**Case Reports**

# Peripheral Nerve Sheath Tumor (Schwannoma), arising from an unidentifiable nerve in the neck, with Horner's Syndrome

B. I. Siddiqui, M. A. Noorani, S. A. Zaidi*, I. A. Shaikh
Departments of Surgery and Biological and Biomedical Sciences*, Dow Medical College and Civil Hospital, Karachi.

## Introduction

Schwannomas are relatively rare tumors arising from Schwann cells that sheath the peripheral nerves. Histologically classified as cellular Antoni type A and hypocellular Antoni type B areas, these are encapsulated and highly vascular tumors. They may be found anywhere in the body but 25 to 45 per cent of extracranial schwannomas occur in the head and neck region.[1] We describe a case of a 50-year-old woman who presented with a swelling on the right side of neck along with Horner's syndrome. The tumor was excised without any further neurological deficit but the nerve of origin could not be identified.

## Case Report

A 50-year-old woman with a long history of hypertension as a co-morbid, presented with a swelling on the right side of neck for seven years which had gradually increased in size. She also complained of dyspnoea on lying down flat without a pillow. There was no history of pain or dysphagia. General physical examination was unremarkable. Significant findings included a BP of 160/100 mmHg and Horner's syndrome on the right side which comprised of partial ptosis and miosis of the right eye, and hemianhydrosis.

On local examination, the swelling was 8x10 cms. in size, extending from posterior triangle and encroaching into the anterior triangle. Superiorly it was approximately at the level of hyoid bone and did not extend beyond the clavicle, inferiorly. The consistency was solid with well-defined edges. It was mobile in both quadrants and did not move on deglutition or tongue protrusion. The carotid pulse was pushed anterio-medially but there was no sign of venous engorgement.

Ultrasonograph showed a complex, well encapsulated, predominantly solid mass, suggestive of a benign growth of the right lobe of thyroid. Further workup by thyroid scan and fine needle aspiration cytology (FNAC) was advised. Thyroid Scintograph showed normal thyroid status with hormonal assay in normal range. FNAC revealed spindle like cells arranged in cellular and loose areas which were suggestive of a Nerve Sheath Tumor (Schwannoma).

Magnetic resonance imaging (MRI) showed a well defined, heterogeneous mass in the neck,appearing completely encapsulated. The mass returned heterogenous high T2 signals from its substance. It also displaced the vascular and other soft tissue structures without any evidence of invasion. This was most likely a nerve sheath tumor arising from cervical sympathetic chain.

It was decided to excise the tumor with all possible efforts to save the nerve of origin to avoid any neurological deficit. Cardiac fitness for general anesthesia was obtained after a month of antihypertensive therapy. On operation the tumor was found to be encapsulated and highly vascular. Although the nerve of origin was unidentifiable, it was definitely not arising from cervical sympathetic trunk or vagus nerve. It was 9 x 6.5 x 7.5 cms. in dimensions, oval in shape and had pushed the neuro-vascular bundle anterio-medially. Histopathological examination of the excised mass revealed features consistent with schwannoma. A small nerve was also seen compressed on one side of the lesion but there was no evidence of malignancy.

The initial post operative period was unremarkable but the patient developed severe hypertension on the second post operative day in which she was treated appropriately and was discharged on the eighth post operative day. The Horner's syndrome did not improve but no further neurological deficits were observed.

Figure 1. Clinical Photograph showing the neck mass and ptosis of the right eye.

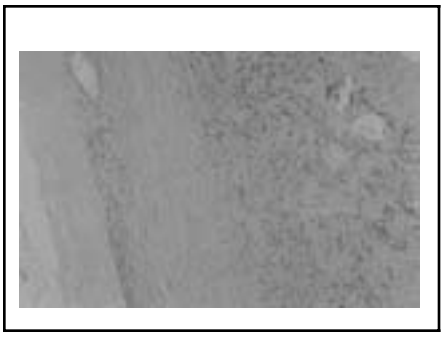

Figure 2. Photomicrograph of the excised tumor showing cellular Antoni A and hypocellular Antoni B areas with the formation of verocay bodies. A nerve is seen compressed at the periphery.

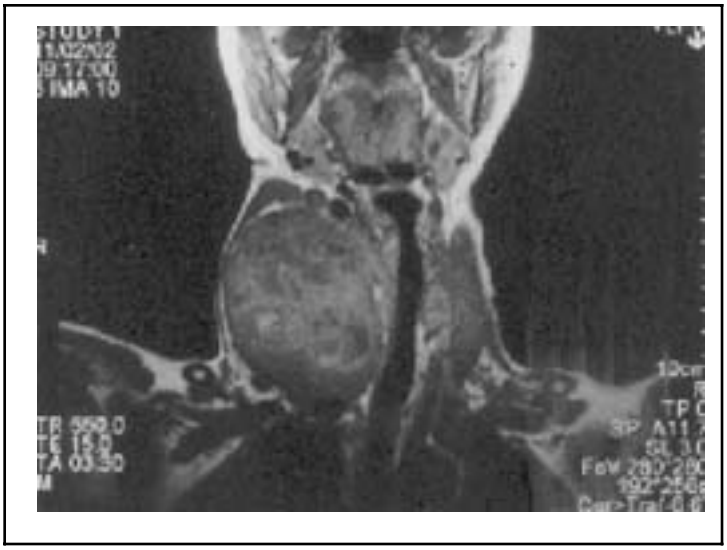

Figure 3. Magnetic resonance imaging showing heterogeneous high T2 signals from the substance of the mass. The carotid sheath and trachea are also seen displaced.

## Discussion

Schwannomas presenting with Horner's syndrome are an extremely rare clinical entity. They have been previously described as arising from cervical sympathetic chain and vagus nerve.[2,3] These tumors are usually solitary, asymptomatic cervical masses occurring within the upper carotid sheath or parapharyngeal space.[4] They are slow growing benign tumors, and must be suspected whenever examining the neck for a solitary swelling of long standing.[5] Preoperative diagnosis can be difficult, even with the aid of computed tomography, magnetic resonance imaging, ultrasound, and angiography.[6]

Although the MRI in this case, showed the mass displacing both the common carotid artery and the internal jugular vein anteriorly and medially, suggesting it's origin from the cervical sympathetic chain[7], further supported by a preoperative diagnosis of Horner's syndrome but we were surprised to find it arising from one of the small branches of some unidentifiable peripheral nerve.[8] Horner's syndrome in this patient was likely to be caused by the pressure effect on the cervical sympathetic chain proximal to the superior cervical ganglion.[9] As operative excision remains the treatment of choice, every attempt to spare the involved nerve must be made even at the expense of leaving a part of the benign tumor behind. These tumors have practically no tendency for malignant change.[5]

Hence the presence of Horner's syndrome does not guarantee the origin of the lesion from cervical sympathetic chain since Schwannomas arising from vagus nerve in the neck also present with Horner's syndrome.[3] Malignant Schwannomas are best treated with wide excision where possible.

## References


1. Colreavy MP, Lacy PD, Hughes J, et al. Head and neck schwannomas - a 10 year review. J Laryngol Otol 2000;114:119-24.
2. Ganesan S, Harar RP, Owen RA, et al. Horner's syndrome: a rare presentation of cervical sympathetic chain Schwannoma. J Laryngol Otol 1997;111:493-5.
3. Gilmer-Hill HS, Kline DG. Neurogenic tumors of the cervical vagus nerve: report of four cases and review of the literature. Neurosurgery 2000;46:1498-503.
4. Myssiorek DJ, Silver CE, Valdes ME. Schwannoma of the cervical sympathetic chain. J Laryngol Otol 1988;102:962-5.
5. Sharaki MM, Talaat M, Hamam SM. Schwannoma of the neck. Clin Otolaryngol 1982;7:245-51.
6. Hood RJ, Reibel JF, Jensen ME, et al. Schwannoma of the cervical sympathetic chain. The Virginia experience. Ann Otol Rhinol Laryngol 2000;109:48-51.
7. Furukawa M, Furukawa MK, Katoh K, et al. Differentiation between Schwannoma of the vagus nerve and Schwannoma of the cervical sympathetic chain by imaging diagnosis. Laryngoscope 1996;106:1548-52.
8. Sakao T, Noguchi S, Murakami N, et al. Neurilemmoma of the neck; a report of 35 cases. Nippon Geka Gakkai Zasshi 1990;91:407-10.
9. Clarke CR. Neurological disease. In: Kumar P, Clark M, eds. Clinical Medicine. 4th ed. London, W.B. Saunders, 1998, pp. 1007-1104.