

# Cervical Spine Nerve Sheath Tumor: Surgical Experience of a Clinical Case Series.

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## Abstract

**Background Data:** Cervical spine nerve sheath tumors (SNTs) represent a special location of interest and challenge for neurosurgeons because the lesion in this region tend more to have extradural and extraspinal component (dumb-bell tumors) than dorsal or lumbar region. These tumors extension is most likely because of short intradural root in the cervical region. Dumb-bell tumors require special surgical strategy and approaches to allow complete removal of these tumors from both intraspinal and extraspinal components without traction on the spinal cord.

**Purpose:** to evaluate our experience in management of cervical spinal nerve sheath tumors with special consideration for dumb-bell tumors that operated by multidisciplinary team and discussing our result.

**Study Design:** A prospective descriptive clinical case study.

**Patients and Methods:** This was a prospective study including nine patients suffering from cervical spinal nerve sheath tumors who were treated from January 2006 to December 2012. Each case was analyzed according to sex, age, clinical presentation, magnetic resonance imaging (MRI) of the brain and whole spine, surgical intervention, pathology and outcomes. The patients were followed in our outpatient clinic, where they were assessed clinically and functionally by VAS score and JOA score as well as radiologically by MRI of the cervical spine.

**Results:** There were nine cases in this study with cervical SNTs with age ranged from 30 to 56 years in eight cases with one case reported at 6 years. There were 5 females and 4 males. Five cases in this study (56%) were dumb-bell tumors and four cases were only intraspinal. All dumb-bell tumors were operated by combined posterior and anterior approach at the same session. Six cases (67%) were schwannoma, two cases (22%) were neurofibromas and one case was malignant nerve sheath tumor. The results were generally good. 80% of patients with preoperative pain (78%) had moderate to complete pain relive. All cases with preoperative cord related motor deficit (56%) had improvement to satisfying degree.

**Conclusion:** SNTs are uncommon lesions and tend to have extradural component in the cervical region. Proper clinical and radiological evaluation should be performed to exclude Neurofibromatosis 1 or 2 with associated spinal and /or intracranial tumors. Multidisciplinary team management provides optimum result for removal of dumb-bell tumors but the posterior approach should be performed first to prevent neurological manipulation. The incidence for recurrence and reoperation of small residual tumor are generally low. (2013ESJ059)

**Key Words:** Dumbbell Neurofibroma, Spinal nerve sheath tumors, Cervical Spine, Neurofibromatosis

## Introduction

Spinal nerve sheath tumors (SNTs) constitute, together with meningiomas, the most common intradural-extramedullary tumors with equal frequency for each. This term (SNTs) include neurofibroma, schwannoma and spinal malignant nerve sheath tumors.<sup>1</sup> SNTs occur in nearly equal distribution throughout various levels of the spine.<sup>1,5</sup> Thus, these lesions were reported more in the dorsal region more than cervical and lumbar region.<sup>5</sup>

Cervical SNTs represent a special location of interest and challenge for neurosurgeons because the lesion in this region tend more to have extradural and extraspinal component (dumb-bell tumors) than dorsal or lumbar region.<sup>1</sup> Seppala et al,<sup>17</sup> suggest that these tumors in the cervical spine are more likely to extend into the extradural space because of short intradural root in the cervical region. Dumb-bell tumors require special surgical strategy and approaches to allow complete removal of these tumors from both intraspinal and extraspinal components without traction on the spinal cord.<sup>1,6,12,13</sup>

This series aim to evaluate our experience for the management of cervical spinal nerve sheath tumors with special consideration for dumb-bell tumors that operated by multidisciplinary team and discussing our result.

## Patients and Methods

This was a prospective study including nine patients suffering from cervical spinal nerve sheath tumors who were treated from January 2006 to December 2012. Each patient was analyzed according to sex, age, clinical presentation, magnetic resonance imaging (MRI) of the brain and whole spine, surgical intervention, pathology and outcomes. The patients were followed in our outpatient clinic, where they were assessed clinically by their history and physical and neurological examination. Sensory manifestations were assessed using VAS to assess neck pain. At each evaluation, patients were asked to quantify their overall pain using a VAS pain score ranging from zero to 10. Patients were also surveyed in regards to their use of pain killers. Degrees of improvement in myelopathy and muscle power were assessed by Japanese Orthopedic Association Score (JOA). Radiological follow up by MRI of the

cervical spine. The mean follow-up duration was 20 months (ranging from 4 m to 3 years).

### Surgical Procedure:

Intraspinal cervical SNTs were approached by simple posterior approach even with small extension into the neural foramen. Dumb-bell tumors with large intraspinal and extraspinal component are approached by combined anterior and posterior approach at the same session. This combined approach performed by neurosurgical and surgical oncology team specialized in neck surgery but the posterior approach were performed first to avoid spinal cord traction, which can happen if the anterior cervical approach performed without intraspinal tumor component first being untethered from the spinal cord. This combined approach by different specialized surgical teams allow complete (or near complete) removal of such large tumors at the same session with perfect results and relatively short operative time for each team.

### Anesthesia:

Extra effort and care must be taken during the step of intubation and positioning to avoid spinal cord injury. The neck should be kept in neutral position and fiberoptic intubation was considered at any difficulty. In case of large extraspinal component distorting the anatomy of the neck and prevent intubation, tracheostomy was performed. All operations are preceded by IV antibiotic and intraoperative steroids.

### Posterior Approach:

The patient was placed in prone position and sufficient laminectomy was performed to ensure complete exposure of the tumor and unilateral partial facetectomy were routinely performed in tumors with extension into the neural foramen. Operating microscope was used for opening the dura that tacked laterally to the muscle and then the arachnoid layer was opened. Tumor removal in this study started with initial debulking of such soft to firm tumors from within the capsule followed by dissection and removal of the remaining part especially if there was a well-defined plane between the tumor and the spinal cord. Such policy was adopted to protect the spinal cord and nerve roots from excessive traction. The remaining part of the tumor was inspected properly to identify the origin from the dorsal nerve root which was sacrificed in all cases to prevent recurrence. Care was taken at this

last step to avoid injury of the ventral motor roots. The part inside the neural foramen was followed, dissected and marked by a stitch to facilitate its removal through the anterior approach without neurological injury. Finally, proper closure of the dura, muscle and fascia layers was performed.

#### **Anterior Approach:**

With patient in supine position; the head and neck was elevated 20° and the head was turned towards the contra lateral surgical side, while the ipsilateral shoulder was slightly elevated and kept on slight traction toward the leg. Longitudinal incision in the neck was done through the skin, the subcutaneous tissue and platysma muscle in same level. The upper and lower flaps were raised in the subplatysmal plain. Incision through the general investing fascia and anterior border of the sternocleidomastoid muscle mobilized posteriorly.

The carotid sheath was identified and dissected from the tumor. Open the posterior border of the investing fascia from the posterior border of sternocleidomastoid muscle, identification of scalenus muscles and phrenic nerve.

If the tumor rises at C5 and C6 care must be taken to avoid the subclavian vessels injury. If the tumor arises at C3 and C4 level, the submandibular gland was retracted in upward and superiorly, together with superficial lobe of parotid gland, hypoglossal nerve and then dividing the posterior border of the digastric muscle. After that the parapharyngeal space was opened. Common carotid artery with its branches (internal and external) dissected from surrounding structures and retracted to expose the tumor

## **Results**

There were nine patients in this study with cervical NTSs with age ranged from 30 to 56 years in eight cases with one case reported at 6 years. There were 5 female and 4 males. Radicular symptoms in the form of brachialgia and less commonly upper limb weakness and/or dermatomal sensory loss was the most common manifestations and were reported in seven cases (78%) followed by cord related motor weakness manifestations in five case (56%). Three

patients (30%) had bladder manifestations. Huge neck mass was reported in two cases. Five cases in this study (56%) were dumb-bell tumors with intraspinal and extraspinal component and four case were only intraspinal. It is worth mentioning that all cases occur in fairly equal distribution at different level of subaxial cervical spine as shown in table 1

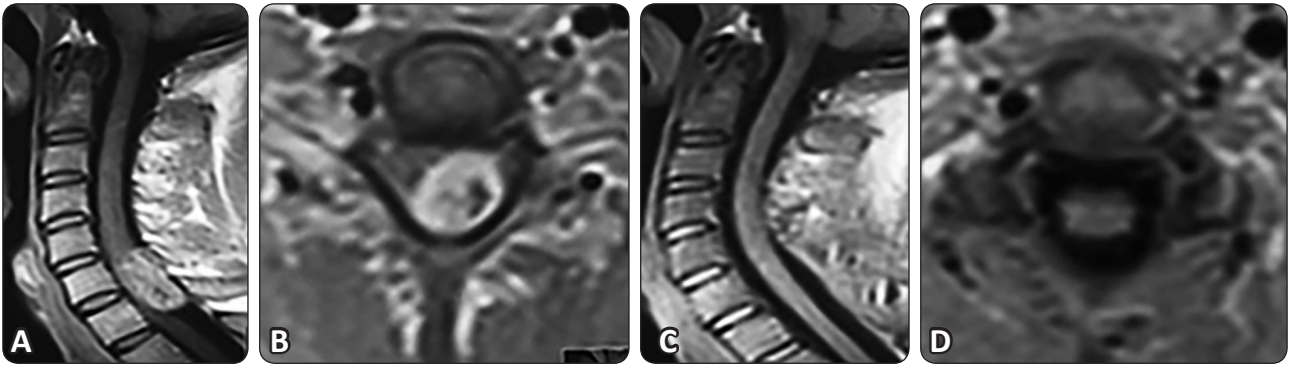
All 5 dumb-bell tumors were operated by combined posterior and anterior approach at the same session. Tracheostomy was needed in one case with large extraspinal component distorting the neck anatomy and displace the trachea which make endotracheal intubation even with fiberoptic impossible. Total removal was achieved in 3 of 5 patients. It should be mentioned that the two cases with incomplete removal were those presented with huge neck mass and the remaining part was in the extraspinal compartment. The four cases confined to intraspinal compartment were removed completely through the posterior approach.

#### **Pathology:**

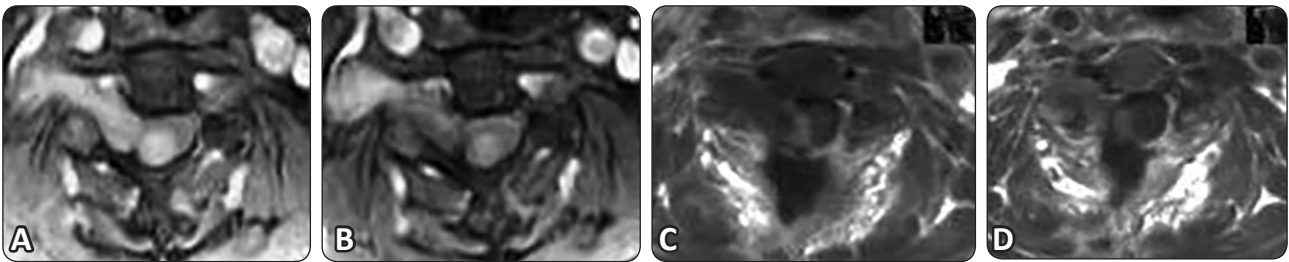
Six cases (67%) were schwannomas, two cases (22%) were neurofibromas (these patients have neurofibromatosis type I) and one case was malignant nerve sheath tumor.

#### **Clinical Outcome:**

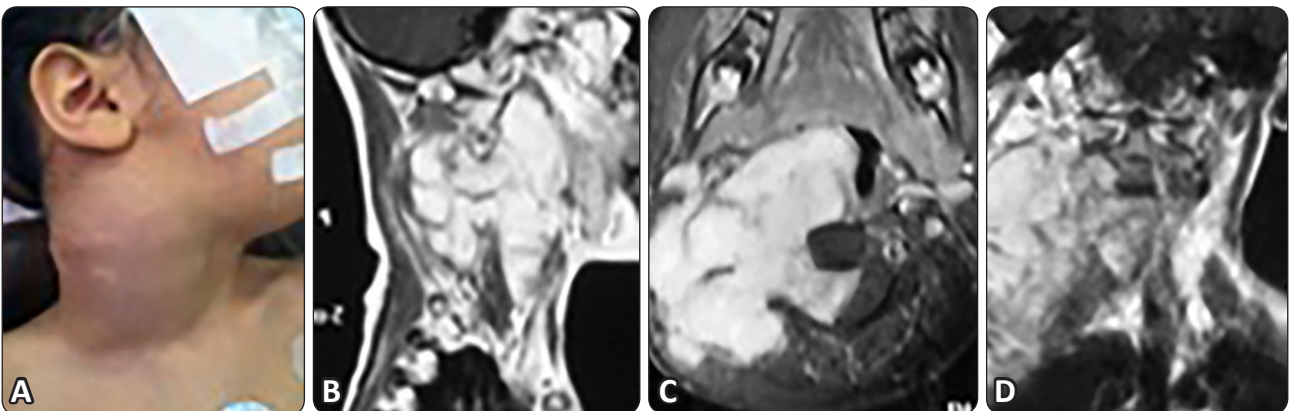
The results were generally good. All patients with myelopathy showed significant improvement. The Average JOA score preoperatively was  $8.51 \pm$  showed improvement to  $15.21 \pm$  at 12 month postoperatively. The average VAS score decreased from  $7.50.8 \pm$  preoperatively to  $20.6 \pm$  at 12 month postoperatively. All differences were statistically significant ( $P > .05$ , paired t test). None of the cases in this study showed postoperative neurological deterioration secondary to spinal cord manipulation. Two patients (22%) with dumb-bell tumors developed new radicular motor weakness without any improvement in the follow up period. Long term survival without recurrences even in case with subtotal removal were reported except in one case of malignant spinal nerve sheath tumor in whom the tumor recurred and re operated after six months. This child died ten months from the first surgery.



**Figure 1.** a,b: preoperative MRI cervical spine sagittal and axial views with contrast show intraspinal SNTs, c,d: postoperative MRI show complete removal of the tumour.



**Figure 2.** a,b: preoperative MRI cervical spine axial views with contrast show dumb-bell SNTs, c,d: postoperative MRI show complete removal of the tumor



**Figure 3.** a: giant malignant SNTs bulging through the right side of the neck, b,c,d sagittal, axial, coronal MRI images showing huge SNTs encroaching upon spinal cord, column, and adjacent structures.

**Table 1:** General data of our 9 patients included in this study.

Age	Sex	Clinical Manifestation	Location	Level	Pathology	Tumor Resection	Recurrence
56	♂	Radiculopathy Lt UL weakness	Intra/extra spinal	C3-4	benign	total	No
41	♀	Radiculopathy Neck mass	Intra/extra spinal	C4-5	benign	subtotal	No
6	♂	Myelopathy Neck mass	Intra/extra spinal	C3-6	malignant	subtotal	Yes
30	♂	Radiculopathy	Intra spinal	C6-7	benign	total	No
40	♀	Radiculopathy Neck mass	Intra spinal	C3-5	benign	total	No
45	♀	Myelopathy	Intra spinal	C2-3	benign	total	No
36	♂	Radiculopathy	Intra/extra spinal	C6-7	benign	subtotal	No
38	♀	Radiculo/ Myelopathy	Intra spinal	C3-4	benign	total	No
35	♀	Radiculo/ Myelopathy	Intra/extra spinal	C4-5	benign	total	No

## Discussion

Spinal cord tumors represent 10-20 % of central nervous system tumors. Intramedullary location represents the largest percentage in children while intradural-extramedullary tumors represent around 60% in adults, most of which are meningioma and SNSTs with nearly equal frequency for each.<sup>1,2,3</sup>

Cervical SNSTs are usually used to describe spinal neurofibroma, schwannoma and malignant nerve sheath tumors.<sup>1,4,16</sup> These tumors are reported mainly in adults with less common incidence in elderly and children.<sup>3,5,17</sup> In this study, the age of presentation ranged from 30-56 years with only one case reported in a ten years old child. These tumors have equal sex distribution but in this study there was a slight female predominance.

SNSTs usually originate from the dorsal (sensory) spinal nerve root and hence radicular symptoms are the most common manifestation.<sup>11</sup> Most of cases in this study (78%) represented by pain which was usually radicular but sometimes poorly localized and radicular weakness was evident in 22% of them. Myelopathy in the form of cord related motor weakness and bladder manifestation were reported in 56% and 25% respectively. This coincides with Levy's series<sup>11</sup> of 66 cases and Seppala's series<sup>17</sup> of 187 cases of spinal SNSTs. In addition, huge neck

mass was the main presenting symptom in two cases of this study. This late diagnosis was related to the neglect of these patients or their relatives. SNSTs may be part of Neurofibromatosis type 1 (Von Recklinghausen's disease) or neurofibromatosis type 2 and hence routine survey should be done to exclude this syndrome.<sup>1</sup> This survey was directed mainly to exclude other operable lesions that belong to this syndrome as other SNSTs elsewhere in the spine, optic glioma, acoustic tumors and any other intracranial lesion. Hence, routine MRI of the brain and whole spine should be performed in all cases. Halliday et al,<sup>8</sup> in a series of 68 patients with SNSTs reported 21 (31%) patient with neurofibromatosis type 1 and 2. In our small series there are only two cases (22%) with neurofibromatosis type 1.

The MRI with gadolinium as mentioned before is the most important investigation for preoperative evaluation of such patients. These tumors appear mostly isointense on T1 weighted images and hyper intense on T2 weighted images. All SNSTs enhance with gadolinium.

Posterior approach was the usual corridor for removal of intraspinal tumors performed in this study.<sup>9</sup>

In dumb-bell tumors with intra and extraspinal extension, there are several approaches reported

in the literature. The posterolateral approach was introduced by Sen and Sekhar<sup>15</sup> for removal of tumors in the cervical spine and foramen magnum. This approach, based on posterolateral dissection of muscles in anatomical layers, provided adequate exposure of the posterior cervical spine, vertebral artery and the vertebral triangle of the neck. Another described approach for such tumors is that described by Hakuba et al,<sup>7</sup> which is the anterolateral transuncodiscal. This procedure consists of anterior cervical discectomy and unilateral unectomy, and removal of the posterolateral parts of the related vertebral bodies in front of the tumor. This work can be extended to resection of a segment from the lateral part of the vertebral body in case of large tumors. This was followed by bone fusion.

In this study none of the previously mentioned approaches was used as the transuncodiscal approach is a complicated one with extensive bony work and also the approach (extreme lateral) is more suitable for anterolateral lesions of the foramen magnum in which exposure of the vertebral artery allow more lateral access to this region. We preferred the more simple combination of the standard posterior and anterior approaches at the same session. In addition, a combined team work of neurosurgeons for posterior approach and general surgeons specialized in neck surgery allow optimum removal of these tumors with minimal risk and effort for each team.<sup>1,6,12,13</sup>

The surgical results for such tumors in this study are generally satisfactory and coincides with other studies.<sup>1,11,17,18</sup> All of our cases with cord related motor deficit showed good degree of improvement without any case with new neurological deficit secondary to spinal cord manipulation. On the other hand, we have two cases with postoperative new radicular weakness that follow complete removal of large dumb-bell tumors. The incidence of motor roots (C5-8) deficit after surgery in most studies ranged from 0-33% and the elderly are more prone for such complication.<sup>10,14</sup>

In general, recurrence of SNTs is low. In one of the largest series dealing with such tumors and included 187 cases reported by Seppala et al,<sup>17</sup> only 20 cases have postoperative residual tumor. Nine of them have stationary course throughout 19 year follow up and in the remaining 11 cases, there are increase in size of the residual tumor that necessitated

reoperation in only two cases. In this study, all cases with only intraspinal component were totally removed without any recurrence and 3 of 5 cases with dumb-bell tumors are sub totally removed with recurrence and reoperation in one case with malignant nerve sheath tumor. Hence, leaving small part attached to important structures in the neck or foraminal motor root can be considered.

## Conclusion

SNTs are uncommon lesions and tend to have extradural component in the cervical region. Proper clinical and radiological evaluation should be performed to exclude Neurofibromatosis 1 or 2 with associated spinal and/or intracranial tumors. Multidisciplinary team management provides optimum result for removal of dumb-bell tumors but the posterior approach should be performed first to prevent neurological manipulation. The incidence for recurrence and reoperation of small residual tumor are generally low.

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## الملخص العربي

أورام غمد عصب العمود الفقري العنقي: الخبرة الجراحية لسلسلة من الحالات السريرية  
البيانات: أورام غمد الأعصاب في العمود الفقري العنقي يمثل موقع خاص من الاهتمام والتحدي لجراحي الأعصاب  
لأن الآفة في هذه المنطقة تميل أكثر لعنصر خارج الجافية وخارج النخاع من الظهرية أو منطقة أسفل الظهر. هذه  
الأورام في العمود الفقري العنقي أكثر عرضة لتمتد إلى الامام خارج الجافية بسبب قصر الجذر العصبى  
تتطلب الأورام الكبيرة التى تمتد للامام وتشبه جرس البكم استراتيجيات جراحية خاصة والنهج للسماح بإزالة كاملة  
من هذه الأورام من كل المكونات داخل النخاع وخارج النخاع دون الجر على الحبل الشوكي  
الغرض: إن الهدف من هذا العمل هو تقييم تجربتنا لإدارة الأورام غمد الأعصاب في العمود الفقري العنقي مع  
تركيز خاص على الأورام التى تشبه جرس البكم تعمل من قبل فريق متعدد التخصصات ومناقشة نتيجة لدينا.  
تصميم الدراسة: دراسة حالة سريرية مستقبلية  
المرضى وطرق: وكانت هذه دراسة مستقبلية بما في ذلك تسعة مرضى الذين يعانون من أورام عنق الرحم غمد  
العصب الشوكي الذين عولجوا خلال الفترة من يناير ٢٠٠٦ إلى ديسمبر ٢٠١٢.  
تم تحليل كل حالة حسب الجنس والعمر والسريرية، والتصوير بالرنين المغناطيسي. الدماغ والعمود الفقري كله،  
والتدخل الجراحي، علم الأمراض والنتائج. وتم متابعة المرضى في العيادة بواسطة الفحص الاكلينكى والتصوير  
بالرنين المغناطيسي للعمود الفقري العنقي  
النتائج: كانت هناك تسع حالات في هذه الدراسة تراوحت ٣٠-٥٦ عاما في ثماني حالات مع حالة واحدة ذكرت في  
٦ طفل عمرة ٦ اعوام. كان هناك نسبة اكبر فى الإناث. وكانت خمس حالات في هذه الدراسة (٥٦%) أورام تمتد للامام  
مع عنصر داخل النخاع وخارج النخاع وكانت أربعة حالات داخل النخاع فقط. وتم ازالة الاورام التى تمتد للامام عن  
الطريق الجراحي والخلفى مجتمعين وكانت مجمل النتائج مرضية.  
الخلاصة: هذا النوع من الاورام فى الفقرات العنقية يميل ان يمتد الى الامام خارج عنق العصب ويجب ان تكون  
الجراحة شاملة ازالة الجزء الامامى ويفضل لذلك فريق طبى متعدد الاختصاصات و يجب عمل رنين مغناطيسى على  
المخ وجميع مستويات العمود الفقري فى هذا النوع من الحالات للتأكد من عد وجود اورام اخرى