



LARYNGEAL SCHWANNOMA - A RARE CLINICAL ENTITY

MIRIA MATHEWS

Department of ENT, CHRISTIAN MEDICAL COLLEGE

Abstract :

Laryngeal schwannomas are rare tumors of the larynx. They can present with subtle complaints and nonspecific features on clinical evaluation. The diagnosis is made on histopathological examination. The definitive management of this condition is complete surgical excision. We present a case of laryngeal schwannoma in a 24 yr old male to emphasize on this rare clinical entity its presentation, clinical features, diagnosis and surgical management.

Keyword :

Schwannoma, Neurofibroma

INTRODUCTION

Neurogenic tumors of the larynx are extremely rare. Around 40% of all the neurogenic tumors are seen in the head and neck region with the predominant area involved being the parapharyngeal space(1). There are two types of laryngeal neurogenic tumors described in literature. They are neurofibromas and schwannomas. They differ in the cells of origin. Neurofibromas arise from the perineural fibrocytes and schwannomas from the perineural Schwann cells. The most common site of involvement is supraglottis. They are benign, slow growing tumors which are seen on clinical examination as a submucosal mass in the supraglottis. Schwannomas are seen more commonly than neurofibromas. This condition is seen more commonly in females.

CASE REPORT

A 24 year old male presented to our Out Patient Department with complaints of difficulty in swallowing for the past 10 months. It was associated with progressive hoarseness of voice for the past 4 months. There was no history of any breathing difficulty. On fibre optic endoscopy, a smooth submucosal mass was seen arising from the interarytenoid region. Bilateral vocal cords were mobile.

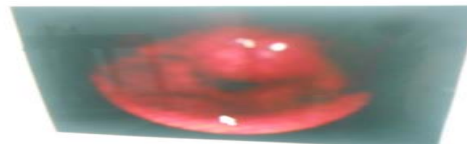


Figure 1: Laryngoscopic view of the submucosal growth over the interarytenoid region. Computed tomography (CT) demonstrated a 22 × 23 × 28 mm well defined, round to oval, mass arising posterior pharyngeal wall growing under intact mucosa. Compared to muscle it was hypodense, slightly inhomogeneous with a clear capsule and no sign of infiltrative growth or cartilaginous destruction (Figure 2).

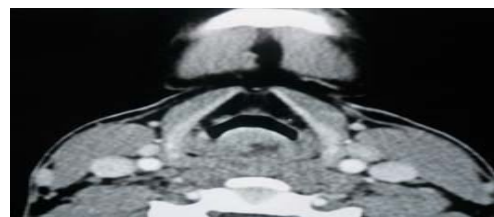


Figure 2A: CT image axial section showing well encapsulated isodense mass arising from posterior pharyngeal wall

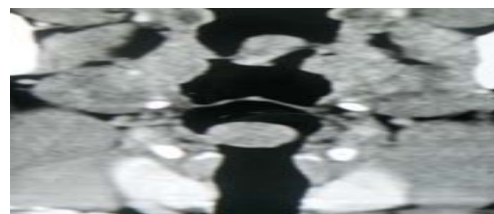


Figure 2B: CT image coronal section showing mass arising from posterior pharyngeal wall



Figure 2C: CT image sagittal cut showing mass arising from the interarytenoid region. Suspension microlaryngoscopy was performed under general anaesthesia, with trans-oral laser assisted excision biopsy. The findings were consistent with the fibre optic examination. The tumor was excised in toto. The patient was extubated primarily and recovered well. The histopathological evaluation confirmed the diagnosis of laryngeal schwannoma (Figure 3).

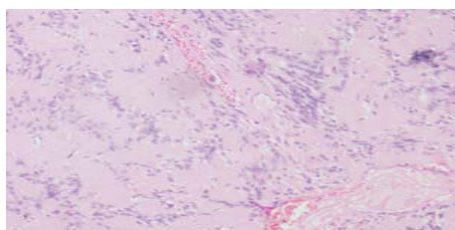


Figure 3A: Histologic appearance of Antoni A regions. Haematoxylin- Eosin, Magnification x100

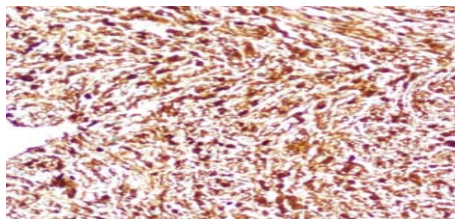


Figure 3B: S-100 positivity of tumor tissue

The patient was followed up for a year post operatively with fibre optic endoscopic examination of the larynx and was found to be free of disease.

DISCUSSION

Neurogenic tumors comprise just 0.5% to 1% of all benign tumors of the larynx(2). Laryngeal schwannoma was first described by Suchanck in 1925(3). Presently there are around 150 cases of such cases described in literature(3). Schwannomas can develop from any nerve in the body except olfactory and optic nerve which lack Schwann cells(4). They are well encapsulated and thus can be removed completely. However in neurofibromas which arise from the perineural fibrocytes, the tumor is non-encapsulated and is seen intertwined with neural fibres which makes it impossible for complete excision(5). Neuromas commonly arise from the internal branch of the superior laryngeal nerve(2). Malignant transformation is seen in 10% of neurofibromas but very rarely seen in schwannoma(3). Clinically, patients can present with varying symptoms owing to the mass effect caused by this slow growing tumor. These may be complaints of change in voice or globus sensation in the throat. As the tumor grows, there can be difficulty in swallowing and noisy breathing or difficulty in breathing. They can also extralaryngeally to present as a mass in the lateral aspect of neck. There is also a case report of asphyxia and death due to a large laryngeal schwannoma (6). Indirect laryngoscopy and fibre optic endoscopy of the larynx is mandatory in evaluating this condition. It is usually seen as a submucosal mass. 80% of these tumors arise from the aryepiglottic

folds(2). The true cord on the affected side can have impaired mobility or can even be fixed due to mass effect of the tumor(7). In some cases, the large size of the tumor can obscure the view of true cords(8). CT imaging reveals a well-defined, mucosal mass hypodense mass which shows no signs of infiltration into surrounding tissues or destruction of the surrounding bony-cartilaginous framework (9). However in T1 weighted contrast study, the mass is isointense with inhomogeneous enhancement with gadolinium and T2, hyperintense(10). The diagnosis is made based on histology. On histopathological examination, characteristic Antoni A and B areas are seen. Antoni A areas are regions of compact cell bundles with nuclei arranged in palisades. Two compact well aligned rows of cells separated by fibrillary cell process are called Verocay bodies. Antoni B areas comprises of loosely arranged cells in a myxoid matrix. Enger and Weiss put forth criteria for diagnosing schwannoma on histology. It includes a clear capsule, presence of Antoni A and/or Antoni B areas and immunoreactivity to S-100 which is diagnostic for schwannoma(7).

The definitive treatment modality is complete surgical excision. Since the diagnosis is made based on histology, initially direct laryngoscopy with biopsy of the lesion is done to confirm diagnosis. For larger lesion with airway compromise, a temporary tracheostomy may be required to secure the airway. External approaches such as median or lateral thyrotomy or median or lateral pharyngotomy approaches may be required for complete removal of larger lesions(8). However transoral laser assisted excision is the treatment of choice for smaller tumors(11). Though rare, recurrence after surgical excision has been reported in few cases of laryngeal schwannoma(7). The differential diagnosis for schwannoma is a neurofibroma of the larynx. In cases of laryngeal neurofibromatosis, clinical evaluation for familial neurofibromatosis should be done such as café a le spots on physical examination and MRI imaging to rule out neuromas of cranial nerves and spinal nerves. In cases of solitary laryngeal schwannoma, dermatological and gynaecological evaluation should be sort for since few studies have shown as association with skin and breast cancer especially with benign solitary schwannomas(12). Other diagnosis can include adenoma, chondroma, laryngeal cyst and internal laryngocoele.

CONCLUSION

Laryngeal schwannomas are benign slow growing neurogenic tumors. They can present with subtle symptoms such as change in voice and if unattended to can cause life threatening complications such as stridor. They are seen as a smooth mucosa covered mass usually in the region of the aryepiglottic folds. The Diagnosis is made based on histology. Definitive management is complete surgical excision. A transoral approach should be the surgery of choice in small to moderate sized tumors. Complete surgical resection is important as the disease can recur if not removed completely.

BIBLIOGRAPHY

1. Arora N, Jain K, Bansal R, Jc P. Laryngeal schwannoma - A rarely occurring benign tumor. *Otolaryngol Online J*. 2015 Mar 8;5(1.5):60-6.
2. Ebmeier J, Reineke U, Gehl H-B, Hamberger U, Mlynski R, Essing M, et al. Schwannoma of the larynx. *Head Neck Oncol*. 2009 Jul 8;1:24.
3. Zbären P, Markwalder R. Schwannoma of the true vocal cord. *Otolaryngol--Head Neck Surg Off J Am Acad Otolaryngol-Head Neck Surg*. 1999 Dec;121(6):837-9.
4. Chiu C-C, Chou S-H, Wu C-C, Liang P-I, Lee K-W. Obstructive laryngeal schwannoma in a young female.

- World J Surg Oncol [Internet]. 2015 Feb 7 [cited 2015 Dec 19];13. Available from: <http://www.ncbi.nlm.nih.gov/pmc/articles/PMC4332926/>
5. Jones SR, Myers EN, Barnes L. Benign neoplasms of the larynx. *Otolaryngol Clin North Am*. 1984 Feb;17(1):151–78.
 6. Gardner PM, Jentzen JM, Komorowski RA, Harb JM. Asphyxial death caused by a laryngeal schwannoma: a case report. *J Laryngol Otol*. 1997 Dec;111(12):1171–3.
 7. Rosen FS, Pou AM, Quinn FB. Obstructive supraglottic schwannoma: a case report and review of the literature. *The Laryngoscope*. 2002 Jun;112(6):997–1002.
 8. Lo S, Ho WK. Schwannoma of the larynx-an uncommon cause of vocal cord immobility. *Hong Kong Med J Xianggang Yi Xue Za Zhi Hong Kong Acad Med*. 2004 Apr;10(2):131–3.
 9. Schaeffer BT, Som PM, Biller HF, Som ML, Arnold LM. Schwannomas of the larynx: review and computed tomographic scan analysis. *Head Neck Surg*. 1986 Aug;8(6):469–72.
 10. Malcolm PN, Saks AM, Howlett DC, Ayers AB. Case report: magnetic resonance imaging (MRI) appearances of benign schwannoma of the larynx. *Clin Radiol*. 1997 Jan;52(1):75–6.
 11. Rognone E, Rossi A, Conte M, Nozza P, Tarantino V, Fibbi A, et al. Laryngeal schwannoma in an 8-year-old boy with inspiratory dyspnea. *Head Neck*. 2007 Oct;29(10):972–5.
 12. Das Gupta TK, Brasfield RD, Strong EW, Hajdu SI. Benign solitary Schwannomas (neurilemmomas). *Cancer*. 1969 Aug;24(2):355–66.

