

CASE SERIES

Four Cases of Cervical Sympathetic Chain Schwannoma and Review of Literature

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ABSTRACT

Schwannomas are benign, solitary and well-differentiated tumors originating from schwann cells. They may originate from any of the peripheral, cranial or autonomic nerves of the body with the exception of the olfactory and the optic nerves. This tumor most often presents as a slow growing asymptomatic solitary neck mass which rarely undergoes malignant transformation. Schwannomas arising from the cervical sympathetic chain are very rare. Only < 65 cases have been reported in the literature to date. Computed tomography with contrast medium or magnetic resonance imaging is essential to the initial workup for cervical sympathetic chain schwannoma (CSCS). Surgical excision is the treatment of choice for this tumor. Only surgical observation of the lesion and the nerve from where it originates, and histologic examination of the specimen, can lead to a correct diagnosis. We report four cases of cervical sympathetic chain schwannomas presented in our department during last two and half years which were treated by surgical excision.

Keywords: Cervical sympathetic chain, Horner's syndrome, Parapharyngeal tumor, Schwannoma.

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INTRODUCTION

Schwannomas were first described by Verocay in 1908. Since then they have been called as neurilemmomas, solitary nerve sheath tumors, perineural fibroblast tumors and recently as schwannomas according to the World Health Organization (WHO) classification.¹ Nearly, 45% of all schwannomas occur in the head and neck area and the commonest site in the head and neck area is the

parapharyngeal space. In the parapharyngeal space, schwannomas may arise from the last four cranial nerves or the autonomic nerves, the vagus being the most common site. Cervical sympathetic chain schwannomas (CSCSs) are uncommon^{2,3} and most often appear as an asymptomatic, slow-growing, solitary neck mass. Horner's syndrome is rarely apparent on clinical examination.⁴ Four cases of CSCSs presented in our department during last two and half years which were treated by surgical excision are reported here.

CASE REPORTS

Case 1

A 41-year-old lady presented with an asymptomatic swelling in the left upper neck of 5 years duration. Clinical examination revealed a firm 7 × 4 cm swelling in the left carotid triangle. It was mobile, nontender, and nonpulsatile, with no associated bruit. Oropharyngeal examination revealed a visible bulge in the left tonsil and posterior pillar. On indirect laryngoscopy, both vocal cords were mobile. Cranial nerve examination was normal.

Ultrasonography (US) showed a 7 × 4 cm mass displacing the carotid arteries slightly forward. Contrast-enhanced computed tomography scan of the neck revealed a well-defined contrast-enhanced mass of 7.2 × 4.2 cm in the left parapharyngeal mass pushing the carotid artery anteriorly and compressing the internal jugular vein. Fine needle aspiration cytology (FNAC) showed benign spindle cells.

A provisional diagnosis of left parapharyngeal mass was made and was taken up for excision of the mass through a transcervical approach under general anesthesia (GA). An upper transverse neck incision was put. The internal jugular vein, the common carotid artery with its external and internal branches and the vagus nerve were displaced anteriorly. The tumor was found to originate from the cervical sympathetic chain and did not involve the vagus, hypoglossal, spinal accessory, glossopharyngeal or lingual nerves. The upper part of the mass was extending to the parapharyngeal region and it was easy to recognize the cervical sympathetic chain inferior to the tumor. The mass was eccentric to the nerve and was surrounded by a well-defined capsule. Complete surgical removal of the mass, without sacrificing nerve

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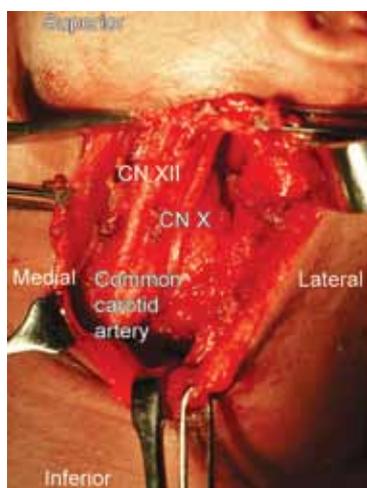


Fig. 1: Intraoperative photograph after tumor excision showing the carotid bifurcation, vagus nerve and hypoglossal nerve



Fig. 2: A pear-shaped 7 × 4 cm encapsulated tumor is excised

fibres was possible as the capsule was easily separable from the underlying fibers (Figs 1 and 2).

Surprisingly, there was no Horner's syndrome in the postoperative period. Histopathological examination of the specimen confirmed the tumor to be a schwannoma.

Case 2

A 22-year-old gentleman presented with an asymptomatic swelling in the right upper neck of 2 years duration. Clinical examination revealed a firm 8 × 5 cm swelling in the right carotid triangle. Magnetic resonance imaging (MRI) of neck showed a heterogeneously enhancing lesion of 8.1 × 5.2 cm in the right parapharyngeal space pushing the internal jugular vein and carotid artery anteriorly (Fig. 3). Excision of the mass through a transcervical approach under GA was done. Peroperatively, the tumor was found to arise from sympathetic chain displacing the IJV, carotid artery and vagus nerve anteriorly. Here, the capsule was not separable from the underlying fibers and the nerve was forced to sacrifice for the complete removal of the tumor. Postoperatively, patient developed right

Horner's syndrome (Fig. 4). Final histopathology report was consistent with schwannoma.

Case 3

A 75-year-old lady presented with an asymptomatic swelling in the left upper neck of 2.5 years duration. Clinical examination revealed a firm 5 × 2.5 cm swelling in the left carotid triangle. Contrast-enhanced CT scan of neck showed a heterogeneously enhancing hypodense lesion of 4.5 × 2.85 cm at the level of carotid bifurcation on left side displacing the IJV and carotid artery anteriorly. Peroperatively, the tumor was found to arise from left cervical sympathetic chain displacing the IJV, carotid artery and vagus nerve anteriorly (Fig. 5). Here also, the capsule was inseparable from the nerve and so was sacrificed for the complete removal of the tumor. Postoperatively, the patient developed left Horner's syndrome. Histopathology report was suggestive of schwannoma.

Case 4

A 22-year-old gentleman presented with an asymptomatic swelling in the right upper neck of 4 years duration. Clinical examination revealed a firm 6 × 3.5 cm swelling

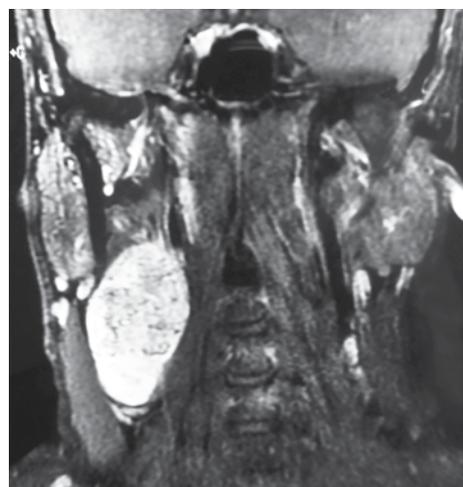


Fig. 3: Magnetic resonance imaging of neck showing a heterogeneously enhancing lesion of 8.1 × 5.2 cm in the right parapharyngeal space



Fig. 4: Postoperative picture showing right Horner's syndrome



Fig. 5: Intraoperative photograph showing the tumor arising from the left cervical sympathetic chain displacing the IJV, carotid artery and vagus nerve anteriorly

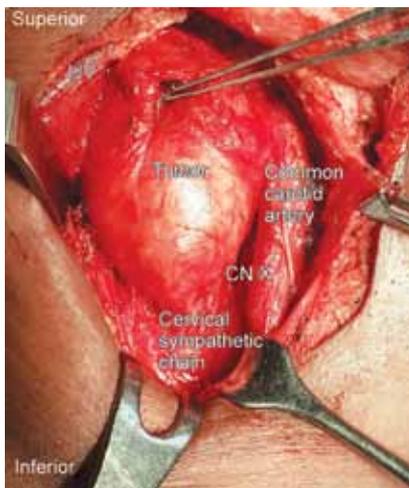


Fig. 6: Intraoperative photograph showing the tumor arising from the right cervical sympathetic chain

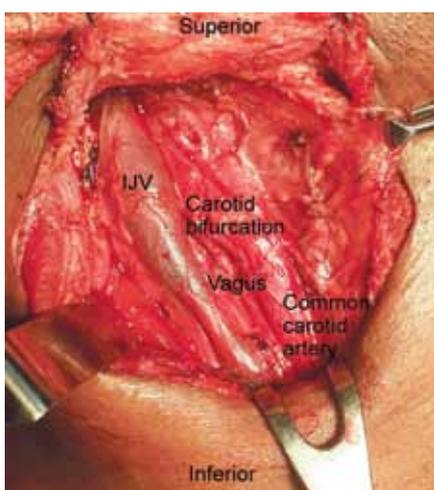


Fig. 7: Intraoperative photograph after complete tumor excision showing the carotid bifurcation, common carotid artery, vagus nerve and IJV

in the right carotid triangle with prominent carotid pulsations over the swelling. Contrast-enhanced CT scan of neck showed a well enhancing, well-defined swelling

of $4.5 \times 3.6 \times 2.5$ cm in the right carotid space posterior to the carotid bifurcation and distal common carotid artery, compressing and displacing the IJV laterally. Peroperatively, the common carotid artery and its bifurcation and vagus nerve were found to be draped over the tumor anteriorly, and the IJV was displaced laterally. The origin of the tumor was from right cervical sympathetic chain (Fig. 6). Here, also the tumor was fusiform in shape and the capsule was inseparable from the nerve and so was sacrificed for the complete removal of the tumor (Fig. 7). Postoperatively, the patient developed right Horner's syndrome. Histopathologically, the tumor was suggestive of schwannoma.

All the four patients are now free of disease and are under follow-up.

DISCUSSION

Cervical sympathetic chain schwannomas are rare, benign tumors originating from the superior or middle part of the cervical chain⁵ and typically located in the retrostyloid compartment of the parapharyngeal space. Most of the cervical sympathetic chain schwannomas present as asymptomatic solitary neck masses. They grow slowly, approximately 3 mm per year.⁶ They are usually seen in patients between 20 and 50 years of age. Frequency is the same in both sexes and malignant change is rare.³ Features of nerve compression are rare because the cervical sympathetic trunk lies in a relatively loose fascial compartment. Preoperative Horner's syndrome is unusual and has only been reported in six cases of CSCS.⁴ Pulsation is an atypical finding and suggests the presence of a carotid body tumor at initial workup. Here, angiography may be necessary for the differential diagnosis.⁷ Pulsation may be due to reflection of the carotid artery system or it may be true pulsation caused by the hypervascularity of the schwannoma.

Computed tomography with contrast medium or MRI is essential to the initial workup for CSCS. Schwannomas typically have well-delineated margins. They commonly show higher attenuation than adjacent muscle on contrast-enhanced CT, but may be isodense or, less commonly, of lower attenuation than the adjacent muscle. On MRI, schwannoma appears to have intermediate signal intensity on T1-weighted images and high-signal intensity on T2-weighted images.

Replacement of vascular structures due to the mass effect of the schwannoma on radiological imaging may give an idea of the origin of the lesion. A mass pushing the internal carotid artery or common carotid artery anteriorly is suggestive of a schwannoma originating from the sympathetic chain or vagus nerve. Schwannomas

originating from the vagus nerve cause separation of artery and vein on US Doppler since they grow between the common carotid artery and internal jugular vein or internal carotid artery and internal jugular vein. No separation is seen between the artery and the vein in CSCS.⁸ In addition, the course of the vagus nerve can be seen with ultrasound. In CSCS, the vagus nerve courses superficial to CSCS, whereas there is a connection with the tumor in vagal schwannomas.⁸

If the tumor presents as a pulsatile mass, carotid body tumor must be considered. A 'salt and pepper' pattern on postgadolinium MRI sequences is commonly seen in carotid body tumor, but it is not pathognomonic as this may be found in other hypervascular lesions and in CSCS.⁹⁻¹¹ In carotid body tumor, CT and MRI display a homogeneous and intense pattern of enhancement following intravenous contrast, while enhancement in schwannoma is less intense and dishomogeneous. Splaying of the carotid bifurcation, typical of carotid body tumor, can also be found in schwannoma arising from the lower four cranial nerves or the sympathetic chain;¹² the main imaging criterion to differentiate CBT and a nerve sheath tumour is hypervascularity. This can be demonstrated by USS, contrast CT, MRA and conventional angiography. Demonstration of contrast agent accumulation, the absence of arteriovenous shunts and low-degree vascularity in DSA should suggest a schwannoma.

Treatment of CSCS is total excision of the lesion. During surgery, the appearance of the tumour can suggest the right diagnosis, i.e. it presents as a fusiform or eccentric mass to the nerve, surrounded by a capsule. Complete surgical removal of the mass, without sacrificing nerve fibre, is possible only when the capsule is easily separable from the underlying fibers. In our first case, we could remove the mass without sacrificing cervical sympathetic chain, as the capsule was easily separable from the underlying fibres. When dissection of the capsule from the nerve is not easy and there are no signs of malignancy, functional loss can be minimized by opening the capsule longitudinally and removing the tumor from inside.¹³ Most of the time, CSCS cannot be removed without sacrifice of some nervous fibers or section of the sympathetic trunk as in our second, third and fourth cases. 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.^{13,14}

CONCLUSION

Cervical sympathetic chain schwannomas are rare tumors. These lesions are commonly asymptomatic or present with nonspecific symptoms, and accurate preoperative diagnosis is not always easy. Imaging examinations cannot reveal the exact origin of the tumor. Only surgical observation of the lesion and the nerve from where it originates, and histologic examination of the specimen, can lead to a correct diagnosis. An accurate preoperative workup is useful for surgical planning and informing the patient about any possible complications.

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