

Case Report

Parapharyngeal schwannoma: a case report

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Received: 17 December 2019

Revised: 05 February 2020

Accepted: 27 February 2020

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ABSTRACT

Schwannomas are uncommon neurogenic tumors that are typically benign, slow growing, and asymptomatic. Upto 45% of schwannomas arise from the head and neck region, the sites of origin are: face, scalp, intracranial cavity, orbit, nasal and oral cavities, parapharyngeal space, middle ear, mastoid, larynx, and medial and lateral regions of the neck. The age distribution is 30 to 70 years and male:female 3:1. Symptoms and signs are neck swelling (54%), pain (11%), cranial nerve palsy (10%), oropharyngeal swelling (8%), dysphagia (6%) and trismus, otalgia (11%). This case is being presented because it is the rare case of neck swelling. A young male patient aged 22 yrs, presented with complains of swelling in right side of neck since 6 months. Patient also complains of difficulty in swallowing, difficulty in breathing and change in voice since 4 months. On examination of neck, a diffuse swelling noted at right level III neck, extending from the superior border of thyroid cartilage up to the level of hyoid bone. Horizontally the swelling was extending from mid thyroid cartilage till the anterior border of sternocleidomastoid muscle. On examination of swelling, a pink smooth globular mass seen pushing the right aryepiglottic fold anteriorly obscuring the laryngeal inlet. No pooling of saliva. elective tracheostomy was done, through lateral cervical approach, mass was excised in toto and extubated after 7 days.

Keywords: Schwannoma, Neurilemmoma, Parapharyngeal tumour

INTRODUCTION

Exclusively these schwannomas are common in 8th, 10th cranial nerves and parasympathetic chain. Hypoglossal (11th cranial nerve) schwannomas are rare. Tumours of parapharyngeal space are rare accounting approximately for 0.5% of all head and neck tumours. Schwannomas of vagus nerve must be differentiated from the carotid body and glomus vagale tumour because the distinction may influence treatment planning. Surgical treatment is complete surgical excision, transcervical approach with recurrence being rare. We describe a case of schwannoma extending from the superior border of thyroid cartilage up to the level of hyoid bone. Horizontally the swelling was extending from mid thyroid cartilage till the anterior border of sternocleidomastoid muscle. Cervical vagal schwannoma is a benign, slow-growing mass, often

asymptomatic, with a very low lifetime risk of malignant transformation in general population, but diagnosis is still a challenge.¹

CASE REPORT

A 22 years old male patient presented with a gradually progressive swelling in right side of the neck since 6 months showed in Figure 1. Patient also complains of difficulty in swallowing, difficulty in breathing and change in voice since 4 months. The following symptoms are tabulated in Table 1.

Examination showed a diffuse swelling noted at right level III neck, extending from the superior border of thyroid cartilage up to the level of hyoid bone. Horizontally the swelling was extending from mid

thyroid cartilage till the anterior border of sternocleidomastoid muscle. On examination of swelling, a pink smooth globular mass seen pushing the right aryepiglottic fold anteriorly obscuring the laryngeal inlet. No pooling of saliva showed in Figure 2.

Table 1: Symptoms.

S. no.	Symptoms	Percentage (%)
1.	Neck swelling	54
2.	Pain	11
3.	Cranial nerve palsy	10
4.	Oropharyngeal swelling	8
5.	Dysphagia	6
6.	Trismus	11
7.	Otalgia	11

P<0.001.



Figure 1: Right side neck swelling.

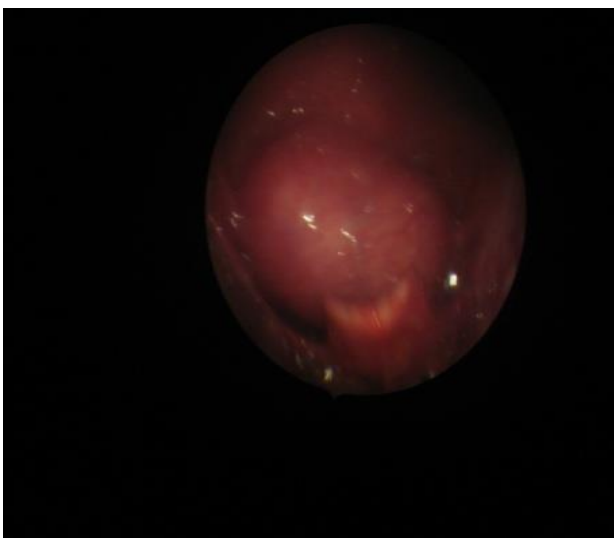


Figure 2: Indirect laryngoscopic examination.

Fine needle aspiration cytology (FNAC) showed adequate cellularity comprising of mesenchymal tissue fragments of spindle cells separated by fibrillar stroma.

These spindle cells have bland looking pallasading nuclei with pointed ends suggestive of schwannoma. Histopathologically showed antoni A and antoni B bodies, pallasading nucleus and verocay bodies suggestive of schwannoma showed in Figure 3.

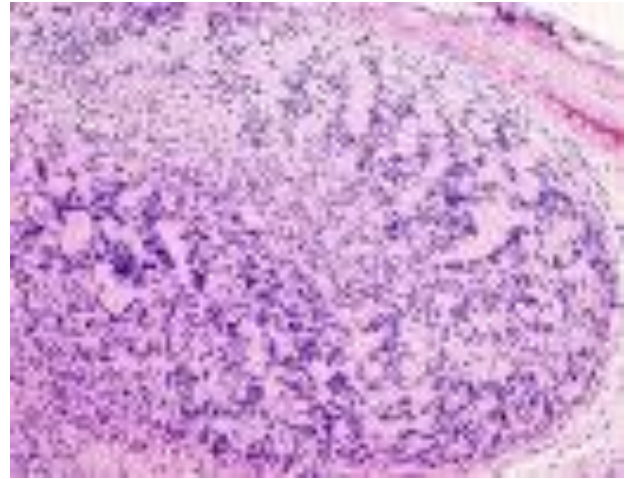


Figure 3: Histopathology, 10X.

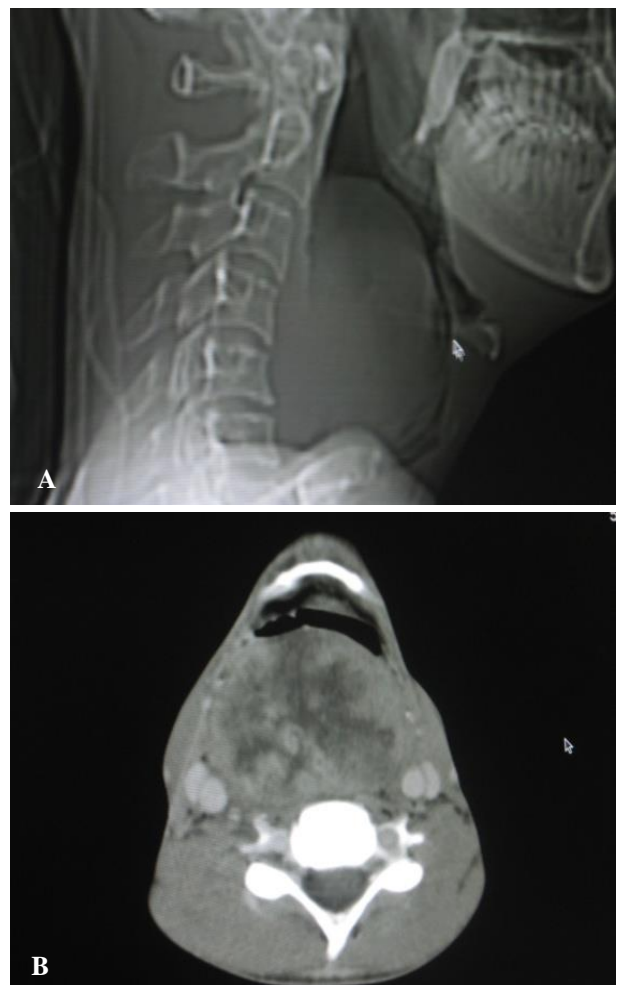


Figure 4 (A and B): Computerized tomography of neck with contrast.

Computerized tomography of neck with contrast showed a well-defined soft tissue lesion in the right parapharyngeal space extending to retropharyngeal space, well encapsulated. It showed displacement of carotid and IJV laterally showed in Figure 4.

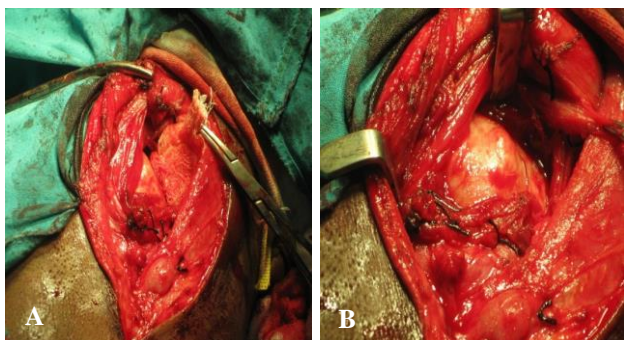


Figure 5 (A and B): Complete surgical excision with tracheostomy done.

Complete surgical excision of the mass by transcervical approach was planned due to the sheer size and location. Tracheostomy was done to secure the airway and for general anaesthesia. The drain was removed after 72 hrs and tracheostomy stoma was closed after 1 week. Patient was discharged with Ryle's tube feeding for about 3 weeks showed in Figure 5.

DISCUSSION

Using immuno-histochemistry, Schwannoma typically shows bundles of spindle cells, which are strongly positive for S-100 protein.² Neurogenic tumours of the Parapharyngeal space are rare, with 55% of these tumours being schwannomas and approximately half of these arising from the vagus nerve.³ In this case, the tumour showed two main histological patterns, consistent with a diagnosis of benign schwannoma. The more cellular areas (Antoni A) include palisading of cell nuclei of the spindle cells & round cells (verocay bodies); the less cellular areas (Antoni B) contain edematous stroma in which fibres and cells form no distinctive pattern.³

Most schwannomas occur between the 2nd and 6th decade of life, affecting both genders equally, and have minimal risk of the malignant transformation. Schwannoma presents as a slow-growing, fixed and painless mass. But in our study the swelling was mild painful in right side of neck, which was mobile in the horizontal but not in vertical direction. When schwannoma arises in the neck, in some cases hoarseness of voice is the most common symptom. However as in this case, a pathognomonic clinical sign is a cough on palpation of the mass.⁴

The term "neurilemmoma" is the preferred nomenclature. The disease presentation, differential diagnosis, and the surgical approach is discussed. The proper treatment for this tumor is surgical removal with preservation of the vagus nerve if possible and reanastomosis

and/or nerve grafting if that is not possible.⁵ For benign lesions conservative surgical excision is the treatment of choice bearing in mind possible vagal or sympathetic chain injury. Malignant schwannomas are best treated with wide excision where possible.⁶

Vagal nerve schwannoma usually occurs between the third and fifth decades of life, it does not show sex predilection both sexes being equally affected and it most often presents as a painless, slow-growing, lateral neck mass. The treatment of choice is complete surgical excision with preservation of the neural pathway, when it is possible. These tumours, in fact, are almost always benign and a conservative surgical approach is emphasized by most of the authors.⁷ Most of them are neurilemmomas, neurofibromas, and paragangliomas. In the management of neurilemmomas, because they are benign and encapsulated, intracapsular enucleation is often performed in an effort to preserve function of the nerve.⁸

Nerve sheath tumors arising from the cervical vagus nerve are extremely rare. These tumors most often present as asymptomatic, slowly enlarging, lateral neck masses and therefore often come initially to the attention of otolaryngologists and general surgeons. Because they are nerve tumors, however, neurosurgeons must be able to recognize and treat these rare entities.⁹ Microscopic evaluation confirmed the diagnosis of schwannoma, demonstrating antoni A areas (densely cellular, arranged in short bundles or interlacing fascicles) and antoni B areas (less cells, organized with more myxoid component). The tumor was S-100 positive. Antoni A areas constituted approximately 5% of tumor volume.¹⁰

Schwannoma of the vagus nerve should be differentiated from other tumours in the neck before planning surgery to minimize the risk of nerve injury due to proximity of the tumour to the vagus nerve. FNAC, Ultrasonography, CT scan, magnetic resonance imaging (MRI) are all preoperative diagnostic methods, although MRI appears to be the investigation of choice for the diagnosis and identification of the nerve of origin.

Surgical excision (trans-cervical approach) is the best surgical treatment of choice for vagal schwannoma and the surgical approach taken will depend on the tumour size, location and proximity to the vagus nerve and vessels of the neck. The main post-operative complication is hoarseness of voice due to Vocal cord palsy, sometimes associated with coughing and choking while eating. This patient developed right vocal cord palsy after surgery, and despite undergoing right medialization thyroplasty, his voice was permanently affected.

CONCLUSION

Schwannoma arising from the vagus nerve in the neck is a rare tumour that is usually asymptomatic but requires

surgical excision. Physician need to be aware of the diagnostic work-up and differential diagnosis of a neck mass, the surgical treatment and the post-operative complications and pre-operative planning is also important. The prognosis of schwannoma after complete surgical excision is good, recurrence is rare.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Manjunath K, Singh A, Smitha V. Parapharyngeal schwannomma: a case report. *Int J Otorhinolaryngol Head Neck Surg* 2020;6:799-802.