ANCIENT VARIANT SCHWANNOMA OF THE LARYNX – A CASE REPORT

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ABSTRACT

Ancient schwannomas are the slow growing, benign neurogenic tumor of peripheral nerves infrequently seen in larynx supposed to be originating from internal branch of superior laryngeal nerve that may remain undiagnosed, accidentally diagnosed or may cause difficulties in the differential diagnosis with other benign or malignant tumors. Here we present a case of laryngeal schwannoma diagnosed by postoperative histopathology report.

Keywords : False vocal cord, Laryngx, Schwannoma

INTRODUCTION

Schwannoma also known by the name of neurinoma or neurilemmoma is the slow growing, benign, well encapsulated tumor arising from the schwann cell. Schwannoma commonly occurs in head and neck region (45%), intracranial being more common than extracranial. Vestibular nerve intracranially and Cranial nerve IX, X, XI, XII at Para pharyngeal regions are the most common location of schwannomas. Laryngeal neurogenic tumor like schwannoma or neurofibroma occurs rarely representing not more than 1.5% of all the benign neoplasm of larynx.[1]Laryngeal schwannoma are thought to arise from internal branch of superior laryngeal nerve, however few reported case from recurrent laryngeal nerve is present.[2] Ancient variant of schwannoma is one of the various histological types. It also shows nuclear atypia and pleomorphism sometime making difficult to separate from malignant counterpart.[3,4] Due to the limited availability of the literatures that too dominated only by case reports, characteristics clinical features of schwannoma are still not specified. Endoscopy and imaging may be helpful but are not confirmatory. Histopathology is the gold standard for the diagnosis of laryngeal schwannoma supported by immunohistochemistry. It is a radioresistant tumor and complete surgical excision is the treatment modalities of choice.[1,5]

CASE REPORT

A 33 years old male presented in outpatient clinic of Department of ENT – HNS, TUTH with complain of persistent hoarseness for last 3 months. He also relates its exaggeration on exertion besides development of shortness of breath. He was an ex – smoker (3-5 sticks per day for 8 years) and has left 6 months prior to presentation and consumes alcohol socially. Intermittently he used to have worrisome foreign body sensation in throat for a year not associated with dysphagia or odynophagia. There were no known comorbidities, no past head and neck surgeries or exposure to radiation. On indirect laryngoscopy Pinkish to grayish endolaryngeal smooth, globular mass visualized, which was confirmed by nasopharyngolaryngoscopic examination to be arising from the left False vocal cord large enough

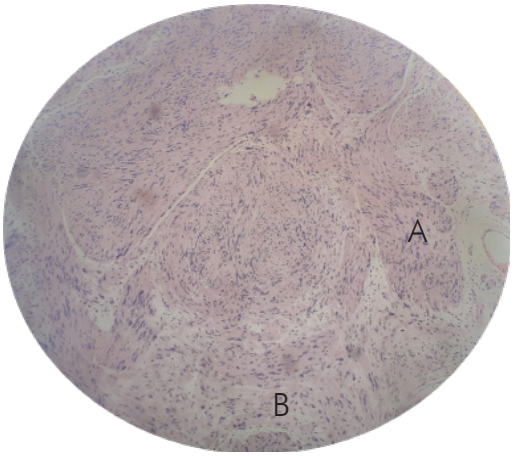

Figure I. Histologic photomicrographs demonstrating a hypercellular area composed of spindle cells with Verocay bodies (A→Antoni A) and hypocellular areas (B →Antoni B)

(~ 2x2 cm) to occlude the view of posterior 2/3rd of glottis chunk. Bilateral vocal cord mobility was not impaired. Preliminary diagnosis of left false vocal cord polyp was kept and microlaryngoscopic excision was done.

Intraoperatively tumor was found to be arising from undersurface of left false vocal cord and ventricles. Tissue collected was sent for histopathological confirmation. Macroscopically it was excised in multiple pieces, appears grey white, firm in consistency and altogether measuring 3x2cm. It was assured that no residual tissues were left behind. Microscopically well circumscribed mass with hyper-cellular and hypo-cellular (Antoni B) areas were seen. Cellular areas showed pallisading spindle shaped cells with wavy nuclei in a fibrillary background (Antoni A). Cells focally display moderate degree of atypia however no mitotic figure or necrosis seen. Areas of degenerative changes were seen like hemorrhages, hyalination to called it as ancient variants of Schwannoma (Figure I). The spindle cells were also positive for S – 100. Patient was discharged on first post-operative day with uneventful hospital stay. Nasopharyngolaryngoscopic examination performed after 2 weeks and 6 months of surgery revealed no residual lesion with bilateral true vocal folds freely mobile with improvement in voice.

DISCUSSION

Neurinomas was the term used for Schwannomas as described by Verocay in 1908, which was later modified by Stout in 1940 as schwannoma. Swchank was the first to describe in literature about schwannoma of the larynx in 1925. In the larynx most common location of these tumors is supraglottis, sites being aryepiglottic fold (80%) and false cords (20%). Schwannomas can occur in all age groups with variable gender predilection. However, most commonly seen between second to fifth decades as in our case. Most of the literature suggests female predominance in contrast to the case presented.[4,5] As they are slow growing tumor, clinically patients presents with prolonged history mainly credited to a mass effect with a variable complaints of change in voice, dysphagia,globus sensation, dyspnea on exertion or on supine position, apneic spells at sleep. Probability of going undiagnosed or accidental diagnosis on evaluation of hoarseness is usual scenario. There is always a risk of patient presenting in respiratory distress due to mechanical obstruction requiring immediate airway management with tracheostomy; although rarely reported.[6] Gardener et al. reported a case of asphyxia death due to laryngeal schwannoma causing upper airway obstruction.[7] As in our case no identifiable cause for occurance of this tumor was apparent. Schwannoma mostly are idiopathic, its association with neurofibromatosis has been noted. Besides the radiation, no other risk factor has been related to the development of laryngeal schwannoma. Literature has shown the occurance of laryngeal schwannoma following trauma during thyroid surgeries.[8] Laryngoscopic examination shows the regular, ovoid submucosal mass with or without mass effect. CT scan and MRI shows well defined, hypodense submucosal mass without erosion of surrounding structures and variable intensity on T1-weighted imaging with strong enhancement on T2-weighted Images following gadolinium injection respectively. Imaging is helpful in determining size and extent of the mass but cannot differentiate various benign condition of larynx.[1,4,5,6] Based on the clinical feature, examination and imaging differentials like laryngeal cyst, laryngoceles, chondromas, adenomas, mucoceles, lipomas and neurofibromas should be considered.[9] It is vital to differentiate a schwannoma from a neurofibroma which has a higher potential of recurrence and malignancy (approximately 10%). Main differentiating feature is that schwannoma grows externally to the nerve fibres and thus can be completely separated from nerve of origin without effecting its function, while in neurofibroma, the tumour is interlaced with the parental nerve fascicle and difficult to separate out.[1] Histopathological examination forms the main diagnostic and differentiating tool. Our case meets three criteria proposed by Enzinger and Weiss for the diagnosis of schwannoma. It was well encapsulated, consists of Antoni A and Antoni B areas and was positive for S 100.[10] Cyst formation, calcification, hemorrhage, hyalinization, fibrosis etc. are the degenerative changes associated with ancient schwannoma.[4,10] Degenerative changes itself suggest that the mass has been present for a

long duration which is not matching with our case as he gave history of persistent voice change for 3 months only. However, taking the globus sensation which was intermittently occurring for a year suggest the fact that laryngeal schwannoma can present without any significant symptoms.[5,9] Laryngeal schwannoma can be approached trans-orally or externally or both. It depends on the size and location of tumor. Small sized and endolaryngeal schwannomas are excised with microlaryngoscopy by using conventional instruments or carbon dioxide or potassium titanyl phosphate laser. Microlaryngoscopic excision reduced the risk of injury to superior laryngeal or recurrent laryngeal nerve and no recurrence with complete excision had been reported. External approach es are generally for larger tumor or tumor that are difficult for complete removal trans-orally. It includes median or lateral thyrotomy, lateral pharyngotomy, trans-hyoidpharyngotomy or extralaryngeal approach and has an advantage of proper exposure and better visualization.[2]

CONCLUSION

Laryngeal schwannoma, although rare entity, should be considered in the differential of vague hypopharyngeal or laryngeal symmptoms. Diagnosis is possible only through the histopathology and hence pathologist should also be aware about the possibility of laryngeal schwannnoma specially Cellular and Ancient variant which may mimics malignant peripheral nerve sheath tumor and misleads diagnosis. Complete surgical excision of the tumor is the treatment of choice with no recurrence and good overall prognosis.

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