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Case Report

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# CERVICAL SYMPATHETIC TRUNK SCHWANNOMA:A CASE REPORT

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## INTRODUCTION

Schwannomas are a rare nerve benign neurological tumours, arising from the nerve sheath and can arise from Schwann cells of myelinated nerves and has a rare malignant transformation incidence.

25% - 45% of all Schwannomas can rise in head and neck region, from last four cranial nerves, peripheral or autonomic nerves.

Microscopically can be composed of two cellular zones

- Antoni type A cells which is densely arranged with spindle shapedShwann cells with areas of palisading nuclei.
- Antoni type B cells which have hypocellular arrangement with largequantity of myxoid tissue.

Extracranial schwannomas can usually present as a solitary, well demarcated lesions, with gradual onset. Cervical sympathetic chain schwannomas are extremely rare.

Computed Tomography scan and Magnetic Resonance Imaging are the bestdiagnostic tools. Complete surgical excision is the treatment of choice with low recurrence rate if the tumor excised completely.

Horner's syndrome being one of the most common complications due to thedivision of the sympathetic trunk during the excision of the Schwannoma.

This case is describing a 56 years old male patient who presented with Right sided upper neck mass noticed 10 days before the presentation to ENT clinic, asymptomatic, with diagnostic tools being Carotid Doppler ultrasound, Computed tomography scan Angio, and Neck Magnetic Resonance with contrast. Tumour was excised completely through a transverse upper cervical incision with no complications developing post operatively.

The pathological and radiological evaluation, treatment and post-operative complications of this tumour are discussed below.

## CASE PRESENTATION

A 56 years old male patient, with no significant medical history, single kidney, presented to Otorhinolaryngology clinic with Right sided upper neck mass, which was noticed around 10 days prior to presentation. The patient has no family history of Neurofibromatosis 1 or 2.

No history of pain, dysphagia, dyspnoea, hoarseness or headache. No history of neck trauma or any other associated lumps.

On physical examination, there was a hard rubbery firm 4 X 6 cm lump in the Right carotid triangle of the neck, mobile, not hot nor tender, non pulsatile with no bruit. Oropharyngeal examination was normal, Flexible fibro-optic laryngoscopy showed Right sided supraglottic bulge with active mobile both vocal cords. Basic blood tests were within normal levels.

Neck Ultrasonography, Carotid Doppler US showed no abnormalities.

Neck Computed tomography angio scan showed 3.3 x 3 cm soft tissue mass lesion with minimal enhancement medial to the right sternocleidomastoid musclebehind the right submandibular angle, located between and displacing the right jugular vein (laterally) and internal and external carotid arteries (medially).

Neck magnetic resonance imaging with contrast was done and showed Right parapharyngeal oval shaped well defined mass measuring 5.2 x 3.4 x 3.6 cm whichappears pushing external and internal carotid arteries anteromedially and internal jugular vein laterally, likely presenting Schwannoma.

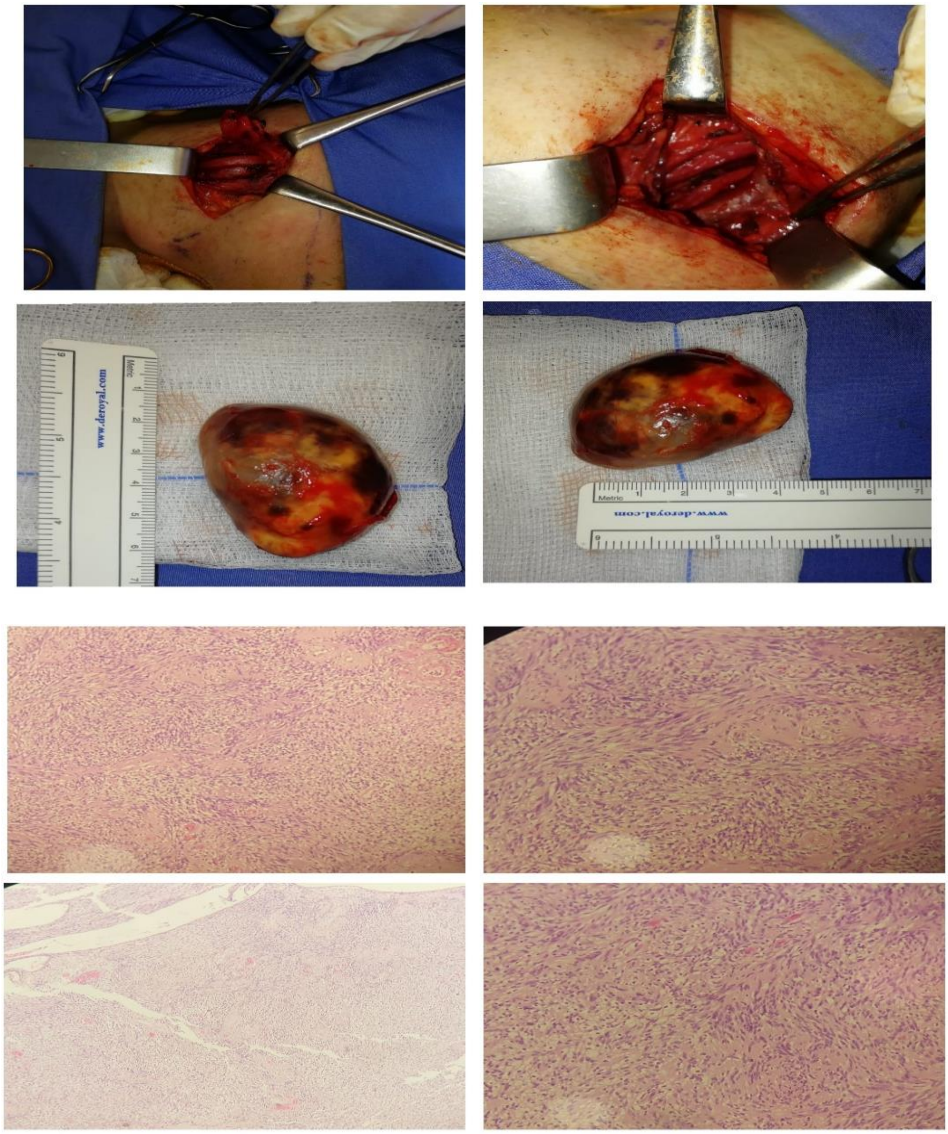
FNA was done and was not informative.

Under general anaesthesia, and Through a trans cervical transverse incision, about two cms below the inferior border of the Right body of mandible, dissection of the superficial tissue was done in layers, with lateralization of the sternocleidomastoid muscle and identification of a big oval encapsulated mass displacing the internal and external carotid arteries medically and internal jugular vein laterally, originating from cervical sympathetic chain trunk.

The mass was dissected from the carotid arteries and excised completely enbloc.

Post operatively, the patient had good smooth recovery with no signs of Horner's syndrome. Histopathology report confirmed the diagnosis of schwannoma, showing features of Antoni type A and Antoni type B cells.

Follow up for six months showed no recurrence.



Biphasic spindle cell tumor with hypercellular and hypocellular areas (Antoni A & Antoni B areas) low and high power magnification

**CLINICAL DISCUSSION**

Schwannomas, alternatively referred to as neurilemmomas or neurinomas, are benign tumors of the nerve sheath that originate from Schwann cells.

They are frequently encountered and are known to manifest predominantly in the head and neck region, accounting for approximately 25-45% of cases.

Cervical Sympathetic Chain Schwannomas (CSCS) originating from the superior or middle sections of the cervical chain, are relatively uncommon and are commonly observed in individuals aged between 20 and 50 years.

Most of these tumors present as asymptomatic slowly

growing neck masses with equal distribution among males and females.

Exceedingly uncommon in children, extradural schwannomas affecting the high cervical spinal roots have been reported. Similarly, schwannomas of the Vagus Nerve (VNCS) are also exceptionally rare.

Regarding the clinical presentation of cervical smapathatic shwanoma Schwannomas occurring in the head and neck region have the potential to affect any peripheral cranial or autonomic nerve. The signs and symptoms associated with these tumors can vary greatly and are often nonspecific. The presentation of the tumor can also be heavily influenced by the specific nerve it originates from, as well as its location and size. In the early stages of the disease, notable signs or symptoms are typically not observed. However, the most common symptom is the presence of a solitary, slow-growing mass in the neck.

As the tumor progresses, the signs and symptoms experienced by the individual are determined by the size of the schwannoma and the anatomical structures it affects. These may include dysphagia, nasal obstruction, painless swelling of the cheek, and dyspnea. The occurrence of neural deficits in patients with schwannomas is a topic of debate among experts. Some authors have reported that most patients exhibit neural deficits, while others have observed neural deficits primarily in cases involving large tumors or neural compression, typically seen in intracranial or skull base tumors.

Tumors originating from sensory nerves can manifest as pain, while those originating from the vagus nerve can manifest as hoarseness and a sensation of a lump in the throat. On the other hand, tumors originating from the facial nerve can present as facial paralysis. It is worth noting that Horner's syndrome is rarely observed in cases of schwannoma affecting the cervical sympathetic chain. In the case of a schwannoma affecting the vagus nerve, hoarseness is the most commonly observed symptom. In some instances, palpation of the mass may trigger a paroxysmal cough, which is a unique characteristic of vagus nerve schwannomas. Clinicians should be alerted to the possibility of a tumor in the vagus nerve sheath when this sign is present, particularly if the mass is located along the medial border of the sternocleidomastoid muscle.

The identification of a schwannoma presents challenges due to the lack of specific information provided by the patient's medical history and clinical examination.

However, advancements in medical technology such as fine needle aspiration (FNA), magnetic resonance imaging (MRI), and computed tomography (CT) have significantly improved the accuracy of diagnoses, reducing the occurrence of misdiagnoses. These imaging techniques not only assist in confirming the diagnosis but also offer valuable insights into the size, location, and extent of thetumor, as well as the surrounding anatomical structures. Consequently, this information plays a crucial role in the planning of surgical interventions.

The usefulness of fine needle aspiration (FNA) remains a topic of debate among experts. Many authors advise against using open biopsy or FNA for these masses. When examined under a microscope, schwannomas typically display a biphasic histological pattern known as Antoni A and Antoni B.

Antoni A areas are characterized by high cellularity, with spindle-shaped cells arranged in bundles, palisades, or whirls. Verocay bodies, which are groups of compact parallel nuclei, are also commonly observed. In contrast, Antoni B areas have fewer cells and may exhibit additional histopathological features such as cystic degeneration or xanthomatous changes. These areas do not display a distinct pattern. Immunostaining for S100 is often highly pronounced in schwannomas, particularly in Antoni A areas. This immunostaining can help distinguish neoplasms in the peripheral nerve sheaths from other types of tumors.

Schwannomas can be identified in ultra sonographic images by their distinctround or elliptical shape, which is accompanied by a well-defined border.

Additionally, the internal echo within the tumor reflects its histology. The patterns observed in these images may vary from homogeneous to heterogeneous, and there is a possibility of cystic changes being present. Ultrasonography proves to be more diagnostically useful when the diameter of the nerve from which the tumor originates is larger, and it is often observed that the tumor is connected to a distinguishable nerve.

Schwannoma diagnosis is better achieved through MRI rather than CT due to its higher sensitivity and specificity. The imaging characteristics of schwannoma include being hypo attenuated on unenhanced CT and exhibiting a mixed degree of enhancement when compared to skeletal muscle.

MRI scans can reveal certain signal characteristics that are commonly seen in schwannomas. These characteristics include isointensity or hypo intensity on T1- weighted images, intense enhancement on T1-weighted images with contrast (particularly on delayed sequences), and heterogeneously hyper intensity on T2-weighted images (which is caused by the varying degree of cellularity within the tumor). Additionally, there are typically no significant flow voids observed in schwannomas on MRI scans.

**Differential diagnosis**

1. **Vagal schwannoma** involves distinguishing it based on its impact onneighboring blood vessels.

2. **Carotid body tumors** or Paragangliomas can be identified by the presence of T2-weighted flow voids (resembling salt and pepper) and arterial enhancement.
3. **Metastatic lymph nodes** and minor salivary gland tumors, however, can be differentiated by their location. While metastatic lymph nodes can occur in various regions, minor salivary gland tumors are typically found in the parapharyngeal space rather than the perivertebral space.

Surgery is the preferred treatment option for schwannomas; however, considering their eccentric origin and non-aggressive invasion, monitoring the symptoms is also a viable alternative.

The majority of schwannomas are enclosed within a capsule. In cases where nerve fibers encompass the surface of the tumor, it is possible to perform intracapsular enucleation while safeguarding the integrity of the nerve fibers. This can be achieved by creating a small longitudinal incision in the capsule. According to Valentino et al, this technique of intracapsular enucleation, which preserves the nerve fibers, results in a retention of nerve function by more than 30% compared to tumor resection with a primary anastomosis. To ensure the preservation of neurological functions, the use of a nerve stimulator or a microscope can be employed during the procedure of intracapsular enucleation.

The prevalence of recurrence of schwannomas is a subject of significant debate, particularly when comparing total resection of the tumor along with nerve fibers and intracapsular enucleation. Zbren et al conducted a study and concluded that there is no significant difference in the recurrence rates between these two procedures. However, it should be noted that if only a partial removal of the tumor is performed, the likelihood of recurrence has been observed to increase.

## CONCLUSION

Cervical sympathetic chain schwannoma is a rare benign slowly growing tumor arising from the schwann cells of the cervical sympathetic trunk, with difficult preoperative diagnosis to establish. The management of choice is complete surgical resection of the tumor to avoid recurrence.

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