



VOCAL CORD SCHWANNOMA-A RARE CASE PRESENTATION



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Abstract

Objectives: Neurogenic tumors of the vocal cord are extremely rare. The goal of this report is to advert to this rare disease, to review and discuss diagnostics, differential diagnoses and treatment options. **Study Design:** Retrospective case report and review of the literature. **Methods:** Case report of a schwannoma of the vocal cord and review of the literature regarding neurogenic tumors of the larynx. **Results:** Neurogenic laryngeal tumors typically involve the supraglottic larynx, rarely the glottis. They can cause globus sensation, dysphagia, dysphonia and upper airway obstruction. Imaging does not yield a definite diagnosis. The only curative treatment option is complete surgical resection. **Conclusions:** A definite diagnosis can only be made histologically. Endoscopic resection for smaller lesions and external approaches for larger lesions are recommended treatment options.

INTRODUCTION

About 45% of all neurogenic tumors occur in the head and neck region and are mostly located in the parapharyngeal space^{1,2}.

Two types of neurogenic tumors must be distinguished: Schwannomas and neurofibromas. Schwannomas emanate from perineural Schwann cells, and are well encapsulated, growing adjacent to the parental nerve but extrinsic to the nerve fascicles². Neurofibromas on the other hand derive from perineural fibrocytes, and are not encapsulated and are usually intertwined with the parental nerve fascicles^{2,3}.

Multiple neurofibromas are observed in Neurofibromatosis. The location of Schwannoma or neurofibroma within the larynx is very uncommon. They represent 0.1% to 1.5% of all benign laryngeal tumors, schwannoma being slightly more frequent than neurofibroma⁴. 80% are located in the aryepiglottic fold, 20% in the false or true vocal cords^{5, 6, 7}. They usually grow submucosal; with a few reports describing polypoid growth⁶. There seems to be a slight female preponderance^{2, 6}. The

internal branch of the superior laryngeal nerve is most likely the nerve of origin^{8,9}

CASE REPORT

A 54-year-old man was referred by ENT physician to Department of Pathology at Katuri Medical College, Guntur for clinical diagnosis and treatment of a mass discernable endoscopically under the intact mucosa of his right vocal cord. He had a 2- to 3-year history of hoarseness and dyspnoea on exertion with no complaint of dysphagia. He had a 15 pack-year smoking history. Further medical history was unremarkable. Physical examination was normal.

Fiberoptic laryngoscopy revealed a submucosal mass within the right vocal cord obstructing the view of the hypo-mobile vocal cord. The mobility of the left vocal cord was normal.

Computed tomography (CT) demonstrated a 12 × 13 × 8 mm well defined, round to oval, mass in the right vocal cord growing under intact mucosa. Compared to muscle it was hypodense, slightly inhomogeneous with a clear capsule and no sign of infiltrative growth or cartilaginous

destruction. The tumor was resected as far as possible through the incision in the right vocal cord and sent for histopathological evaluation. Patient had an uneventful postoperative recovery.

Histopathology showed densely cellular areas, composed of compact elongated cells with palisading (Antoni A pattern) and a less cellular, loosely texture pattern in which cells often contained lipid (Antoni B pattern). Immunohistochemical studies showed reactivity for S100 protein. These features were consistent with schwannoma of the vocal cord (Figure 1 and 2).

DISCUSSION

Two different types of neurogenic tumors of the larynx have been described: Schwannomas and neurofibromas. Both entities are rare and comprise only about 0.1% to 1.5% of all benign laryngeal tumors⁴. Neurofibromas are encountered more frequently in Neurofibromatosis. Malignant transformation is reported in 10% of neurofibromas while in schwannoma it is very uncommon². Neurogenic tumors of the larynx are most frequently located in the aryepiglottic fold or in the true or false vocal cords^{5,6,7}.

Vocal cord schwannoma is a rare benign tumor arising from the internal branch of the superior laryngeal nerve⁸. Schwannomas affect patients in all age groups, more commonly occurring in sixth and seventh decades¹⁰. It is more common in females¹¹.

The clinical symptoms of the disease are those usually associated with a slow growing lesion of the larynx: Over a period of years the patient gradually develops hoarseness, globus sensation, dysphagia, dyspnoea on exertion with inspiratory, sometimes biphasic stridor^{2, 6}. Some patients complain about dyspnoea in the supine position which seems to be associated with the location of the lesion^{6, 12}. One case of asphyxial death due to laryngeal schwannoma is reported.

The true vocal cord on the affected side is usually immobile or hypomobile^{6, 13, 14}, although some authors report normal mobility^{9,15}. In many cases, such as in our reported case, the bulgy supraglottic tumor obstructs the view of the true vocal cord^{11, 16}. Also, the mass effect of a large tumor can mimic fixation of the cricoarytenoid joint ("pseudo-fixation")⁶.

The diagnostic workup should include indirect and fiberoptic laryngoscopy which usually reveals a submucosal mass in the described location. Such a lesion coupled with impaired vocal cord mobility should draw attention towards a neurogenic tumor. In CT scans of the disease, most authors describe a well defined, hypodense submucosal mass without signs of infiltrative or destructive growth¹¹

A definite diagnosis can only be made histologically. Schwannomas almost exclusively are comprised of spindle cells with long, oval nuclei and indistinct cell membranes. These Schwann cells either form cellular regions with compact cell bundles with nuclei lining up in palisades (Antoni A regions) or edematous regions with loosely arranged cells in a myxoid matrix prone to degeneration (Antoni B regions). Two compact rows of well aligned nuclei separated by fibrillary cell processes are called Verocay bodies. Axons are usually not found in Schwannomas. A clear capsule, the presence of Antoni A and/or Antoni B areas, and intense immunoreactivity for S-100 protein are criteria for the histologic diagnosis of Schwannoma^{2,6}

The main differential diagnosis of a laryngeal schwannoma is neurofibroma. Other differentials include other benign tumors like laryngeal cyst, laryngocele, adenoma, and chondroma. Schwannomas must be distinguished from neurofibromas because recurrence and malignant transformation is relatively more in neurofibromas^{17,18}.

CONCLUSION

Schwannoma is a solitary, encapsulated, slow-growing benign tumor and commonly occurs in women. Neurofibromas may be single or multiple. Neurofibromas are intertwined within the nerve fascicles, in contrast to schwannoma which is encapsulated¹⁸.

Surgical removal is the treatment of choice. Small tumors can be removed by endoscopic excision and large tumors by an external surgical approach. Histopathology and immunohistochemistry are required for confirmation of diagnosis. Recurrence or malignant sarcomatous changes following surgical treatment are rare^{1,2}.

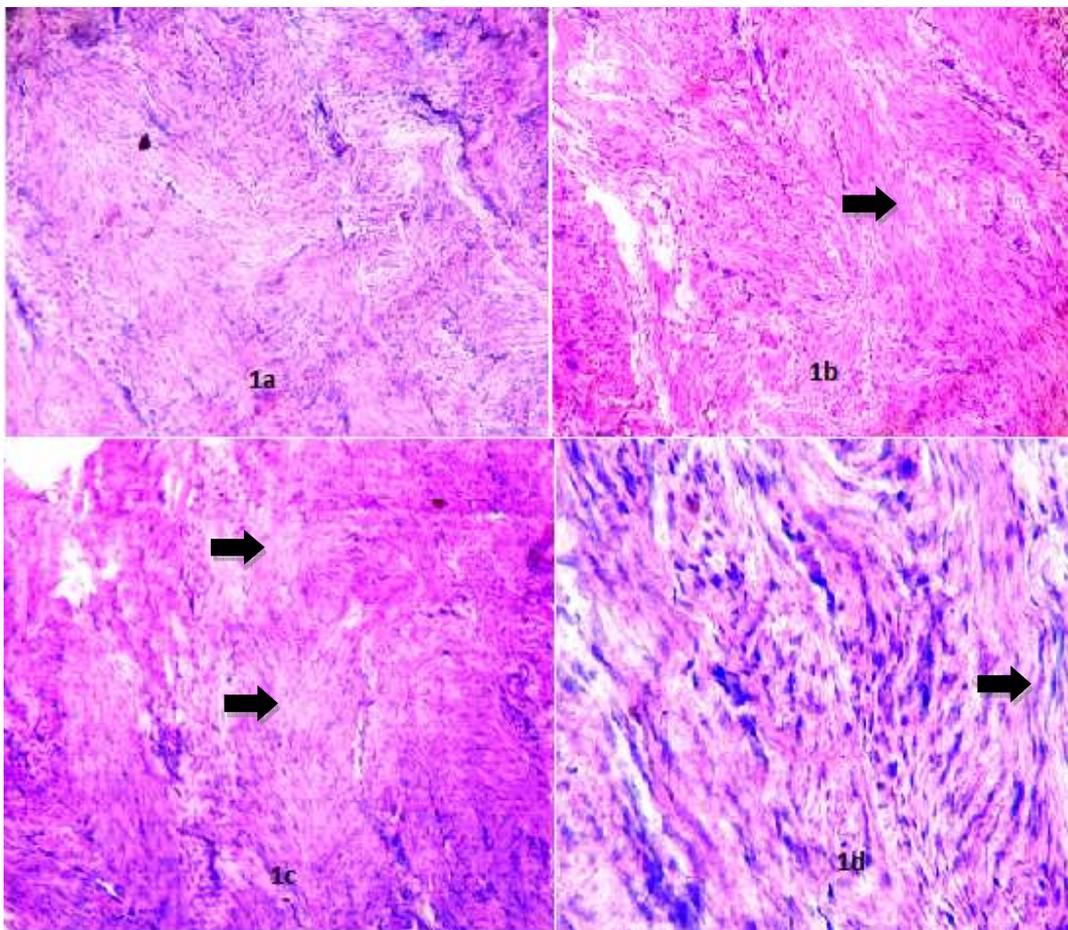


Figure 1 Schwannoma: 1a-Showing Antoni A and Antoni B areas (H&E, 100x),
1b-Showing areas of hyalinization (H&E, 100x),
1c-Showing Verocay bodies (H&E, 100x),
1d- Showing Schwann cells (H&E, 400x).

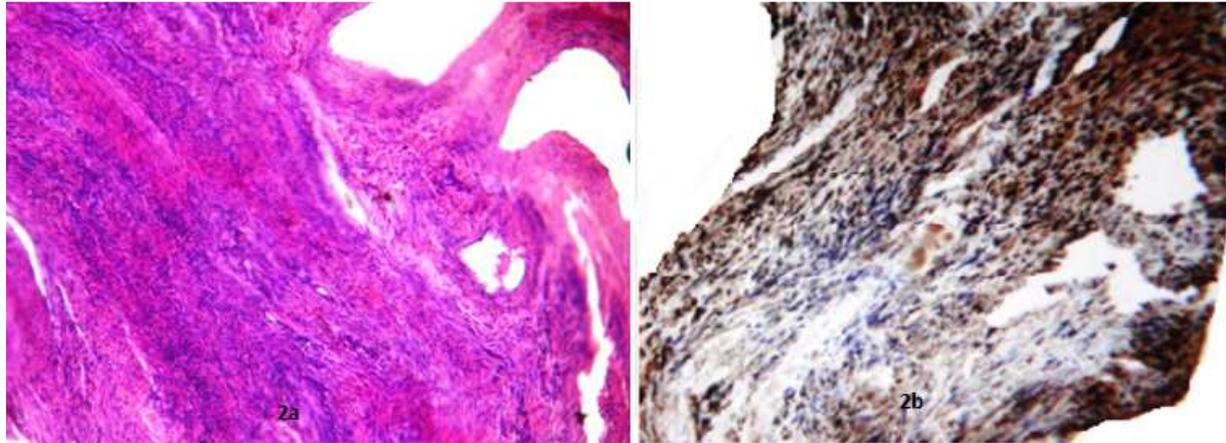


Figure 2 2a- Showing a Schwannoma with cystic change (H&E, 100x), 2b-Showing S-100 positivity in Schwannoma (IHC S-100,100x)

REFERENCES

1. Sanghvi V, Lala M, Borges A, Rodrigues G, Pathak KA and Parikh D: Lateral thyrotomy for neurilemmoma of the larynx. *J Laryngol Otol* 1999, 113(4): 346-8.
2. Zbaren P and Markwalder R: Schwannoma of the true vocal cord. *Otolaryngol Head Neck Surg* 1999, 121(6):837-9.
3. Lusk MD, Kline DG and Garcia CA: Tumors of the brachial plexus. *Neurosurgery* 1987, 21(4):439-53.
4. Jones SR, Myers EN and Barnes L: Benign neoplasms of the larynx. *Otolaryngol Clin North Am* 1984, 17(1):151-78.
5. Palva T, Jokinen K and Karja J: Neurilemmoma (schwannoma) of the larynx. *J Laryngol Otol* 1975, 89(2):203-7.
6. Rosen FS, Pou AM and Quinn FB Jr: Obstructive supraglottic schwannoma: a case report and review of the literature. *Laryngoscope* 2002, 112(6):997-1002.
7. Fini-Storchi I and Frosini P: Laryngeal neurinoma. A case report and review. *ORL J Otorhinolaryngol Relat Spec* 1997, 59(3):1825.
8. Nanson EM: Neurilemoma of the larynx: a case study. *Head Neck Surg* 1978, 1(1):69-74.

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9. Cadoni G, Bucci G, Corina L, Scarano E and Almadori G: Schwannoma of the larynx presenting with difficulty swallowing. *Otolaryngol Head Neck Surg* 2000, 122(5):773-4.
 10. Woodruff JM, Kourea HP, Louis DN and Scheithauer BW: Schwannoma. In: Kleihues P, Cavenee WK, editors. *WHO Classification of Tumours: Pathology and Genetics of Tumours of Nervous System*. 2nd ed. Lyon, France: IARC Press; 2000. p. 164-6
 11. Jamal MN: Schwannoma of the larynx: Case report, and review of the literature. *J Laryngol Otol* 1994; 108: 788-90.
 12. Gardner PM, Jentzen JM, Komorowski RA and Harb JM: Asphyxial death caused by a laryngeal schwannoma: a case report. *JLaryngol Otol* 1997, 111(12):1171-3.
 13. Meric F, Arslan A, Cüreoglu S and Nazaroglu H: Schwannoma of the larynx: case report. *Eur Arch Otorhinolaryngol* 2000, 257(10):555-7.
 14. Ho WK, Lo SD and Ng RW: Schwannoma of the larynx. *Ear Nose Throat J* 2005, 84(9): 554-5.
 15. Taylor J, Stiefel M and Park SY: Schwannoma of the true vocal fold: a rare diagnosis. *Ear Nose Throat J* 2006, 85(1):52-3.
 16. Baumann I, Paasch S and Maassen MM: [Progressive oropharyngeal dysphagia. Neurinoma of the larynx]. *Hno* 2002, 50(1):62-3.
 17. Al-Otieschan AT, Mahasin ZZ, Gangopadhyay K, al-Dayel and Jamshed A: Schwannoma of the larynx: Two case reports and review of the literature. *J Otolaryngol* 1996; 25: 412-5.
 18. Enzinger FM and Weiss SW: Benign tumors of the peripheral nerves. In: Enzinger FM, Weiss SW, editors. *Soft tissue tumors*. St Louis: Mosby; 1988. p. 725-35.