

High Cervical Spinal Schwannoma; Microneurosurgical Management: An Experience of 15 Cases

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Abstract-

Purpose: Schwannoma arising from high cervical spinal root are common among the spinal schwannomas. Here, we prospectively recorded the clinical features, radiological features, surgical approaches & findings, postoperative follow up & ultimate neurological outcome of high cervical spinal schwannomas and then we evaluated the records retrospectively.

Method: Here, schwannomas arising from C1, C2 and C3 spinal nerve roots were regarded as high cervical spinal schwannoma. All patients with high cervical spinal schwannomas that were consecutively operated micro-neurosurgically from 2006-2010 were included in the study. Postoperatively all patients were followed up regularly both clinically and neuro-radiologically (MRI of cervical spine).

Results: Average follow up was 31.5 months. The mean age of the series was 35.8 years (range 10-61 years). There were 8 male and 7 female patients. The mean duration of symptoms at the time of presentation was 32 months (range 06 months-5 years). Two schwannomas were completely extradural, seven were intradural and rest six were interdural or hourglass type (both extra and intradural) as identified during surgery. The standard midline posterior approach was used in all patients. A C2 hemilaminectomy or C2 laminectomy with or without cutting of posterior arch of atlas was used for most intradural and large interdural C2 schwannomas. Tumor removal was complete in all cases. Preservation of the nerve root fibers was not possible in 9 cases and was possible only in 3 cases. In two patients CSF leak developed after operation. One patient who had severe myelopathic features with bed sore failed to improve and expired 5 months after operation. Rest of the patients showed postoperative improvement in their preoperative symptoms and returned to their normal life by the end of sixth month. There was no tumor recurrence in any patient till last follow up.

Conclusion: Proper 3-D anatomical orientation & physiological knowledge, deep neuro-radiological observation, pathological appreciations and micro-neurosurgical skill and expertization can make the surgical management of these tumors (in a surgically complex site) simple with gratifying result (i.e.neurological outcome) without extensive bone removal or soft tissue manipulation through a standard midline posterior approach.

Key Words: High cervical schwannoma; C1, C2 and C3 schwannoma; Spinal schwannoma; Micro-neurosurgical management

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INTRODUCTION

Spinal schwannoma and meningioma constitutes the main bulk of spinal extramedullary tumor⁽¹⁾. Schwannoma arising from upper cervical spinal roots are commoner than any other spinal nerves⁽¹⁻³⁾. This type of schwannoma may be intradural, extradural or hourglass form (intra and extradural). In this article, we prospectively recorded the clinical features, radiological features, surgical approaches & findings, postoperative follow up & ultimate neurological outcome of high cervical spinal schwannomas and then we evaluated the records retrospectively.

METHOD

In this series, schwannomas arising from C1, C2 and C3 spinal nerve roots were regarded as 'high cervical spinal schwannoma'. All patients who were diagnosed as a case of high cervical spinal schwannoma on the basis of MRI findings and confirmed by preoperative finding and postoperative histopathology were included in this series. All patients were consecutively operated micro-neurosurgically in the department of Neurosurgery, Dhaka Medical College Hospital, Islami Bank Hospital and Ibn Sina Hospital, Dhaka, Bangladesh from 2006-2010. Preoperatively all clinical features and radiological findings were recorded for comparison with that of post operative follow up. Operative procedures and per-operative findings were noted properly. Histopathological report and early post operative complication (if any) were also recorded carefully. Postoperatively all patients were followed up regularly both clinically and neuro-radiologically (MRI of cervical spine). Clinical features, radiological features, surgical approaches & peroperative findings, postoperative follow up and ultimate neurological outcomes were studied.

RESULTS

Total number of patients was 15. Average follow up was 31.5 months. Patient's particulars, clinical presentations, locations & origin of tumor, operative approaches,

and ultimate outcomes are summarized in Table 1. The mean age of the series was 35.8 years (range 10-61 years). There were 8 male and 7 female patients. The mean duration of symptoms at the time of presentation was 32 months (range 06 months-5 years). In one of the case, duration of symptoms was 60 months and 24 months before operation, she became completely quadriplegic with complete loss of sensory and autonomic functions. She also developed bed sores three months before operation. There was mild to moderate myelopathic features in 12 cases. One of the patients presented with severe myelopathy. Two patients had no features of myelopathy. Preoperative MRC muscle grade was also shown in the Table No 1. All patients had neck pain but in one case it was severe. No patient had features of Type-1 or Type-2 neurofibromatosis or schwannomatosis.

Plain x-ray cervical spine was available in 06 cases. But no specific inference could be drawn from the radiograph. CT scan was done in two cases without significant finding. Magnetic resonance imaging was the principal investigation and was done in all patients both pre-operatively and postoperatively. The schwannomas were mostly isointense on T1-weighted images and hyperintense on T2-weighted sequences (Figure 1-5). The lesions in general had smooth contours, a homogenous appearance, uniformly mild to moderate contrast uptake. All were solid tumor, no cystic or partially cystic tumor was found in the series. In MRI vertebral arteries (VA) were clearly identified and were found mostly lateral, anterior-lateral or anterior-superior to the tumor. There was no encasement of VA in any case of the series.

The tumors varied in size and ranged from 1 × 2 cm to 7 × 4 cm. Two schwannomas were completely extradural (Figure 3 & 4), seven were intradural (Figure 2) and rest six were hourglass type (both extra and intradural, Figure 1 & 5) as identified during surgery. In four cases tumor entered largely in paraspinal area (Figure 6). These tumors extended mostly lateral, anterolaterally or anteriorly to the cord.

During operation standard midline incision was used in slightly flexed position of neck in prone position. The standard midline posterior approach was used in all

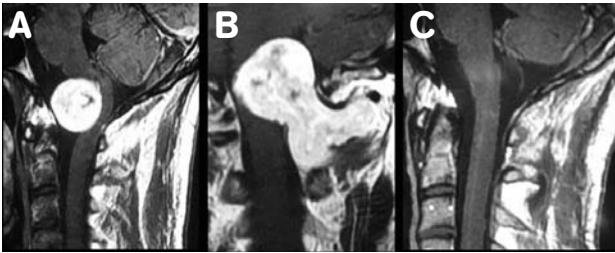


Figure 1. Pre operative contrast MRI of cranio-vertebral (CV) Junction A- sagittal view, B-Coronal view showing C1 schwannoma with intra and extradural extension. Intradural component caudo-cranially and anteriorly, C- Post operative contrast MRI of CV junction sagittal view showing no residual tumor.

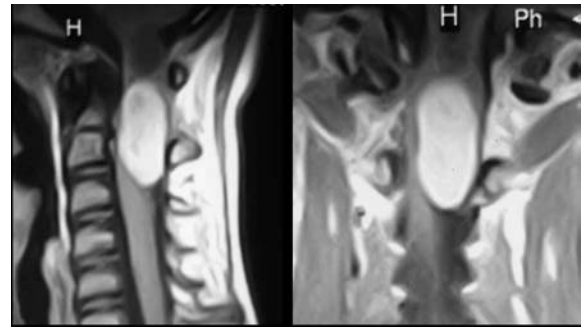


Figure 2. Contrast MRI of cervical spine A-Sagittal view B-coronal view showing complete intradural C2 schwannoma.

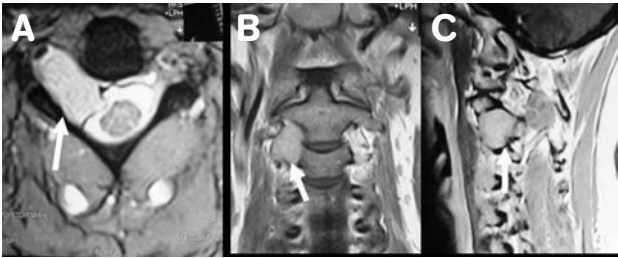


Figure 3. MRI of cervical spine A- T2W axial, B-T1W coronal, C- T1W sagittal view showing extradural (intraforaminal) C3 schwannoma (white arrow indicating tumor).

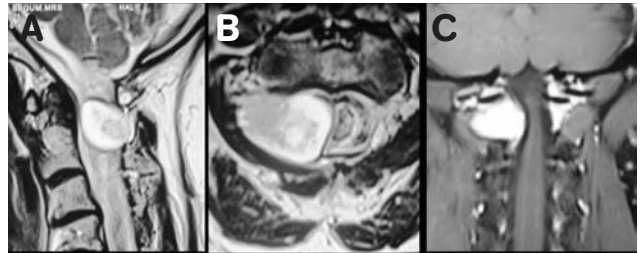


Figure 4. MRI of cervical spine T2W images, A-sagittal image, B axial image and C-coronal image showing extradural C2 schwannoma (Right).

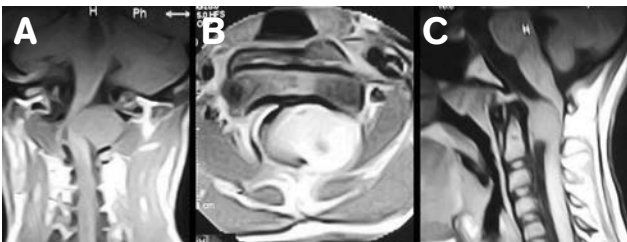


Figure 5. MRI of cervical spine A-Coronal view (T1W), B-axial view (contrast), C-sagittal view showing C2 intra and extradural schwannoma (left).

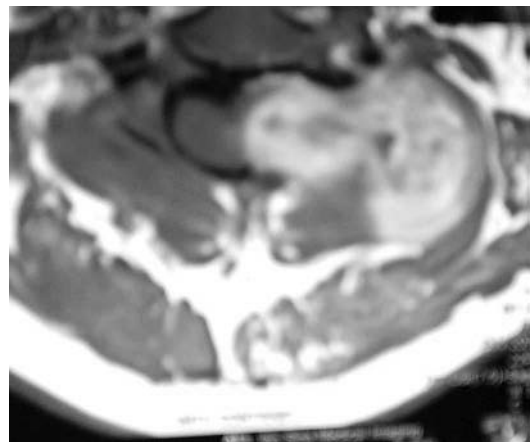


Figure 6. Contrast MRI of cervical spine axial view showing typical intra-extradural and dumbbell shaped high cervical schwannoma.

patients. The incision extended from the occipital protuberance down to the C4 spinous process. The C2 spinous process was widely exposed on the side of the tumor and the paraspinal muscles were dissected subperiosteally to expose the C2 lamina & the pedicle and the lateral aspect of the arch of the atlas. For tumor originating from C3, C3 lamina was also exposed. In extradural and

small hourglass (interdural) tumor originating C2 were excised completely without any bony resection through intratumoral resection technique.

Table 1. Patient's particulars, clinical presentations, size, locations & origin of tumor, operative approaches, complications, and ultimate outcomes

Sl No	Age/sex	Nerve involved, Location, (size)	C/F (MRC grade)	Operation	Complication/s	Last F/U	Remark
1.	48/F	Rt C2 root, Extradural on C2 pedicle (2×1 cm)	Neckpain and occipital headache	Rt posterolateral approach between C1 and C2	None	Symptoms gone	C2 nerve root and ganglion preserved
2.	52/F	Rt C2, extradural over C2 pedicle (4×3 cm)	Neckpain, rt sided weakness (4+/5) with features of UMNL	Rt posterolateral approach between CC1 and C2	None	Complete recovery	C2 root and ganglion preserved
3.	13/F	Lt C2, extra-intradural, over C2 pedicle and paraspinal ara, (4.5×3.5 cm)	Neckpain, headache, quadripareisis (MRC grade 1/5), autonomic disturbance	Lt posteriolateral approach between C1 C2	None	Complete recovery including autonomic functions	C2 root and ganglion sacrificed
4.	10/M	Lt C2, intrdural (4×2.5 cm)	Neckpain, quadripareisis (3+/5)	Posterior approach C1,C2,C3 laminectomy	None	Symptoms resolved completely	Nerve root preserved partially
5.	61/M	Lt C3, intradural, (4×3 cm)	Neckpain, quadripareisis (3/5)	Posterior approach	None	Symptoms relieved completely	Nerve root partially preserved
6.	28/F	Lt C2, intradural, (2×2 cm)	Neckpain, quadripareisis (Lt>Rt)	Poerior and Lt postero-lateral approach	None	Complete recovery	Nerve root partially preserved
7.	30/M	Lt C2, intra and extradural extending into paraspinal area, (5.5×2.5 cm)	Occipital headache, quadriplegia, autonomic disturbance	Posteriolateral approach	None	Recovered motor(4/5) and autonomic s function but persitent spasticity	Lateral dural hole could not be closed during operation, C2 root sacrificed
8.	40/F	Rt. C1, intradural, lower clivus to C2 (4×3.5 cm)	Headache, neckpain, quadripareisis (2-/5)	Posterior and rt posterior-lateral approach with suboccipital craniectomy	None	Recovered completely	C1 sacrificed. No residual on MRI
9.	22/M	Lt C2 root, intra and extradural extending into paraspinal area (6×2.5 cm)	Headache, neckpain, quadripareisis 3-/5	Posterior and posterior-lateral approach	CSF leakage (transient)	Complete recovery	Lateral dural hole could not be closed preoperatively, C2 sacrificed.
10.	32/M	Lt C2, intradural, (3×2 cm)	Neckpain, quadripareisis (3-/5)	Posterior approach	None	Sympyoms recovered completely	C2 root was not preserved
11.	51/M	Rt C2, intra and extradural, (3×2.5 cm)	Neckpain, quadripareisis (2/5)	Rt postero-lateral approach	CSF leak, reexploration needed	Recovered completely	C2 sacrificed.
12.	53/F	Rt C3, extradural upto inter vertebral foramen (1.5×2 cm)	Neckpain	Posteriolateral approach	None	Neckpain gone	C3 root preserved
13.	17/M	Rt C2, intradural (3×2 cm)	Syncopal attack, Repeated fall, signs of UMNL in all limbs	Posterior approach	None	Complete recovery	Nerve root could not be prserved
14.	29/F	Lt C2, intradural, (3×4 cm)	Neckpain, Quadriplegia, Posterior approach	Posterior approach	None	No neurological recovery	Nerve root not preserved, Expired 5 months after operation from bed sore, UTI and hypostatic pneumonia
15.	52/M	C1, intradural and extradural, (7×4 cm) with paraspinal extension.	Neckpain, quadripareisis (2+/5).	Posterior approach with midline suboccipital craniectomy	None	Complete recovery	C1 root could not be preserved

[M=male, F=female, Rt=right, Lt=left, MRC=medical research council, CSF=cerebro spinal fluid, C/F=clinical features, F/U=follow up, Sl=serial, UTI=urinary tract infection, UMNL=upper motor neuron lesion, C1=cervical 1st spinal nerve, C2=cervical 2nd spinal nerve, C3=cervical 3rd spinal nerve]

In most cases, a C2 hemilaminectomy or C2 laminectomy with or without cutting of posterior arch of atlas was used for most intradural and large intra-extradural C2 schwannomas. One of the C3 extradural schwannoma was removed by exposing the canal through lateral mass (Figure 3), facet joint on that side was compromised where fusion done after tumor removal. In rest of the cases joint instability was not suspected after tumor removal. Other case of C3 schwannoma was removed by removing C2 and C3 lamina. In C1 nerve root schwannoma, posterior arch of atlas excision (more on the tumor side) with small suboccipital craniectomy was used for tumor removal. Among the 15 cases, 2 were C1 (Figure 1), 2 were C3 (Figure 3) and rest 11 was C2 schwannoma (Figure 2, 4 & 5). In no case VA came in view and no volunteer effort was made to see the VA. In C2 schwannoma venous sinus seemed to be pushed back and compressed. It did not make any troublesome venous bleeding in any case. When the posterior surface of the tumor was exposed, its posterior dural wall was incised and intratumoral debulking was performed.

Most of the tumors were firm in nature; some were soft and friable. Bleeding was minimum in most of the cases. No cyst or cystic degeneration was found in any of the cases. Preservation of the nerve root fibers was not possible in 9 cases. In 3 cases nerve roots were partially preserved and total nerve root preservation was possible only in 3 cases. Tumor removal was complete in all cases. Two patients developed CSF leak through the wound post operatively. One patient stopped CSF leak by superficial re-suturing of the wound and transient lumbar subarachnoid drain; whereas in the rest of the case re-exploration was needed to close the lateral hole of the dura with obliteration of dead space by mobilization of paraspinal muscle. Histopathological examination confirmed schwannoma.

One patient who had severe myelopathic features with bed sores failed to improve and expired 5 months after operation from bed sore, urinary tract infection and hypostatic pneumonia. In another one case, patient improved neurologically and returned to his daily life with mild motor deficit (MRC grade 4+/5) and mild

spasticity that persisted at last follow up (12 month after operation). Rest of the patients showed postoperative improvement in their preoperative symptoms and returned to their normal life by the end of six months. No patient complained for dysesthesia or hyposthesia in the C2 or C3 dermatome postoperatively. There has been no tumor recurrence in any case till last follow up.

DISCUSSION

At cranio-vertebral junction and higher cervical spinal region meningioma are commoner than schwannoma (3:1). Here other rare tumors are dermoids, teratoma, paraganglioma, intradural extraosseous chordomas⁽⁴⁾. Spinal schwannomas make up approximately 30% of all primary spinal cord tumors. 70% schwannoma arises from sensory root, 20% from motor roots and rest from both motor and sensory roots^(5,6). About 75% schwannomas are intradural, 10% intra-extradural and rest (15%) are extradural⁽⁷⁾. Though some authors have noted a predominance of extradural location of schwannomas at C-2^(2-4,8-12). Among the schwannomas arising from C1, C2, & C3 spinal roots, C2 spinal root tumors are commoner and constitutes 15% of all spinal schwannomas^(1-3,7,9,10-19). In our series, among the C1, C2 and C3 spinal schwannoma, 11(71%) originated from C2 spinal nerve root.

A multiplicity of schwannomas is frequently noted in cases involving NF Type 1. A multiplicity of schwannomas at any spinal level is reported in approximately 4% of cases^(8,15). Multiple schwannoma was absent on our series.

In our series male and female ratio was almost equal, whereas other authors had reported a higher incidence of these tumors in females^(3,6,9). Symptoms and signs are usually diagnostic in nature. However, unusual symptoms of syncopal attacks, migraine headaches, and unrelated motor & sensory symptoms, can lead to misdiagnosis^(2,20-24). In one of our case there was history of syncopal attack and fall.

These tumors are slow growing and usually attain a large size before becoming symptomatic⁽²⁾. Dumbbell-shaped tumors located elsewhere in the spine are gener-

ally classified as intraspinal, foraminal, and extraforaminal⁽¹¹⁾ and can also occur at high cervical region^(8,10,11,25). We conveniently divided these tumor into intradural, intra-extradural and extradural group. Malignant changes of such tumor are marked by infiltration of paraspinal muscles and destruction which is rare⁽²⁾.

These tumors are located lateral, anterolateral, or anterior to the spinal cord. Several posterior, posterolateral, lateral, and anterolateral approaches have been described to surgically approach these tumors^(8,10,11,15,26). But we are convinced from our small experience that standard midline posterior approach is suitable and most appropriate to resect almost all types of C1, C2 and C3 schwannomas with transtumoral resection techniques. A similar surgical strategy had been reported by several investigators in the past^(10,11,27,28).

These tumors are usually moderately vascular and firm inconsistency. Such schwannomas are relatively simple to resect because they have a well-defined arachnoid plane of dissection intradurally and well defined capsule extradurally⁽²⁾. The exposure used in the surgery (midline posterior approach) is standard and quick and there is no need for manipulation of any cranial nerves, blood vessels, or joints to affect exposure⁽²⁾.

Some authors have suggested proximal control of the VA prior to tumor resection^(8,29). There is a low risk of VA injury in this surgery. So preoperative angiography or intraoperative proximal or distal control of the VA does not appear to be necessary^(13,30). Concerning bony stability, after laminectomy of cervical spine, instability or deformity frequently occurred, especially in dumbbell type tumor, in which cases additional bony removal is required to remove the tumor. Although no immediate instability is noted, long time follow up is advised for this problem⁽¹⁵⁾. But if instability is present it should be stabilized immediately⁽²⁾. In our series we did not face any instability.

According to the new WHO classification of brain tumors⁽³¹⁾, tumors of cranial and spinal nerves are classified as-schwannoma /neurinoma (variants: cellular, plexiform, melanotic), neurofibromas (circumscribed, plexiform), malignant peripheral nerve sheath tumor (MPNST) (neurogenic sarcoma, anaplastic neurofibro-

ma, "malignant schwannoma"). Variants of MPNST are epithelioid, MPNST with divergent mesenchymal and/or epithelial differentiation and melanotic⁽³¹⁾. Again histopathologically, spinal schwannoma may be Antoni A and Antoni B type. Type A tissue is highly cellular and demonstrates nuclear palisading and associated Verocay bodies, which may reflect their prominent extracellular matrix and secretion of laminin. Type B tissue is loosely organized with myxomatous and cystic changes and may represent degenerated Antoni A tissue⁽²⁵⁾. Successful tumor resection results in rapid and sustained neurological recovery^(2,27,28). Total tumor resection is advocated by several authors for complete treatment of the disease. The significant chance of early tumor recurrence in a partially excised tumor is known⁽³⁾, and every attempt should be made to remove them completely⁽²⁾.

In spite of the critical location of these tumors, with an anterior or anterolateral extension to the spinal cord and a relationship with vital neural structures & the VA, surgery for C1, C2 & C3 schwannoma is usually very successful. If the anatomy of the tumor in relationship to normal structures in the vicinity is appropriately understood, then surgery in these formidable-looking tumors is relatively uncomplicated⁽⁸⁻¹¹⁾.

CONCLUSION

Among high cervical spinal schwannomas C2 nerve root originating tumors are predominant. Proper 3-D anatomical orientation & physiological knowledge, deep neuro-radiological observation, pathological appreciations and micro-neurosurgical skill and expertization can make the surgical management of these tumors (in a surgically complex site), simple with gratifying result without extensive bone removal or soft tissue manipulation through a standard midline posterior approach.

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