. Schwannoma of the larynx: a case report. *The Journal of Laryngology & Otolaryngology*. 1996;110(03):294-96.
- [11] Sanghvi V, Lala M, Borges A, Rodrigues G, Pathak KA, Parikh D. Lateral thyrotomy for neurilemmoma of the larynx. *The Journal of Laryngology & Otolaryngology*. 1999;113(04):346-48.

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FINANCIAL OR OTHER COMPETING INTERESTS:

None.

Date of Publishing: **Apr 01, 2017**