

Giant Cervico-Dorsal Spine Ectopic Intradural Schwannoma Placed Dorsally Without Attachment to the Nerve Root: Review of Literature

Guru Dutta Satyarthee*, Shikha Satyarthee

The Department of Neurosurgery, All India Institute of Medical sciences, New Delhi, India

Abstract

Spinal intradural schwannoma without attachment to the spinal nerve root is very rare. About six such cases of extra-axial schwannoma without attachment to nerve roots were reported in the literature. Author reports an interesting case of giant cervico-dorsal intradural schwannoma, which was neither attached to the spinal nerve roots nor to the dura or spinal cord observed during the surgery. Patient had unremarkable recovery following the surgery. To the best knowledge of the authors, current case is the second cases describing giant sized cervico-dorsal schwannoma located dorsal to spinal cord in the western literature till date. Briefly pathogenesis, management is reviewed.

Keywords

Giant Cystic Schwannoma, Cervico-Dorsal, Management, Attachment, Ectopic

Received: September 15, 2015 / Accepted: November 20, 2015 / Published online: January 17, 2016

© 2016 The Authors. Published by American Institute of Science. This Open Access article is under the CC BY-NC license.

<http://creativecommons.org/licenses/by-nc/4.0/>

1. Introduction

Schwannomas are benign tumor of peripheral nerve sheath. [1-5] It accounts for 25% of spinal tumors. [6, 7] Although schwannomas are usually solid tumors, they occasionally exhibit necrosis and cystic degeneration. [7] Schwannoma not connected to nerve root is very rare. [1-5] These are mostly extra-axial located in spinal column, peripheral nerve and rarely along cranial nerves. It was cystic and placed posteriorly to the cord and no attachment to the nerve root was demonstrated during microneurosurgical excision.

2. Case Report

A-42-year old woman presented with six months history of weakness and numbness involving all limbs. However, no spinal deformity or neurocutaneous markers of neurofibromatosis were present. Neurological examination showed grade 3-4 power in all limbs. Sensory examination

revealed a graded sensory loss below C4 dermatomes. Reflexes were brisk in both upper and lower limbs with planter extensor response. Magnetic resonance imaging showed a large cystic intradural extra-medullary lesion lying posterior to spinal cord extending rostrally third cervical vertebra body level to caudal third thoracic vertebral body level. It was showing hypointense signal intensity on T1 weighted magnetic resonance imaging [Fig-1] and hyperintense on T2 weighted images, [Fig-2] causing displacement of compressed spinal cord anteriorly and cord was deformed and pushed to the left side of the dural sac. [Fig-3, 4] No additional lesions were identified. The differential diagnosis based on imaging findings included arachnoid cyst. However, on imaging appearance and location, a metastatic or meningioma is less likely possibility.

Treatment:

The patient underwent C2 to D4 osteoplastic laminoplasty. The dura was opened longitudinally in the midline and a greyish mass lesion was observed. The tumor was resected

* Corresponding author

E-mail address: duttaguru2002@yahoo.com (G. D. Satyarthee)

successfully en block. It was not attached to either nerve root, spinal cord or dura. The postoperative period was uneventful with no fresh neurological deficit in the immediate postoperative period. Histopathological examination of specimen revealed schwannoma Antonio type B. At five month following surgery, she had marked improvement in muscle power and recovery of sensation. At last follow-up 1 year after surgery, she had no deficit.



Fig. 1. MRI sagittal section, T1WI of cervical and dorsal spine showing hypointense lesion located in the intradural compartment extending from C3 cervical vertebrae to the upper dorsal spine.



Fig. 2. MRI sagittal section, T2WI of cervical and dorsal spine showing hyperintense mass lesion located in the intradural compartment displacing of compressed spinal cord anteriorly and rotation of cord towards the left side of the dural sac.

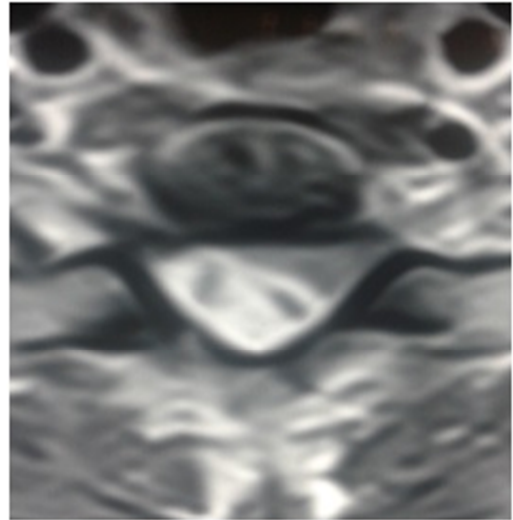


Fig. 3. MRI axial section, T1WI of cervical showing hyperintense mass lesion located in the intradural compartment displacing of compressed spinal cord anteriorly and rotation of cord towards the left side of the dural sac.

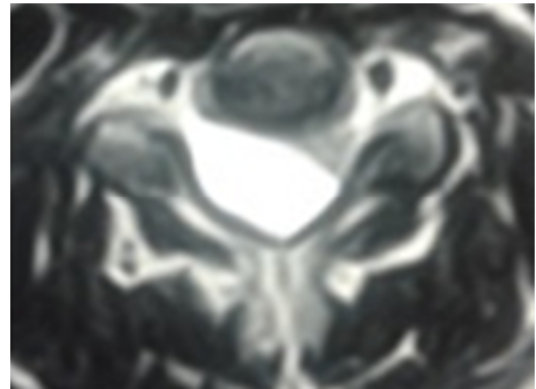


Fig. 4. MRI axial section, T2WI of cervical showing hyperintense mass lesion located in the intradural compartment displacing of compressed spinal cord anteriorly and displacement of cord towards the left side of the dural sac.

3. Discussion

Schwannoma approximately constitute approximately 20% of intraspinal tumor. [8] These are typically intradural extramedullary neoplasm derived from Schwann cells of nerve sheath. The vast majority of spinal schwannoma arises from dorsal and much less frequently ventral nerve roots and therefore located eccentrically in dorsolateral or ventrolateral position in the spinal canal. [7] Rarely schwannoma may arise within the spinal cord itself, indicating an anomalous site of Schwann cell origin. [9, 4-15] The parent nerve of spinal schwannomas is predominant dorsal sensory nerve roots. However, as much as 23% of cervical nerve sheath tumor may have an anterolateral component consistent with ventral root origin. Rarely schwannoma may be located in intramedullary location. [8]

Many hypotheses are made to explain ectopic origin of schwannomas, which were not attached to the nerve roots.

Some authors postulated these schwannomas arise from perivascular nerve plexus surrounding penetrating spinal cord vessels from the anterior spinal artery. [9] Alternatively, neural crest progenitors rest may migrate improperly into the central nervous system parenchyma during embryogenesis. [10] Genesis of midline ventral intradural schwannoma without attachment to nerve root can be explained on the basis of atypically located parent Schwann cells, such those of nervi vasorum of posterior spinal arteries. [7]

Current case is interesting as schwannoma was located in midline, lying posterior to the spinal cord. However, only two case reports exist in the literature with reporting the dorsally located midline schwannoma. [14-15] It was entirely cystic, which is much rare occurrence. Regarding ectopic origin of intraparenchymal schwannoma can be explained as following theories. Redekop supported the theory of distorted embryogenesis [11] while Rigg and Clary postulated its origin could be related to proliferation of schwannoma cells in the perivascular plexus. [12]

Our case is unique with regard to exceptional large size of cystic schwannoma extending from C3 to D3 vertebral body level. Such large size cystic schwannoma was only comparable to two previously reported by Palma et al [5], and Nagańska Nagasaki et al [6] However, our case had giant size located in the midline lying posterior to the spinal cord Nagańska et al. reported a case of a giant schwannoma in a 45-year female. Spinal MRI revealed the presence of an intradurally located, extramedullary lesion lying over lower border of C4 and extending to T4 vertebral body . It was filling spinal canal and also causing compression and distortion of the spinal cord. Nagańska et al emphasized importance of the early diagnosis and early management can provide satisfactory neurological outcome. The current case was similar to those, reported by Nagańska et al. [6]

Palma and Mariottini reported a large cystic ectopic Schwannoma lying anteriorly to spinal cord and extending from the pontomedullary cistern to the upper dorsal spine. The current case was similar to case reported Palma and Mariottini [5]. Our case constituted second giant cystic schwannoma lying dorsal to the spinal cord in the western literature.

References

- [1] George B, Lot G, Velut S: Pathologie tumorale du foramen magnum Neurinome. *Neurochirurgie* 1993; 39: 43-49.
- [2] Goebel NH, Shimkawa K, Schaake, et al: Schwannoma of the sellar region. *Acta Neurochir* 1979; 48: 191-197.
- [3] Bollati G, Galli G, Gandolfini M, et al: spinal intradural Schwannoma without attachment to nerve root. *J Neurosurg* 1982; 57: 701-702.
- [4] Goel A, Bhayani R, Nagpal RD: Unattached intracranial extra-axial schwannoma. *Br J Neurosurg.* 1996; 0: 405-407.
- [5] Palma L, Mariottini A: Cystic ectopic Schwannoma extending anteriorly from the pontomedullary cistern to the thoracic spinal cord: Case illustration. *J Neurosurg (Spine)* 2003; 8: 113.
- [6] Nagańska E, Matyja E, Mossakowski Z, Zabek M.: Giant cervico-thoracic Schwannoma with long clinical history. Case report. *Folia Neuropathol* 1999; 37; 85-88.
- [7] O, Toole JE, Mc Cormick PC. Midline ventral intradural schwannoma of the Cervical spinal cord resected via anterior corpectomy with reconstruction: technical case report and review of literature. *Neurosurgery.* 2003;52(6): 482-6
- [8] Mc Cormick Pc, Post KD, Stein BM Intradural extramedullary tumors in adults. *Neurosurg Clinic N Am* 1990; 1: 592-608.
- [9] 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: 321-323.
- [10] Singer RJ, Clough J, Johnson M, Atkinson JB, et al. Pigmented schwannoma of the ventral spinal cord. *South Med J.* 1999; 92: 532-534.
- [11] Redekop He, Elisevich K, Gilbert J. Fourth ventricular schwannoma. *J Neurosurg* 1990; 73; 771-781.
- [12] Riggs He, Clary WU. A case of Intramedullary sheath cell tumour of the spinal cord. Consideration of vascular nerve as a source of origin. *J Neuropathol Exp Neurol* 1957; 16: 332-336.
- [13] Prakash B, Roy S, Tandon PN. Schwannoma of the brain stem: case report. *J Neurosurg* 1980; 53: 121-123.
- [14] Fischer G, Brochi J. Intramedullary spinal cord tumours. *Stalwarts.* Thieme. 1996 pp 80-81.
- [15] Herregodts P, Vloeberghs M, Schmeding E, Goosens A, Standnik T, D' Haens J. solitary dorsal intramedullary schwannoma. *J Neurosurg* 1991; 4: 816-820.