

## Parapharyngeal Schwannoma- A Rarity: Case Report with Review of Literature

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**Abstract:** Schwannomas are solitary, benign, well-circumscribed, encapsulated, slow-growing tumors originating from Schwann cells of the peripheral nerve sheath. Approximately 30-45% of cases occur in the head and neck region, of which 1-1.5% has been reported in the oral cavity. They are usually asymptomatic and malignant transformation is extremely uncommon. Few cases of intraoral schwannoma occurring in the sites such as vestibule, lips, salivary glands, palate, floor of the mouth, tongue, gingival and the mental nerve region have been reported in the medical literature. Glossopharyngeal schwannomas are rare entities and can arise either from the intracranial or from the extracranial portion of the cranial nerve. Extra cranial schwannomas usually present as a painless parapharyngeal space mass. This report describes a rare case of parapharyngeal schwannoma in 32- year old male patient. We present this case of parapharyngeal schwannoma in view of its rarity and value of radiological investigations in diagnosis the parapharyngeal masses.

**Keywords:** Schwannoma, Neurilemmoma, Parapharyngeal Mass, Peripheral Nerve, Sheath Tumors, Intra-oral schwannoma.

### INTRODUCTION

The schwannomas, also known as Neurilemmomas are typically benign, slow growing, and asymptomatic tumors which are formed by proliferating Schwann cells encompassing peripheral motor and sensory nerves [1]. About 30-45% of all schwannomas originate in the head and neck region. They are reported to occur in the face, scalp, intracranial cavity, orbit, nasal and oral cavities, parapharyngeal space, middle ear, mastoid, larynx, and medial and lateral regions of the neck [2]. Neurilemmoma was first described by Verocay in 1910. He first called it "Neurinoma". Later in 1935, Stout coined the term "Neurilemmoma". Schwannoma occur in the oral cavity at tongue, floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and region of the mental nerve. Histopathologically, more than five schwannoma variants have been described: Ancient schwannoma, Cellular, Epithelioid, Plexiform, Pigmented, Psammomatous melanotic, Microcystic-reticular variant. These tumors gradually grow to large proportions and can compress the nerve of origin or adjacent nerves causing pain and paraesthesia [3, 4]. Primary tumors arising in parapharyngeal space are very rare [5]. The parapharyngeal space, which is situated immediately outside the pharyngobasilar fascia and the superior constrictor muscle, extends from the base of the skull to the level of hyoid bone as an inverted pyramid. It is divided into a post-styloid compartment and a pre-styloid compartment by the

styloid process with muscles and a layer of fascia that extends from, and encompasses, the tensor veli palatini muscle [6, 7]. CT and MRI have had a dramatic influence on the diagnosis and surgical planning of cases arising in the parapharyngeal space since the internal carotid artery is consistently identified on magnetic resonance (MRI) images which demarcates and establishes definite diagnosis.

### CASE REPORT

A 32-year-old male patient came with complaints of swelling in the left part of the throat since 1 year with foreign body sensation and recurrent throat infections. Patient also complained of difficulty and pain during swallowing. Patient is smoker, tobacco chewer and occasional alcoholic. On general physical examination, patient was mild pallor with poor dental hygiene. Intra-oral examination shows swelling of size approximately 2.5 x 2 cm in the left parapharyngeal space with congestion. Uvula is slightly shifted towards the right side (Fig.1). Patient was advised to undergo Computerized Tomography (CT) scan and Fine Needle Aspiration Cytology (FNAC) which gave impression of benign soft tissue tumor (Fig. 2). Patient underwent surgery and the excised swelling was sent for the histopathological examination (HPE). Grossly, a well-defined and encapsulated swelling of size 2.2 x 2 cm received. Cut section show homogenous gray white appearance (Fig. 3). Representative areas were processed and slides were made which revealed Antoni

A (Hypercellular) and Antoni B (Hypocellular) areas with few areas showing amorphous, acellular, eosinophilic material representing Verocay bodies. There are few hyperplastic and congested blood vessels also noted (Fig. 4).



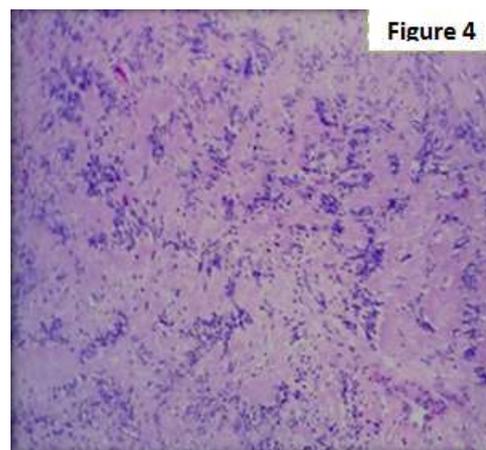
**Fig. 1: Clinical photograph of the patient shows parapharyngeal mass with midline shift of uvula**



**Fig. 2: CT scan image showing mass in the parapharyngeal space**



**Fig. 3: Gross image of the excised parapharyngeal mass**



**Fig. 4: Haematoxylin and Eosin stained photomicrograph showing Antoni- A and Antoni-B areas with Verocay bodies**

#### DISCUSSION

Schwannomas (also referred to as neurilemmomas) are benign encapsulated nerve sheath tumors composed primarily of Schwann cells in a poorly collagenized stroma [8]. It is believed to cause displacement and compression of the adjacent nerve. The parapharyngeal space is located deep within the neck lateral to the pharynx and medial to the ramus of the mandible. In the review of the literature, tumors of the parapharyngeal space are rare, approximately 0.5% of all head and neck tumors [9]. Salivary gland neoplasms are the most common parapharyngeal tumors accounting for 40-50% neoplasms, 20% are of neurogenic origin and enlarged lymph nodes comprise rest 15% to 20% [10]. Parapharyngeal schwannomas arise from the last four cranial nerves and the cervical sympathetic chain. Tumors associated with seventh nerve cause facial palsy. Tumors associated with ninth, tenth or eleventh cranial nerves cause cough and dyspnoea. The tumors involving the twelfth nerve cause impairment of tongue function [3, 11].

Schwannomas of the parapharyngeal space are usually reported to occur in patients between the age group of 30 and 70 years, there is no sex predilection. The tumor is solitary with a smooth surface, and a slow asymptomatic growth is evident; although the clinical symptoms depend on the nerve of origin. Pain and paraesthesia may be found in 50% of the patients. These tumors commonly arise in soft tissue of the head and neck region. About 30% to 45% of schwannomas occur in the head and neck and 1%- 1.5 % only demonstrates an intraoral origin. Intraoral schwannomas are rare with a negligible incidence of 0.5 %. Imaging techniques such as computed tomography (CT) and magnetic resonance (MRI) are very helpful. Initially the schwannoma appears as a well-defined tumor. Later with time cystic changes can appear probably associated with mucinous degeneration, hemorrhages

and necrosis. The differential diagnosis for a mass found in the parapharyngeal space is wide and can include tumors of the deep lobe of the parotid gland, tumors of minor salivary gland origin, metastatic cervical nodes, paragangliomas, branchial cysts, lymphomas, neurofibromas [12].

In schwannoma, classically two histological patterns are defined, Antoni-A (with hypercellularity) and Antoni-B (with hypocellularity). Antoni-A type is formed by fusiform cells with elongated nuclei arranged in a well-organized palisading pattern. Antoni-A areas with compact aggregates of spindle cells, which are separated by collagen and frequently formed parallel palisading arrays (Verocay bodies). Verocay bodies are formed by two compact rows of well-aligned palisading nuclei with intervening cell processes. Antoni-B type is composed of a smaller number of cells and the spindle cells which are randomly arranged within a loose myxomatous stroma [13, 14].

The differential diagnosis should be made with traumatic neuroma, solitary neurofibromas, granular cell tumor, neurofibromatosis, malignant schwannoma (also called neurogenic sarcoma). Glomus tumour is the other common differential for a vagal schwannoma. These are well marginated masses almost always displacing the internal carotid artery anteriorly. Enhancement is intense after contrast administration. Being hypervascular, glomus tumors show rapid initial contrast accumulation in contrast to schwannomas which, being hypovascular, show delayed filling [15]. Pleomorphic adenomas arising in the parapharyngeal space predominantly occur in the pre-styloid compartment originating from either the deep lobe of parotid gland or from the extra parotid salivary gland rests.

Immunohistochemically, schwannomas show positivity for S-100, CD 34, and epithelial membrane antigen (EMA), only in the capsule protein. Although schwannomas are typically benign, they may affect adjacent tissues by expansion with pressure effect. If the schwannoma is completely removed, recurrence rates are extremely low [13]. The treatment of choice is excision. The encapsulated form can be enucleated easily, whereas the non-encapsulated requires normal tissue margins to avoid relapse. If the nerve of origin is visualized, an attempt should be made to separate it carefully to preserve function, although this is sometimes not possible [12]. The prognosis is very good since it does not usually recur, and malignant transformation is rare.

## CONCLUSION

We present here a case of parapharyngeal schwannoma in view of its rarity. Although they are rare in the parapharyngeal space, the unique nature of

presentation imposes a systematic work-up for an accurate diagnosis which includes the histological analysis along with immunohistochemical assay integrated with the clinical data and imaging techniques. Treatment consists of surgical enucleation with periodic follow-ups.

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