



Laryngeal schwannoma - A rarely occurring benign tumor.

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Abstract:

Neurogenic tumors of the larynx are quite rare. We present the case report of a laryngeal schwannoma in a 28 year old male. The goal of this report is to make otolaryngologist aware about rare site of involvement and unusual presentation. A definite diagnosis can only be made histologically and only curative treatment is complete surgical resection.

Introduction:

Schwannomas are benign, slow-growing, encapsulated tumors that arise from the Schwann cells of the nerves. These tumors are usually seen in the 4th and 5th decade of life and are more common in females.

Twenty-five percent to 45% of all schwannomas present in the head and neck with the majority occurring in the parapharyngeal space¹. The larynx remains a rare site. Over 130 cases of laryngeal schwannoma have been reported in the literature since it was first described in 1925 by Suchanck¹. Laryngeal schwannomas are slow growing lesions that usually arise from the false vocal fold or aryepiglottic fold. The most common anatomical site of laryngeal schwannoma is the aryepiglottic fold (80%), followed by the arytenoids, ventricular folds, and vocal fold (20%)². We describe a case of laryngeal schwannoma, which occurred in a young male and was diagnosed only after repeated biopsies because of submucosal nature of the lesion.

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Case Report:

A 28 yr old male presented to the otolaryngology clinic with the complaints of change in the voice for 4-5 months and dull pain on left side of the neck for past 1 month. There was no history of dysphagia and respiratory difficulty.

Patient was a nonsmoker and a nonalcoholic. On examination of the neck, no physical abnormality was seen. On fiberoptic laryngoscopy, a circumscribed smooth mass was seen filling the endolarynx, obscuring the view of the true and false vocal folds. (Fig-1A). Contrast enhanced computed tomography scan (CECT) of the neck showed a heterogeneously enhancing well defined mass filling the supraglottic region of larynx. (Fig-1B). In view of impending respiratory difficulty and difficult preoperative endotracheal intubation score elective tracheostomy was done. A day later patient underwent direct laryngoscopic assessment under general anesthesia. His endolarynx including left pyriform sinus was obscured by the tumor while right pyriform sinus was free of tumor.

In view of the submucosal location and well encapsulated lesion, repeated biopsies on separate occasions were required for making histopathological diagnosis. Finally histopathological examination confirmed the nature of the tumor to be a schwannoma. As the only curative treatment option is complete surgical resection, considering the size of lesion an excision via external approach was planned. A 4x4 cm tumor was removed en bloc by transcervical approach under general anesthesia. (Fig-2A and 2B).

Postoperatively patient stayed well and fiberoptic laryngoscopy done on fourteenth day showed normal mobility of true vocal folds with adequate glottic space (Fig-3). Decannulation was done successfully thereafter.

Discussion:

Benign neurogenic tumors of the larynx consist of the schwannoma and neurofibroma. These tumors are rare and represent 0.1% to 1.5% of all benign laryngeal tumors, with schwannomas being more frequent than neurofibromas³. Origin of the laryngeal schwannoma is presumed to be in the internal branch of the superior laryngeal nerve after it penetrates the thyrohyoid membrane⁴. Schwannoma should be differentiated from neurofibroma due to some reasons:

1) Schwannomas arise from Schwann cells while neurofibromas from perineural fibrocytes thus while schwannoma grows extrinsic to the nerve fiber, in neurofibroma the tumor is within the parental nerve fascicles². This makes preservation of nerve fiber possible during excision of schwannoma. 2) Neurofibroma is associated with greater recurrence rate. 3) Neurofibroma has potential of malignant transformation to neurogenic sarcoma in 10% of the cases whereas malignant degeneration of schwannomas is extremely rare¹.

4) Schwannoma usually occurs as a solitary lesion and is always encapsulated; Pain suggests degenerative changes or hemorrhage in a schwannoma. Neurofibroma are not encapsulated, commonly multiple and are not associated with pain. These are encountered more frequently in Neurofibromatosis, but there is a case report of histologically diagnosed schwannoma in a patient of neurofibromatosis⁵. Thus clinical examination for presence of café au lait spots should be done in all patients. In our patient General physical examination was negative for café-au-lait spots, axillary or inguinal freckling, superficial neurofibromas, or Lisch nodules, as might be expected in neurofibromatosis type II. Beside this an association of benign solitary schwannomas with skin and breast cancer) has also been reported⁶. Other differential diagnosis are condroma, adenoma, laryngeal cyst, internal laryngocele^{2,7}.

A schwannoma on fiberoptic laryngoscopy appears as a smooth submucosal lesion, with a few reports describing polypoidal growth². It may obstruct the view of the laryngeal inlet and result in reduced mobility of the vocal folds. "Pseudofixation" of the cricoarytenoid joint as a result of mass effect of the lesion may be the cause of this reduced mobility². Though Schwannoma and neurofibromas cannot be distinguished radiologically, computed tomography (CT) and magnetic resonance imaging (MRI) are valuable in defining the nature and extent of the lesion. CT scan is preferable to MRI, as it shows the heterogeneous enhancement following contrast administration⁸.

Calcification though rare reflects degenerative change in a long standing schwannoma. Since the diagnosis can only be made histologically, direct laryngoscopy with biopsy of the lesion is usually the first step in treatment. However, in Schwannoma biopsy can be difficult due to the solid capsule of the tumor².

Enger and Weiss established three histological criteria for the diagnosis of schwannoma: encapsulation, presence of Antoni A and/or Antoni B stroma, and S-100 protein positivity⁹. As these tumors are radioresistant complete surgical excision is the only treatment.

For smaller tumors, endoscopic resection with use of the CO₂ laser is done while external approach is required for larger lesions. Lateral pharyngotomy, lateral thyrotomy, and median thyrotomy (laryngofissure) are the possible external approaches. A tracheostomy may be required to secure the airway as in our case. The key to successful treatment is complete excision of the tumor, preservation of laryngeal function, and mucosal coverage of exposed cartilage. Independent of the approach, vocal cord mobility is usually restored even if it was immobile prior to surgery⁵.

Conclusion:

Despite of its rarity, early diagnosis and treatment is vital. As the Imaging defines only the extent of the lesion, diagnosis can only be made histologically. Since the tumor has a solid capsule, repeated biopsy is often required.

Surgical excision is the only curative treatment option and complete resection of the lesion is necessary to prevent recurrence.

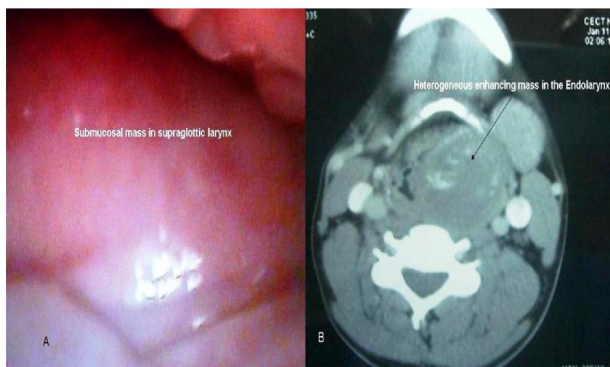


Fig-1(A) Fiberoptic laryngoscopy showing a circumscribed smooth mass filling the endolarynx.

Fig-1(B) Contrast enhanced computed tomography scan (CECT) of the neck showing a heterogeneously enhancing well defined mass filling the supraglottic region of larynx.

Figure 1

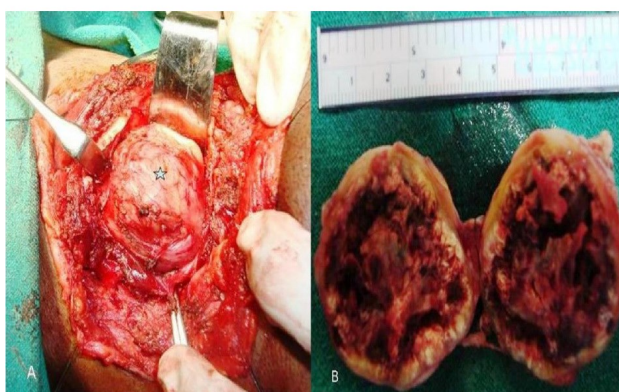


Fig-2(A) Perioperative image showing removal of tumor(*) by transcervical approach.

Fig-2(B) Cut specimen of the tumor showing areas of degeneration.

Figure 2

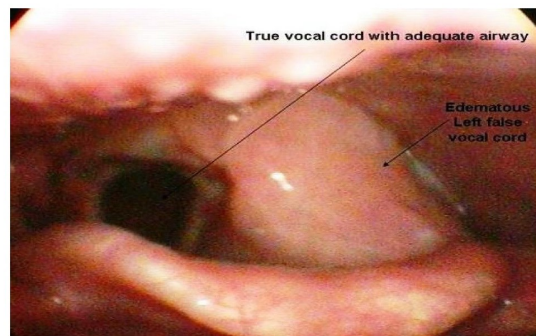


Fig-3 Fiberoptic laryngoscopy showing normal mobility of true vocal cords with adequate glottic space.

Fig 3

References:

1. Zbaren P, Markwalder R. Schwannoma of the true vocal cord. *Otolaryngol Head Neck Surg.* 1999; 121:837–9.
2. Rosen FS, Pou AM, Quinn FB. Obstructive supraglottic schwannoma: a case report and review of the literature. *Laryngoscope.* 2002; 112:997–1002.
3. S. R. Jones, E. N. Myers, and L. Barnes. Benign neoplasms of the larynx. *Otolaryngologic Clinics of North America*, vol. 17, no. 1, pp. 151–178, 1984. :

4. E. M. Nanson, "Neurilemoma of the larynx: a case study," *Head and Neck Surgery*, vol. 1, no. 1, pp. 69–74, 1978
5. Plantet MM, Hagay C, De Maulmont C, Mahe E, Banal A, Gentile A, Cherel P, Mayras C. Laryngeal schwannomas. *Eur J Radiol*. 1995; 21:61–6.
6. Das Gupta TK, Brasfield RD, Strong EW, Hajdu SI. Benign solitary Schwannomas (neurilemomas) *Cancer*. 1969; 24:355–66.
7. Malcolm PN, Saks AM, Howlett DC, Ayers AB. Case report: magnetic resonance imaging (MRI) appearances of benign schwannoma of the larynx. *Clin Radiol*. 1997; 52:75–6.
8. Cadoni G, Bucci G, Corina L, Scarano E, Almadori G. Schwannoma of the larynx presenting with difficulty swallowing. *Otolaryngol Head Neck Surg* 2000;122:773–774.
9. J. M. Woodruff, H. P. Kourea, D. N. Louis, and B. W. Schethauer, "Schwannoma," in *WHO Classification of Tumours: Pathology and Genetics of Tumours of Nervous System*, P. Kleihues and W. K. Cavenee, Eds., pp. 164–166, IARC Press, Lyon, France, 2nd edition, 2000.