



THE AGA KHAN UNIVERSITY

eCommons@AKU

Section of Neurosurgery

Department of Surgery

April 2004

Complete surgical excision of intramedullary schwannoma at the craniovertebral junction in neurofibromatosis type-2

A.A. Siddiqui

A.A. Shah

Follow this and additional works at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_neurosurg

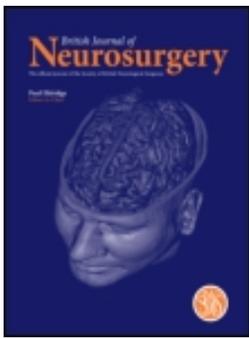


Part of the [Surgery Commons](#)

Recommended Citation

Siddiqui, A. A., Shah, A. A. (2004). Complete surgical excision of intramedullary schwannoma at the craniovertebral junction in neurofibromatosis type-2. *British Journal of Neurosurgery*, 18(2), 193-196.

Available at: https://ecommons.aku.edu/pakistan_fhs_mc_surg_neurosurg/120



Complete surgical excision of intramedullary schwannoma at the craniovertebral junction in neurofibromatosis type-2

AA Siddiqui & AA Shah

To cite this article: AA Siddiqui & AA Shah (2004) Complete surgical excision of intramedullary schwannoma at the craniovertebral junction in neurofibromatosis type-2, British Journal of Neurosurgery, 18:2, 193-196, DOI: [10.1080/02688690410001681109](https://doi.org/10.1080/02688690410001681109)

To link to this article: <https://doi.org/10.1080/02688690410001681109>



Published online: 06 Jul 2009.



Submit your article to this journal [↗](#)



Article views: 19



Citing articles: 4 View citing articles [↗](#)

Complete surgical excision of intramedullary schwannoma at the craniovertebral junction in neurofibromatosis type-2

A. A. SIDDIQUI & A. A. SHAH

Division of Neurosurgery, Aga Khan University, Karachi, Pakistan

Abstract

Intramedullary spinal schwannomas are extremely rare. We describe a case of a 13-year-old female who was found to have an intramedullary schwannoma at craniovertebral junction along with incidental bilateral acoustic neuromas, which was excised completely. To our knowledge, this is the first report of a case of intramedullary schwannoma in association with NF-2 located at the craniovertebral junction. The literature has been reviewed with emphasis on radiological imaging, histopathological diagnosis and technique to achieve complete microsurgical excision.

Key words: *Complete surgical excision, craniovertebral junction, intramedullary schwannoma, spinal cord schwannoma.*

Introduction

Schwannomas (neuromas or neurolimmomas) account for 30% of primary spinal neoplasms and, of those, intramedullary spinal schwannomas represent only 0.3–1.5%.¹ Solitary intramedullary schwannoma (without neurofibromatosis) is the commonest presentation.^{2,3} Association of intramedullary schwannomas with neurofibromatosis type-2 (NF-2) is extremely rare^{4,5} and, to our knowledge, only six cases have been reported, which were mainly located in the cervical and thoracic regions.^{1,4,6–8} This is the first report of a case with intramedullary schwannoma at the craniovertebral junction in a patient with bilateral acoustic neuromas (NF-2). This was completely excised by using microsurgical techniques. The aim of this report is to place emphasis on radiological imaging, histopathological diagnosis and related surgical implications to achieve complete excision.

Case report

A 13-year-old female presented with complaints of pain in the back of neck and weakness of all four limbs for the past 6 months. The weakness was more pronounced on the left side, but with no sphincteric disturbances. There were no complaints of headache, fever, difficulty in breathing or hearing problem. On clinical examination, the motor weakness was more on left side (2/5) than on the right (3/5), with hypertonia and hyper-reflexia in all the four limbs and sustained ankle clonus. There was no

definite pin-prick sensory level, but position sense was impaired bilaterally.

MRI of the craniovertebral junction showed an intraspinal mass lesion. On T1-weighted images, the lesion was hypo- to isointense extending from above the foramen magnum to the C3 level, filling the whole spinal canal along with a segmented syrinx visible below this. On T2-weighted images (Fig. 1) the lesion was hypointense pushing the cord anteriorly. Gadolinium-enhanced study (Figs 2 and 3) revealed heterogeneous enhancement of the lesion with bright peripheral enhancement. There were also bilateral acoustic neuromas, which were confirmed by MRI. Screening audiogram showed no hearing loss bilaterally.

Suboccipital craniectomy with upper cervical laminectomy (from C1 to C4) followed by dural incision and midline myelotomy exposed an encapsulated grayish-white mass at 0.5 cm depth. After intratumoural debulking, it was possible gradually to lift and progressively remove the whole sausage-shaped mass by working in the interface between tumour capsule and spinal cord. Peroperative frozen-section analysis revealed a benign lesion with features in favour of schwannoma, so we proceeded to complete surgical excision of tumour. Subsequent detailed histopathological examination of the mass confirmed the diagnosis of spinal cord intramedullary schwannoma.

Postoperatively, the patient showed a progressive recovery and she was able to walk without support 3 months after surgery. Follow-up MRI (Fig. 4) of cervical spine showed complete excision of the

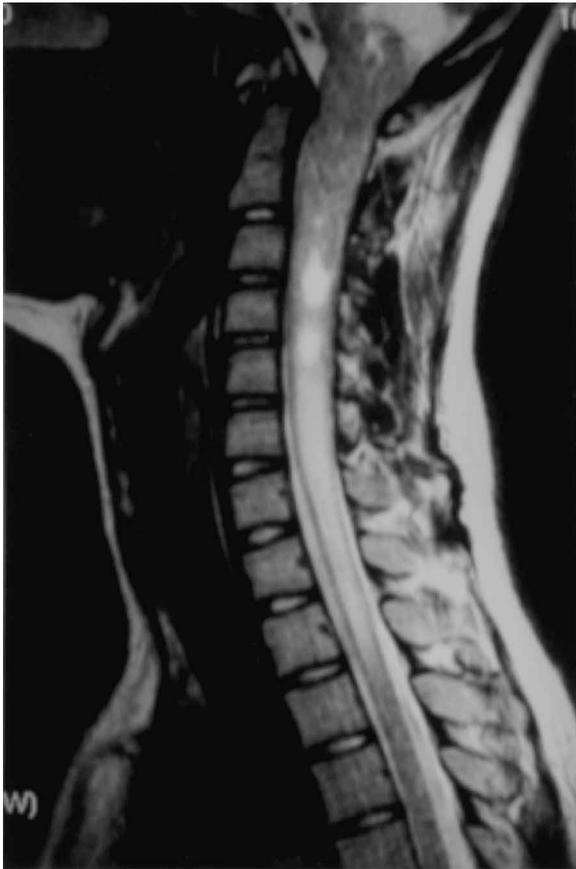


FIG. 1. T2-weighted sagittal imaging showing a mass lesion at craniocervical junction with syringomyelia at its caudal end.



FIG. 2. Contrast-enhanced T1-weighted sagittal imaging showing bright enhancement pattern with well-demarcated tumour periphery. There are also hypointense signals at upper and lower end of mass, possibly due to necrotic areas.

lesion. She is kept under observation for growth of her CP angle tumours.

Discussion

Intramedullary schwannomas constitute 1.1% of all the spinal schwannomas.⁸ Since the earliest report in 1931,⁹ review of reports of all the intramedullary schwannomas up to 1999 showed 57 cases of nerve sheath tumours within the spinal cord.² Nine more cases have been added to the list afterwards including our case.^{6,7,10,11} Among all 66 cases reviewed, intramedullary schwannomas were seen in males/female ratio of 1.5, which remained the same as noted earlier by Drapkin.¹ The ages of patients ranged from 9 to 75 years (mean 41.5). Duration of symptoms ranged from 3 months to 20 years (mean 31.5 months) and varied with the location of the tumour. Intramedullary schwannomas were most frequent in the cervical spinal cord (65%) followed by the thoracic region (22%) and lumbar region (10%).^{1,2,12}

Although most of the cases in the literature were described as solitary intramedullary schwannomas, some of these were also reported occasionally in association with neurofibromatosis (NF).^{4,5} It is noted that there is a trend for NF-2 to harbour intramedullary ependymomas and NF-1 to have

spinal cord astrocytomas.⁴ We found only five cases of intramedullary schwannoma in NF-1 and six cases were seen in association with NF-2.^{4,5,12} Finding intramedullary schwannomas is unusual as the fibres in the central nervous system do not have Schwann cells. The pathogenesis of intramedullary schwannoma is a matter of controversy.^{1,2,9} Hypotheses regarding the origin of intramedullary schwannoma include:

- late neoplastic development of ectopic Schwann cells originating from the embryonic neural ridge during development;
- Schwann cells ensheathing aberrant intramedullary nerve fibres;
- Schwann cells extending along the anterior spinal artery;
- neoplastic growth from Schwann cells at dorsal-root entry zone;
- transformation of pial cells of neuro ectodermal origin into Schwann cells.¹²

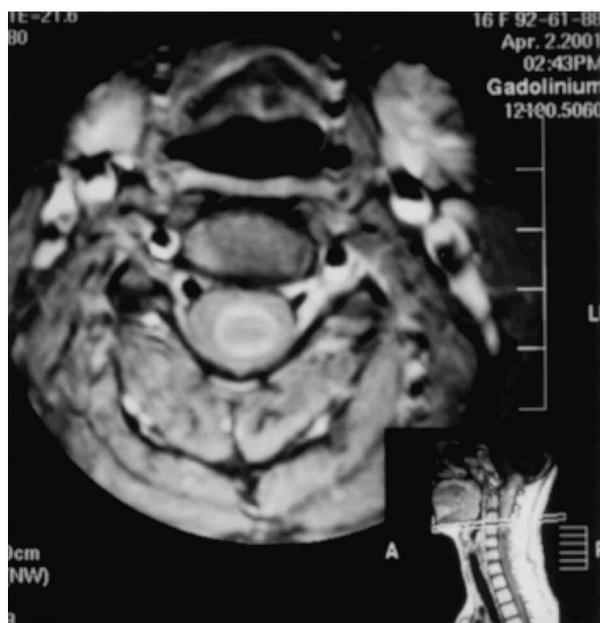


FIG. 3. Postgadolinium T1-weighted axial imaging showing expansion of central spinal cord with contrast enhancement of the mass lesion more marked at the periphery.



FIG. 4. Postoperative contrast-enhanced imaging showing complete resection of tumour mass.

Our surgical findings found dorsal subpial attachment suggestive of origin, probably from the dorsal exit zone of nerve root.

MRI is the most useful method for diagnosis of spinal intramedullary tumours.⁸ Colosima *et al.*³ recently reviewed the findings in intramedullary schwannoma. On T1-weighted images, the tumour

usually gave hypointense signals compared with spinal cord, while in T2-weighted images, the tumour usually showed hyperintense signals, with occasional isointense or low signal areas. The tumour showed marked enhancement on postgadolinium images with well-delineated margins. In our case, there was marked enhancement and the margins were well demarcated. There were small hypointensities at the upper or lower ends of the enhancing mass suggestive of cystic degeneration or necrosis. Differential diagnoses include ependymomas and astrocytomas. Signals intensity and enhancement patterns in both solitary cases (without neurofibromatosis) of intramedullary schwannomas and those in association with neurofibromatosis are similar. Bright contrast enhancement pattern in schwannomas is probably because of open-gap junctions, which are short, straight and patent, communicating freely with relatively large extracellular space. Our case exhibited similar MRI findings, but it was also associated with segmented syringomyelia of cervical spine, which was not described in review of MRI of these tumours.^{7,8}

The aim of surgical intervention in intramedullary schwannoma is complete excision, because of its benign nature.^{4,11} Complete surgical excision of an intramedullary tumour (including Schwannoma) is challenging in neurosurgical practice to avoid exacerbating neurosurgical deficits.^{6,11} Recently, the use of Real-time MRI, CUSA, CO₂-laser along with somatosensory-evoked potentials have revolutionized the sophistication and precision in neurosurgical procedures. Their use in spinal intramedullary tumour is especially helpful to accomplish complete excision.⁶ These efficient tools are not only expensive, but also still not available in many neurosurgical centres like ours. We were able to excise this well encapsulated tumour by using microsurgical techniques only. However, peculiar radiological imaging features and use of preoperative fresh frozen analysis of biopsied tissue were quite helpful, which give enough evidence to know the benign nature of lesion. We think that intramedullary schwannomas can be successfully removed completely due to its soft consistency and well-defined capsular interface. Careful slow progression, intratumoural debulking, good illuminated and clean surgical field, were key to achieve comparable microsurgical results in complete excision of intramedullary schwannomas.

Conclusion

Intramedullary schwannomas are extremely rare in association with NF-2. This is the first reported case located at the craniovertebral junction. Clinical manifestations and radiological imaging features are similar to those of solitary intramedullary schwannomas occurring without neurofibromatosis type-2. Although newer sophisticated neurosurgical techniques have enhanced the precision to achieve complete excision, their cost and availability is a problem.

Microsurgical technique is helpful for achieving complete excision in these benign tumours and can obtain comparable clinical results.

Acknowledgement

I am grateful to my wife, Amna Javaid, for helping me in the preparation of this manuscript.

References

- 1 Drapkin AJ. Intramedullary schwannoma. *J Neurosurg* 1991;75:834–5.
- 2 Binatli O, Ersahin Y, Korkmaz O, Bayol U. Intramedullary schwannoma of spinal cord; case report and review of literature. *J Neurosurg Sci* 1999;43:163–8.
- 3 Colosima C, Cerase A, Denaro L, Maira G, Greco R. Magnetic resonance imaging of intramedullary spinal cord schwannoma; report of two cases and review of literature. *J Neurosurg (Spine 1)* 2003;99:114–17.
- 4 Lee M, Rezai AR, Freed D, Epstein FJ. Intramedullary spinal cord tumors in neurofibromatosis. *Neurosurgery* 1996;38:32–7.
- 5 Patronas JN, Courcoutsakis N, Bromley MC, Katzman GL, MacCollin M, Parry DM. Intramedullary and spinal canal tumors in patients with neurofibromatosis 2: MR imaging findings and correlation with genotype. *Radiology* 2001; 218:434–42.
- 6 Kalangu KK, Couto MT. Radical resection of intramedullary spinal cord tumors without cavitron, ultrasonic aspirator or CO₂ laser: a ‘two stage’ technique. *Surg Neurol* 1996;46:314–16.
- 7 Kodama Y, Terae S, Hida K, Chu BC, Kaneko K, Miyasaka K. Intramedullary schwannoma of the spinal cord: report of two cases. *Neuroradiology* 2001;43:567–71.
- 8 Riffaud L, Morandi X, Massengo S, Carsin-Nicol B, Hersbach N, Guegan Y. MRI of intramedullary spinal schwannomas: case report and review of the literature. *Neuroradiology* 2000;42:275–9.
- 9 Kernohan JW, Terae S, Hida K, Adson AW. Intramedullary tumor of the spinal cord. *Arch Neurol Psychiat* 1931;25:679–99.
- 10 Darwish BS, Balakrishnan V, Maitra R. Intramedullary ancient schwannoma of the cervical spinal cord: case report and review of literature. *J Clin Neurosci* 2002;9:321–3.
- 11 Ogungbo BI, Strachan RD, Bradey N. Cervical intramedullary schwannoma: complete excision using the KTP laser. *Br J Neurosurg* 2000;14:345–8.
- 12 Morimoto T, Sasaki T, Mochizuki T, Takakaru K, Sato A. A thoracic intramedullary neurinoma with multiple intracranial meningiomas: case report. *No Shinkei Geka* 1992;20:423–7.