CASE REPORT

Laryngeal Schwannoma Excision by Lateral Pharyngotomy

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ABSTRACT

Neurogenic tumors of the larynx are extremely rare. Here, we are reporting a case of laryngeal schwannoma presenting as supraglottic submucosal lesion with hoarseness and stridor, which was excised by lateral pharyngotomy approach. This presentation is to further discuss the histology of neurogenic tumors, modes of presentation, differential diagnosis and treatment modalities.

Keywords: Schwannoma, Neurofibroma, Larynx, Stridor, Lateral pharyngotomy.

INTRODUCTION

About 45% of all neurogenic tumors occur in the head and neck region and are mostly located in the parapharyngeal space. 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 fascicles. (Zbaren and Markwalder, 1999). Neurofibromas on the other hand derive from perineural fibrocytes, are not encapsulated and are usually intertwined with the parental nerve fascicles (Lusk et al, 1987). 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 neurofibroma (Jones et al). Around 80% are located on the aryepiglottic fold, 20% in the false or true vocal cords (Palva et al). They usually grow submucosal, with a few reports describing polypoid growth. There seems to be a slight female preponderance (Rosen et al). The internal branch of the superior laryngeal nerve is most likely the nerve of origin (Nanson). The standard treatment option for these tumors is complete surgical excision either by endolaryngeal or external approaches, according to the size and position of the tumor.

CASE REPORT

A 45-year-old lady presented to the department of ENT-HNS with the complaint of hoarseness of voice of one month duration and stridor since last 15 days. There was no history of difficulty in swallowing. Videolaryngoscopy revealed mucosal covered swelling from left aryepiglottic fold falling onto the left pyriform fossa and the glottic chink, with increase in intensity of stridor on pulling the tongue out (Fig. 1). Vocal cords were not visualized on either side. There was no significant medical history.

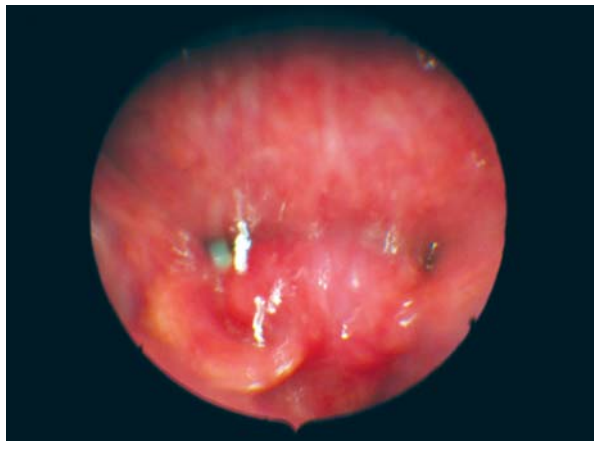

Fig. 1: Endoscopic view of the laryngeal tumor

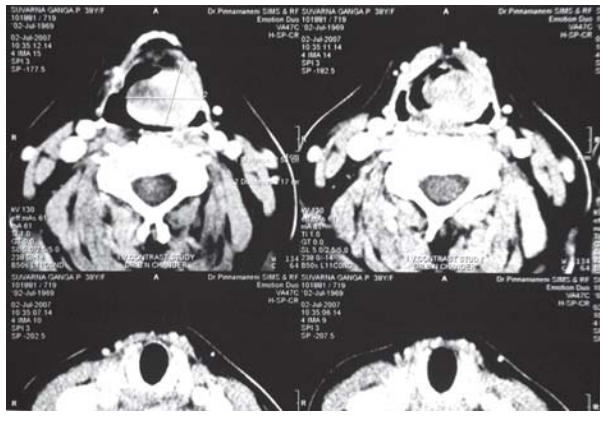

Fig. 2: CT scan showing the tumor occupying the laryngeal inlet

CT neck revealed contrast enhancing solid lesion occupying left laryngeal ventricle, paraglottic space and supraglottic area (laryngeal vestibule) (Fig. 2). In view of the lesion blocking

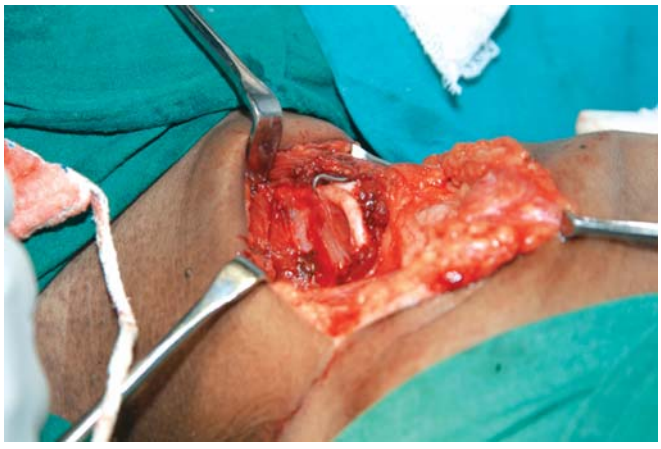

Fig. 3: Lateral pharyngotomy approach

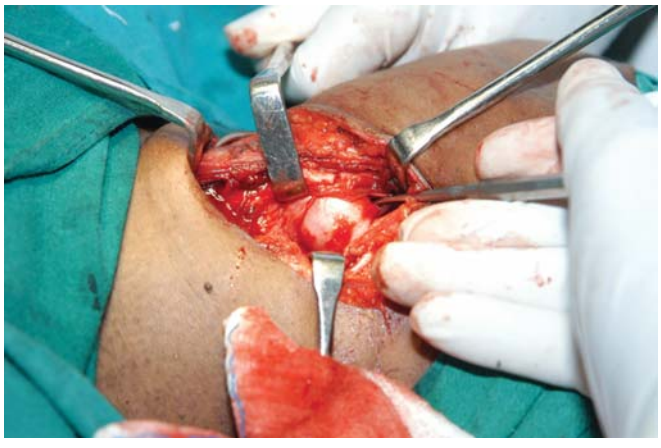

Fig. 4: Smoth tumor before being excised

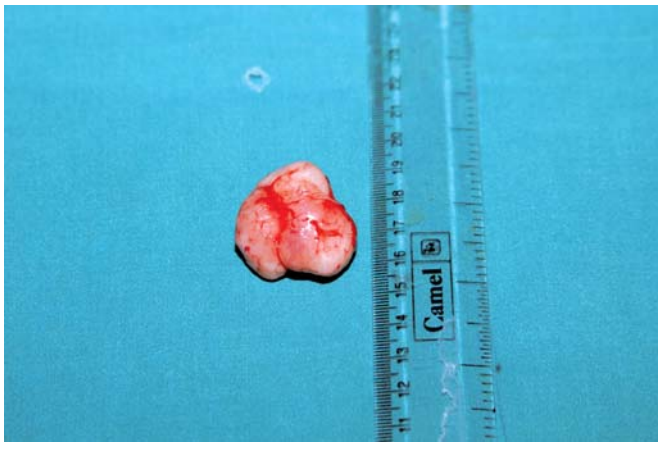

Fig. 5: Excised tumor

the glottic chink we planned for an elective tracheostomy followed by lateral pharyngotomy for excision of the lesion in the same sitting. Horizontal skin crease incision was given over thyroid prominence and subplatysmal flaps were elevated. Strap muscles were retracted laterally and thyropharyngeus was divided vertically at the lateral border of thyroid cartilage (Fig. 3). Mucosal incision was made to enter into left pyriform fossa. A firm glistening bilobed mass of 4 by 4 cm was shelled out from left aryepiglottic fold (Fig. 4). Pharyngotomy incisions were closed and the specimen was sent for histopathology, which revealed schwannoma. We could not decannulate the patient in the immediate postoperative period because of large potential space of paraglottic space causing edema of larynx with obstruction. She was decannulated after four weeks of surgery. Vocal cord movements were normal and so was the voice after decannulation. The patient was followed for two years after surgery without any recurrence.

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 tumors.[1] Neurofibromas are encountered more frequently in neurofibromatosis. Malignant transformation is reported in 10% of neurofibromas while in schwannoma it is very uncommon.[2] Neurogenic tumors of the larynx are most frequently located in the aryepiglottic fold or in the true or false vocal cords.[3]

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, dyspnea on exertion with inspiratory, sometimes biphasic stridor. Some patients complain about dyspnea in the supine position, which seems to be associated with the location of the lesion. One case of asphyxial death due to laryngeal schwannoma is reported.[4]

The true vocal cord on the affected side is usually immobile or hypomobile[5] from the bulk of the tumor, although some authors report normal mobility.[6] In many cases the bulky supraglottic tumor obstructs the view of the true vocal cord on one or both sides, as in this case total laryngeal inlet is obscured by the mass.

The diagnostic work-up 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.[7]

CT scan of the neck shows a well-defined, hypodense submucosal mass without signs of infiltrative or destructive growth with heterogeneous contrast enhancement.[5]

On MRI scanning, the lesion is expected isointense to slightly hyperintense in T1-weighted images with strong, inhomogeneous enhancement of gadolinium, in T2 the lesion is hyperintense.[8]

The differential diagnoses of neurogenic tumors of the larynx include chondroma and adenoma.[9] Also, laryngeal cysts and internal laryngocele should be taken into consideration.[5] Our differential diagnosis at the end of clinical examination were neurogenic tumor *vs* laryngeal cyst, found contrast enhancing solid lesion in CT neck.

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. Neurofibromas like schwannomas exhibit an abnormal proliferation of schwann cells. However, while schwannomas emanate from Schwann cells, neurofibromas emanate from perineural fibrocytes. They are not encapsulated and comprise a variety of cell types: elongated spindled Schwann cells interwoven with axons and collagen fibers. Thus, an important feature of neurofibroma is entwining of the tumor with the parental nerve fascicles while schwannoma grows extrinsic to the nerve fibers.

The surgical separation of the tumor from the nerve is theoretically possible in schwannoma, while in neurofibroma it is impossible.[1]

The only effective therapeutic option in benign neurogenic laryngeal tumors is complete excision. Since the diagnosis can only be made histologically, direct laryngoscopy with biopsy of the lesion will usually be the first step in treatment . However, in schwannoma biopsy can be difficult due to the solid capsule of the tumor.[5] Complete surgical excision of the tumor should be planned according to the individual requirements of each case. Most authors favor external approaches with alternative airway provisions, such as a preliminary tracheotomy in larger tumors. Median or lateral thyrotomy or median or lateral pharyngotomy are recommended. In smaller tumors, endoscopic (laser-assisted) resection of the tumor can be a reasonable treatment option.[2] Independent of the approach, restoration of the vocal cord mobility is possible even if it was immobile prior to surgery.[8] As the tumor size is more than 5 cm and occupying the entire supraglottis we have planned for an external approach and by lateral pharyngotomy approach we could deliver the mass without disturbing the laryngeal framework and the wound healed well without pharyngo-cutaneous fistula.

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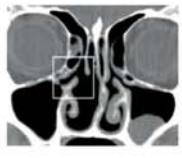

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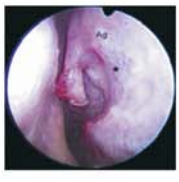

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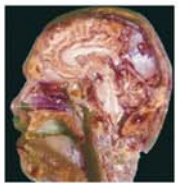

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