

Case Report

Giant Ventral Midline Schwannoma of Cervical Spine : Agonies and Nuances

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Pure ventral midline giant schwannoma is an extremely rare entity. Spinal intradural extramedullary schwannomas commonly occur posterolateral or anterolateral to the spinal cord. A case of a pure midline ventrally situated giant pan cervical extramedullary schwannoma in an 18-year-old male patient with compressive myelopathy and sphincter involvement is presented. Spinal MR imaging showed a midline ventrally situated extramedullary tumor with severe spinal cord compression extending from clivus to C7 vertebra. It was resected through a posterolateral approach. Histology was consistent with a schwannoma. Post operative MR imaging showed no evidence of the tumor. The radiological features, pathogenesis and surgical strategies in management of these difficult tumors are discussed and the relevant literature is briefly reviewed.

KEY WORDS : Extramedullary · Intradural · Giant schwannoma · Spine · Ventral location.

INTRODUCTION

Schwannomas are common benign intradural extramedullary spinal neoplasms⁶. These usually present with subacute to chronic compressive myelopathy. Modern magnetic resonance (MR) imaging has helped immensely in their accurate preoperative identification and pathological diagnosis⁷. Advanced instrumentation and microsurgical techniques have made surgical excision safe and radical. Surgical excision of midline ventral long segment intradural tumors can be formidable and can have potentially serious morbidity¹⁹. An appropriate surgical approach and strategy needs to be outlined to achieve a good outcome.

CASE REPORT

An 18-year-old male presented with progressively worsening neck pain and paraesthesias in both upper limbs for the past 3 years. He had progressive stiffness of both lower limbs, difficulty in walking and straining while micturition

and constipation for the past 2 months. He required support while walking. There was no history of trauma, fever or tuberculosis. There were no neurocutaneous markers. There was weakness of spinal accessory nerves bilaterally in form of paresis of trapezius and sternomastoids. On examination, he had spastic quadriparesis of grade 4. Patient had hypertonia and hyperreflexia of all limbs. Plantars were extensor. MRI of the craniovertebral region and cervical spine revealed a midline ventral tumor extending from lower clivus to C7 vertebra. It was hypointense on T1 and hyperintense on T2-weighted image with irregular thick peripheral enhancement (Fig. 1). The spinal cord was severely compressed and displaced posteriorly. A C1-C7 laminotomy was performed. All lateral masses were preserved. The tumor was midline ventral and the spinal cord was stretched out like a ribbon. The ligamenta denticula were cut bilaterally. All roots were safeguarded. The tumor was yellowish-grey, lobulated, vascular and had a fleshy capsule and suckable. The dissection was commenced from C7 and carried upwards. The tumor was adherent to the ventral surface of the spinal cord in lower part but no definite attachment of the tumor to any nerve root could be found. A radical excision was achieved. The cord was seen to pulsate well at the end. Histological examination revealed features of schwannoma with extensive degenerative change (Fig. 2). Immediately following surgery, spasticity and paresis of all four

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limbs improved. Sphincteric complaints also showed delayed improvement. Post-operative MR showed no evidence of residual tumor (Fig. 3). At follow-up after 1 year patient is doing well.

DISCUSSION

Intradural spinal schwannomas are usually dorsal, lateral or dorsolateral in position⁶. However, as much as 23% of cervical nerve sheath tumors may have an anterolateral component consistent with ventral root origin⁸. Giant spinal schwannomas are defined as those that extend over more than two vertebral levels. Giant spinal schwannomas in ventral midline location are rare^{10,13,15,21}. We report the case of a pure midline ventral cervicodorsal intradural extramedullary schwannoma extending from clivus to C7. The origin of schwannoma may be difficult to judge in long segment schwannomas, as observed in our case. The location of the tumor in our case suggests an alternative site of Schwann cell derivation. Multiple theories exist to explain this aberrant location for a Schwann cell-based tumor. Some authors postulate that these arise from perivascular nerve plexi surrounding penetrating spinal cord vessels from the anterior spinal artery^{2,17,18}. Alternatively, rests of neural crest progenitors may migrate improperly into the central nervous system parenchyma during embryogenesis^{2,12,18,20}. We hypothesize that genesis of a midline ventral schwannoma without attachment to a ventral root requires an atypically located parent Schwann cell, such as those of the nervi vasorum of the anterior spinal artery^{2,17}. Singer et al.¹⁸ have reported a ventrally placed pigmented schwannoma of the C7-T2 region arising from the leptomeninges in the region of the anterior median septum, ventral to the dentate ligaments and anterior rootlets.

Various approaches to access the ventral cervical and cervicothoracic intradural tumors including schwannomas are : 1) anterior cervical or transthoracic approaches^{3,16}, 2) anterolateral approach¹, 3) posterolateral approach^{6,11,13}, and 4) endo-assisted posterolateral approach⁴. In most cases, there is no single approach that provides adequate surgical exposure across multiple spinal levels. O'Toole and McCormick¹⁶ described anterior corpectomy and reconstruction for solid and small schwannoma apposed to the ventral midline surface of the spinal cord producing no significant rotation of the spinal cord and therefore presenting no tumor surface for lateral surgical access. Anterior approach has certain disadvantages like 1) inadequate access

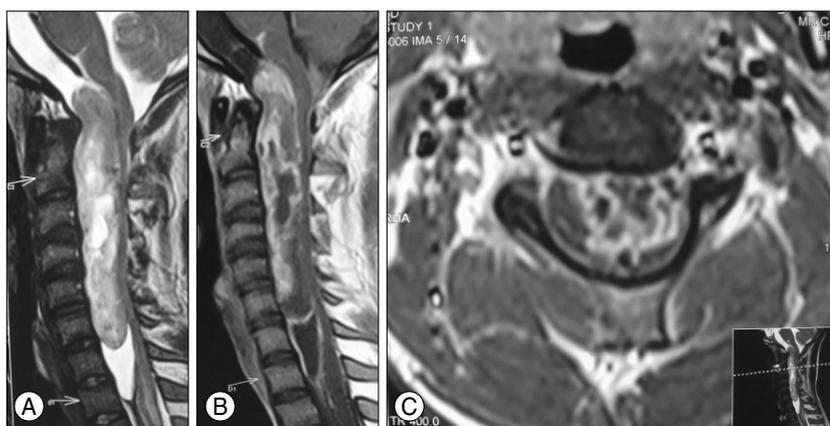


Fig. 1. MRI showing ventrally situated extramedullary schwannoma extending from lower clivus to C7. A : Sagittal T2-weighted image. B : Sagittal T1- contrast enhanced image. C : Axial T1-weighted contrast enhanced image.

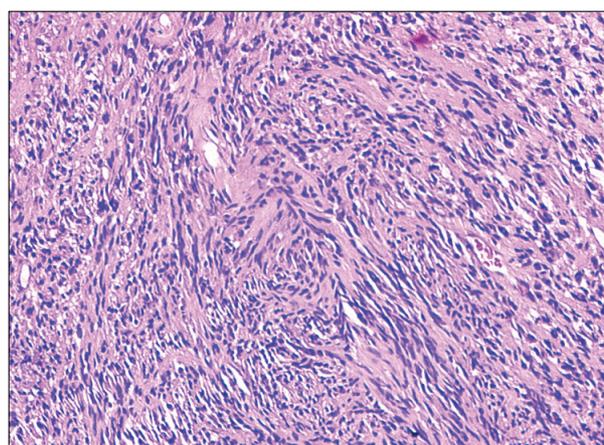


Fig. 2. Microphotograph showing densely packed spindle cells in interlocking fascicles (Antoni type A), intermingled with loosely textured tissue with extracellular clear spaces (Antoni type B). H&E, original magnification $\times 200$.

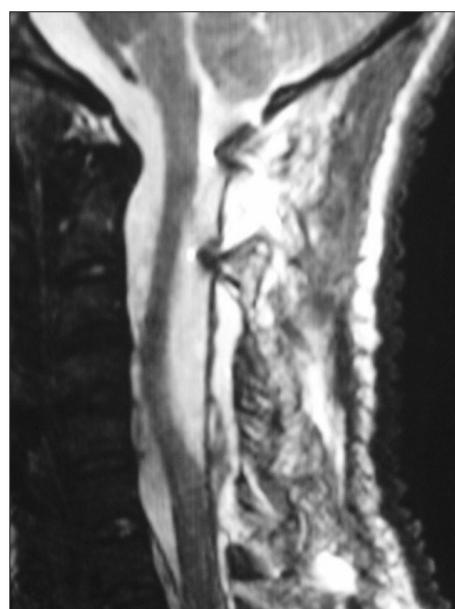


Fig. 3. Sagittal T2-weighted post-operative Modern magnetic resonance image showing no evidence of tumor.

due to deep and constrained field of view, 2) excessive bleeding from epidural venous plexus, 3) need for ensuring spinal stability and meticulous bony reconstruction, and 4) postoperative CSF fistula¹⁶. However, Acosta et al.¹ described anterolateral corridor using modified paramedian transpedicular approach and spinal reconstruction for ventral intradural tumors including schwannomas of the cervical and cervicothoracic spine. Acosta et al.¹ state that removal of the posterolateral elements, and the dorsal vertebral body combined with angulation of the operating table exposes the ventral-most thecal sac and creates a potential space for delivery of tumor without spinal cord retraction or rotation. This approach avoids the difficulties of anterior approach¹.

Traditionally, the approach for resection of intradural spinal tumors has been by posterior laminectomy with or without facetectomy and/or pediclectomy. This is largely a reflection of the posterior and lateral orientation of most intradural tumors. Extensive bone excision and reconstruction is avoided in this approach¹³.

Barami and Dagnew⁴ reported endoscopic assisted removal of ventral intradural spinal tumors through a posterior approach. They postulate that judicious use of endoscope during tumor removal allows minimal retraction of the cord and inspection of the corridors which are difficult to visualize with the microscope. We believe that an endo-assisted posterolateral approach might be the best option for such difficult tumors in near future.

We believe that these tumors form a special subset of schwannomas with different origin and presentation. These manifest with myelopathy rather than radiculopathy due to aberrant origin, therefore present after achieving giant size unlike simple schwannomas originating from roots. Literature on consistency of giant ventral schwannomas is lacking due to very rare occurrence. We have realized that giant ventral schwannomas have softer consistency due to extensive degenerative changes (might be due to relative ischemia caused by huge size) which facilitates the fall of the tumor into the vision after decompression. Surgical approach is determined primarily depending on the location of tumors in spinal canal. However, tumor consistency and pathology should be also considered. Solid ventral midline schwannomas are optimally managed by anterior and anterolateral approaches^{1,3,16}. However; tumors with softer consistency may be approached from posterolateral approach with satisfactory outcome as evidenced by our case.

On preoperative MR imaging, schwannomas usually show hypointense or isointense signal on T1-weighted and hyperintense on T2-weighted images^{6,7}. Homogenous enhancement is common but occasional heterogeneous enhancement revealing intratumoral hemorrhage, cyst or necrosis

is seen suggesting an atypical or aggressive nature^{6,7}. Meningioma and tuberculous granulomas are important differential diagnosis^{5,14}. The hyperintense signal on T2-weighted MR images was suggestive of a relatively soft tumor consistency. MR imaging is helpful to indicate consistency of the tumor, thereby suitability of the posterolateral approach.

A piecemeal decompression bilaterally in between the nerve roots allowed the tumor to express into the operative field and relieved the cord compression in our case. Gentle and slow micro dissection, minimal use of cautery and no retraction of cord parenchyma are the keys to the successful surgery. The use of cautery was kept minimal to avoid thermal injury to the roots and cord. Preservation of roots and lateral masses is essential for long term good functional outcome. The upper third of the tumor was free in the subarachnoid space while the tumor in the middle and the lower third was adherent to the ventral surface of the spinal cord suggesting a caudocranial growth of the tumor.

Laminoplasty reduces the risk of post-operative kyphosis⁹. However, Kim et al.¹⁰ have emphasized that despite of multilevel laminoplasty for huge craniocervical cord tumor the patient may develop swan neck deformity. To prevent this kind of complication, they stressed anterior cervical fixation in addition to posterior laminoplasty or laminectomy. In our patient, although there was no immediate cervical spinal instability, we are closely monitoring the patient. Long term follow up is essential for such complication.

CONCLUSION

Pure midline ventral giant schwannomas are very rarely encountered. These manifest late with myelopathy after achieving giant size due to aberrant origin and location. Surgery for these tumors is potentially formidable. A posterolateral approach with piecemeal micro dissection is feasible in such tumors. It remains unclear whether these schwannomas represent a special variant of schwannomas or an entirely distinct entity. Larger series may answer this question in the future.

References

1. Acosta FL Jr, Aryan HE, Chi J, Parsa AT, Ames CP : Modified paramedian transpedicular approach and spinal reconstruction for intradural tumors of the cervical and cervicothoracic spine : clinical experience. *Spine (Phila Pa 1976)* 32 : E203-E210, 2007
2. Adelman LS, Aronson SM : Intramedullary nerve fiber and Schwann cell proliferation within the spinal cord (schwannosis). *Neurology* 22 : 726-731, 1972
3. Banczerowski P, Lipóth L, Vajda J, Veres R : Surgery of ventral intradural midline cervical spinal pathologies via anterior cervical approach : our experience. *Ideggyogy Sz* 56 : 115-118, 2003
4. Barami K, Dagnew E : Endoscope-assisted posterior approach for the

- resection of ventral intradural spinal cord tumors : report of two cases. **Minim Invasive Neurosurg** 50 : 370-373, 2007
5. Clarke MJ, Krauss WE : Cervical meningioma masquerading as schwannoma : two cases and a review of the literature. **J Neurosurg Sci** 50 : 41-44, 2006
 6. Conti P, Pansini G, Mouchaty H, Capuano C, Conti R : Spinal neurinomas : retrospective analysis and long-term outcome of 179 consecutively operated cases and review of the literature. **Surg Neurol** 61 : 34-43; discussion 44, 2004
 7. De Verdelhan O, Haegelen C, Carsin-Nicol B, Riffaud L, Amlashi SF, Brassier G, et al. : MR imaging features of spinal schwannomas and meningiomas. **J Neuroradiol** 32 : 42-49, 2005
 8. el-Mahdy W, Kane PJ, Powell MP, Crockard HA : Spinal intradural tumours : Part I--Extramedullary. **Br J Neurosurg** 13 : 550-557, 1999
 9. Katsumi Y, Honma T, Nakamura T : Analysis of cervical instability resulting from laminectomies for removal of spinal cord tumor. **Spine (Phila Pa 1976)** 14 : 1171-1176, 1989
 10. Kim SH, Doh JW, Yoon SM, Yun IG : Huge schwannoma extended from craniocervical junction to upper thoracic spine. **J Korean Neurosurg Soc** 34 : 589-592, 2003
 11. Kobayashi Y, Mochida J, Nishimura K, Nomura T, Miyazaki S : Intradural extra-arachnoid schwannoma of the upper cervical spine. **Spinal Cord** 36 : 859-863, 1998
 12. Kodama Y, Terae S, Hida K, Chu BC, Kaneko K, Miyasaka K : Intramedullary schwannoma of the spinal cord : report of two cases. **Neuroradiology** 43 : 567-571, 2001
 13. Mahore A, Muzumdar D, Chagla A, Goel A : Pure ventral midline long segment schwannoma of cervicodorsal spine : a case report. **Turk Neurosurg** 19 : 302-305, 2009
 14. Mirzai H : Tuberculoma of the cervical spinal canal mimicking en plaque meningioma. **J Spinal Disord Tech** 18 : 197-199, 2005
 15. Nagasawa S, Ohta T, Kajimoto Y, Aoki J, Onomura T, Miyaji Y : Giant neurinoma occupying the holocervical and upper thoracic regions : case report. **Surg Neurol** 42 : 157-159, 1994
 16. O'Toole JE, McCormick PC : Midline ventral intradural schwannoma of the cervical spinal cord resected via anterior corpectomy with reconstruction : technical case report and review of the literature. **Neurosurgery** 52 : 1482-1485; discussion 1485-1486, 2003
 17. Riggs HE, Clary WU : A case of intramedullary sheath cell tumor of the spinal cord; consideration of vascular nerves as a source of origin. **J Neuropathol Exp Neurol** 16 : 332-336, 1957
 18. Singer RJ, Clough J, Johnson M, Atkinson JB, Copeland M : Pigmented schwannoma of the ventral spinal cord. **South Med J** 92 : 532-534, 1999
 19. Slin'ko EI, Al-Qashqish II : Intradural ventral and ventrolateral tumors of the spinal cord : surgical treatment and results. **Neurosurg Focus** 17 : ECP2, 2004.
 20. Solomon RA, Handler MS, Sedelli RV, Stein BM : Intramedullary melanotic schwannoma of the cervicomedullary junction. **Neurosurgery** 20 : 36-38, 1987
 21. Sridhar K, Ramamurthi R, Vasudevan MC, Ramamurthi B : Giant invasive spinal schwannoma : definition and surgical management. **J Neurosurg** 94 : 210-215, 2001