

## **Schwannoma of Cervical Sympathetic Chain**

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### **ABSTRACT**

**Schwannomas may occur spontaneously or in the context of a familial tumour syndrome such as neurofibromatosis type 2 (NF2), schwannomatosis and Carney's complex. Schwannoma arising from the cervical sympathetic chain is an uncommon benign nerve tumour and is a diagnostic challenge. This tumour most often presents as an asymptomatic solitary neck mass, with slow-growing and rare malignant degeneration. Definitive pre-operative diagnosis may be difficult and investigations are not usually helpful. Diagnosis relies on clinical suspicion and confirmation is often obtained using surgical pathology. Surgical excision is the treatment of choice for this tumour, with recurrence being rare. Horner's syndrome is a common postoperative neurological consequence but does not appear to cause problems to the patient. we describe our experience with clinical presentation, surgical management, and the outcome of a patient who presented with this pathology**

**Key words:** Schwannoma, Cervical sympathetic chain, Neurofibromatosis, Horner's syndrome

### **INTRODUCTION**

Schwannoma or neurilemmoma are benign neoplasms of Schwann cell origin. Schwannomas constitute 25–45% of tumours of the head and neck[1]. The Schwann cell surrounds peripheral nerve tissue and is believed to originate from the neural crest. They are typically solitary, well-encapsulated, benign tumours characteristically running along the course of a nerve or attached to peripheral, cranial, or sympathetic nerves. Malignant transformation is rare. In the parapharyngeal space, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the most common. Schwannomas arising from the cervical sympathetic chain are extremely rare, with less than 60 cases reported [2]and most often appear as a painless asymptomatic, slow-growing, solitary neck mass. This is because the

cervical sympathetic chain (CSC) runs in a relatively loose fascial compartment and compression is rare[3]. Horner's syndrome is rarely apparent on physical examination [4]. Preoperative diagnosis may prove difficult, for evaluation of neurilemmoma, contrast-enhancing computed tomography (CT) or magnetic resonance imaging (MRI) and ultrasound (US) are the most appropriate and carotid angiography is not necessary. Post-operative ipsilateral Horner's syndrome is to be expected following the removal of the tumour, due to the necessary division of the cervical sympathetic chain, but this does not appear to have an adverse effect on the patient

### **CASE REPORT**

A 45 years old female came with complaints of swelling over the left side of the neck for the past 10 months and mild difficulty in swallowing for 2 months. There was no history of hoarseness or syncopal attacks. Examination revealed a firm swelling of size 6X4cm located in the left mid jugular chain area(fig1), that did not move with deglutition. Indirect laryngoscopy excluded vocal cord paralysis. Ultrasonography showed a hypoechoic homogeneous well-circumscribed mass.MRI showed a solitary,poorly enhancing,non-infiltrative lesion epicentered in left carotid space, laterally displacing left internal jugular vein and left common carotid artery with no invasion (fig2A and 2B). At Surgery, the neck was explored by a skin incision along with the left sternocleidomastoid muscle. Deep to the upper portion of the sternocleidomastoid muscle, an encapsulated, white-coloured, 4× 3 cm tumour was found between the internal and external carotid artery anteromedially, and the internal jugular vein posterolaterally. Vascular structures were not stretched or compressed. The tumour appeared to originate from the sympathetic chain with the nerve roots attached to it (fig 3). The tumour was excised and the histopathology showed benign spindle-shaped cells with verocay bodies (fig 4 ). Postoperative Horner's syndrome was present(fig 5).

### **DISCUSSION**

Schwannoma in the head and neck region may arise from any peripheral, cranial or autonomic nerve, except the optic and olfactory nerve. Schwannomas of cervical sympathetic chains (SCSC)are rare, benign tumours originating from the superior or middle part of the cervical chain [5]and typically located in the retrostyloid compartment of the parapharyngeal space. SCSC occur more frequently in adults 20 to 50 years old, but is even observed in patients aged 5 to 77, without sex preference [1]Schwannoma of the vagus nerve grows between the internal or common carotid artery and the internal jugular vein causing separation of the two vascular structures; no separation has been seen between the artery and the vein in SCSC. A large tumour can displace the internal carotid artery and vagus nerve anterolaterally or the internal carotid artery also in addition to the internal jugular vein. The location and soft-tissue characteristics of the mass, along with displacement of the carotid sheath vessels, were typical of a cervical sympathetic chain schwannoma. Most SCSC appears as asymptomatic and solitary masses in the mid or upper lateral neck that tends to grow slowly, approximately 3 mm per year [6], thus avoiding compression of other structures until advanced stages. These tumours grow at least 2.5-3 cm before they are detected[7]. Horner's syndrome is rarely apparent on physical examination [8].

The main differential diagnosis in the suspect of SCSC is to distinguish it from other pathologies of the parapharyngeal space such as neurogenic tumours involving cranial nerves IX through XII and paragangliomas arising from the vagus nerve or carotid body. Deep lobe parotid

tumours, lipomas and benign and malignant lymphadenopathies can be excluded as they arise in the prestyloid compartment.

Schwannomas of the vagus nerve and the sympathetic chain cannot be distinguished without radiologic signs of separation between the common carotid artery and internal jugular vein. A superficial course of the vagus nerve on the mass or vagal connection with the tumour can be important elements to detect, respectively, a sympathetic or vagal origin of the tumour, but they are difficult to find by preoperative radiology[9].

Since cervical sympathetic chain damage is well-tolerated, restoration of the nerve has only been rarely performed, while in vagal schwannomas the practice of nerve reconstruction is recommended [6]. Thus, the majority of patients who have undergone intervention are reported to manifest some degree of Horner's syndrome, which is the most frequent complication after removal [10].

### CONCLUSION

Head and neck schwannoma though rare should be considered a differential diagnosis of a unilateral slow-growing mass in the head and neck region, particularly in an adult. Schwannomas are always a diagnostic dilemma as they are asymptomatic for a long time, and histopathology is the gold standard for diagnosis. Schwannomas of the cervical sympathetic chain do not present with specific symptoms or imaging signs. Imaging examinations cannot reveal the exact origin of the tumour. Radiological investigations can only narrow the diagnosis. Radiological investigation and fine-needle aspiration cytology make only a small contribution to its preoperative diagnosis, the histopathological examination being much more useful. The majority of patients have undergone intervention without a preoperative diagnosis, but only with a provisional suspect based on the exclusion of other similar tumours.

Malignant schwannomas are best treated with wide excision where possible. The role of adjuvant therapy remains uncertain and irrespective of treatment modality, the prognosis is poor with overall survival of 15 per cent.



**Fig 1. (Swelling in left mid jugular chain area)**

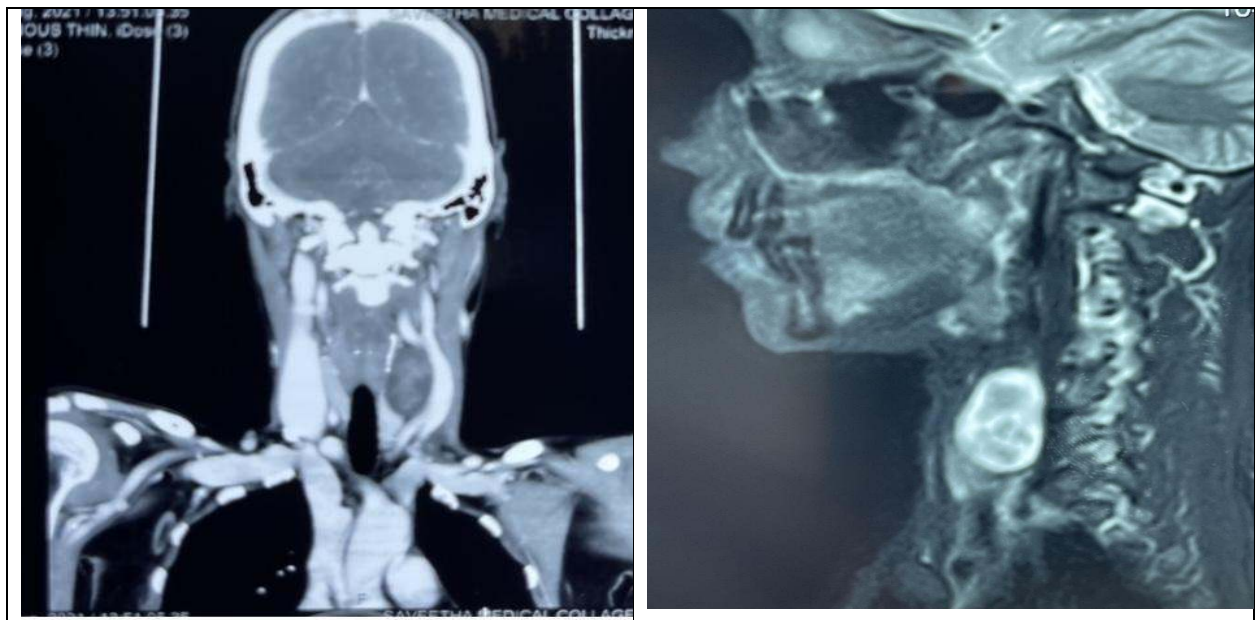


**Fig 2A and 2B (Solitary, poorly enhancing, non infiltrative lesion in left carotid space laterally displacing vessels)**

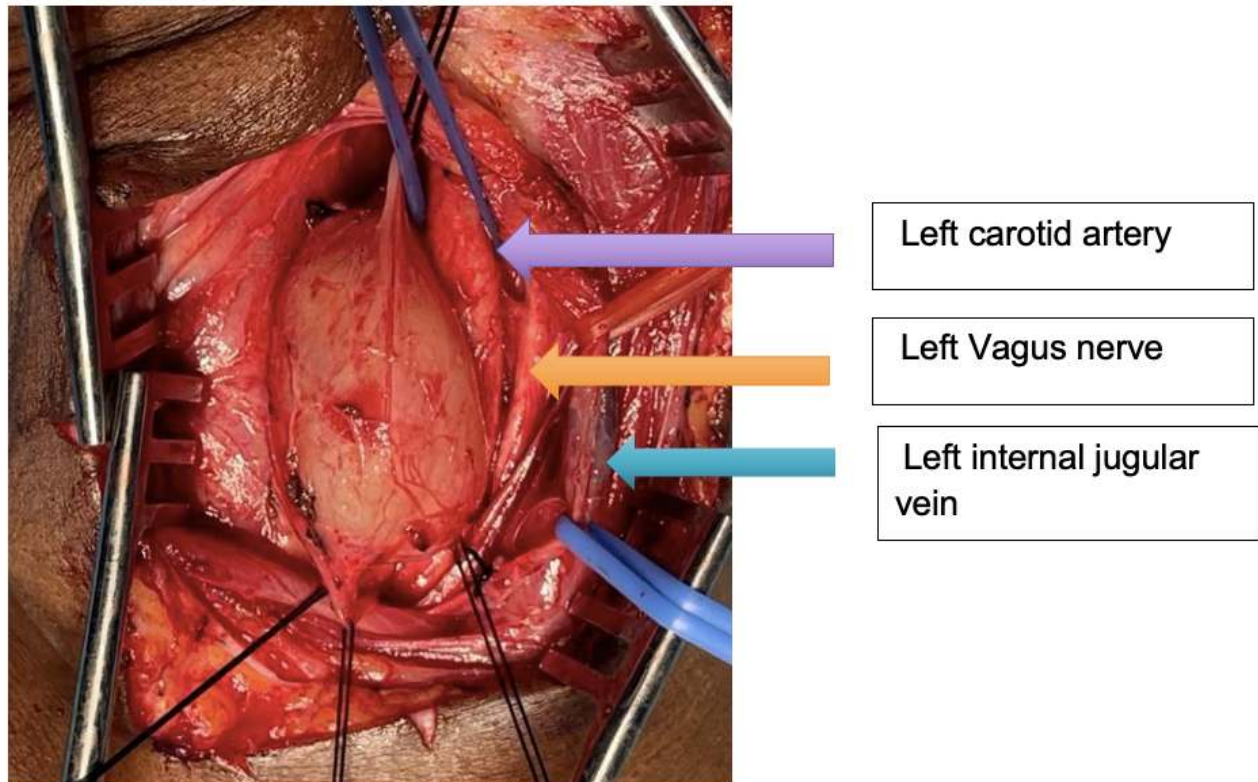


**Fig 3(Fig showing vessels around lesion ,showing no infiltration by lesion)**



**Specimen after excision**

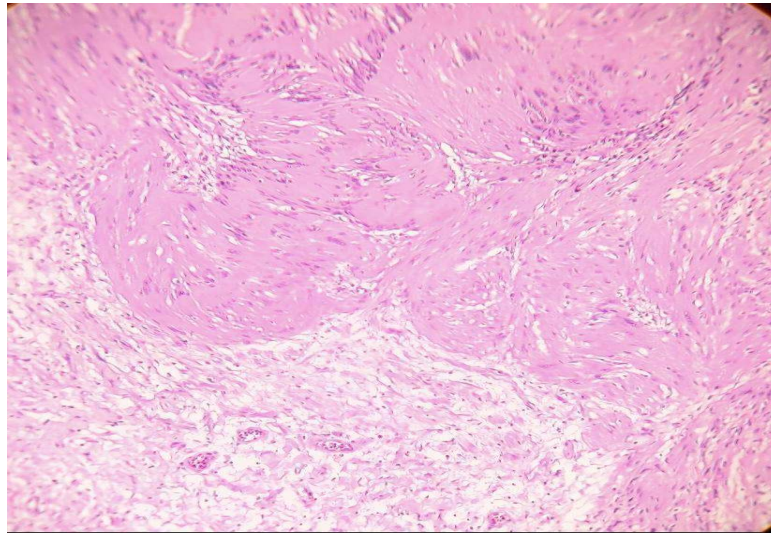
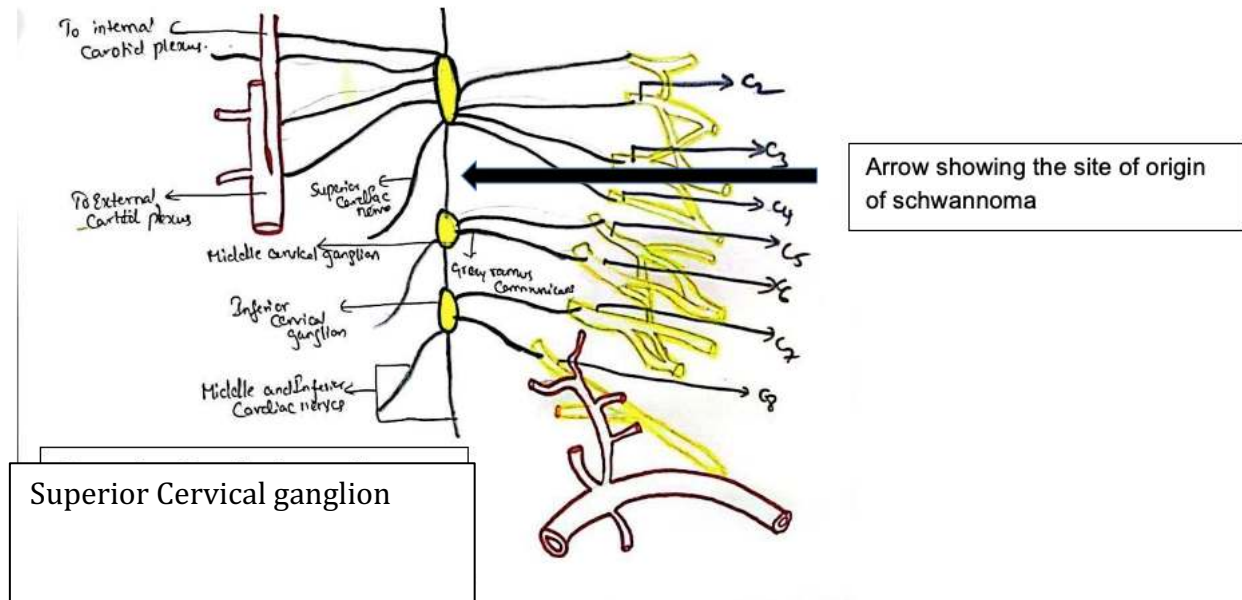


**Fig 4(SlideShowing spindle shaped cells)**



**Superior cervical ganglion(Arrow showing site of origin)**

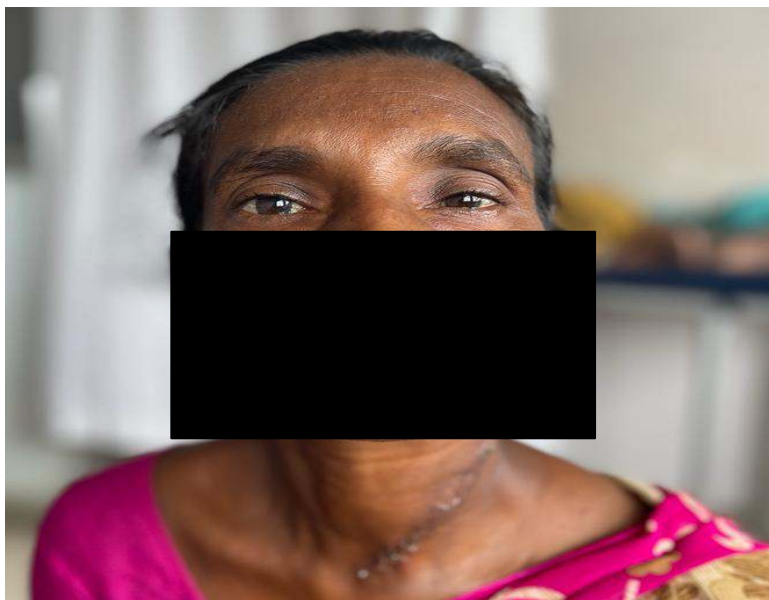


**Fig 5(Horner's syndrome in postoperative period)**

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