Giant Cervical Intraspinal Schwannoma: A Case Report

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Citation

R Prasad, M Hoda, K Singh, V Sharma. *Giant Cervical Intraspinal Schwannoma: A Case Report*. The Internet Journal of Spine Surgery. 2006 Volume 3 Number 1.

Abstract

Nerve sheath tumors are one of the most common intraspinal tumors. They constitute approximately 90% of all intradural extramedullary tumors together with meningioma [1]. They are derived from both Schwan cells and fibroblasts forming a mass that encases mainly dorsal sensory nerve root. They may be multiple and are seen in association with neurofibromatosis. We report a case of giant extradural and intradural extramedullary schwannoma of cervical region extending from Axis (C2) to first thoracic vertebrae (T¬1).

CASE REPORT

A 20-year old man had weakness in right lower and upper limb, difficulty in walking and pain in neck 3 years back with involvement of left half of body for 3 months.

The weakness was gradual in onset which first appeared in right lower limb in distal group of muscles followed by involvement in proximal muscles. Gradually his grip of right hand became weak. Weakness was progressive and extended to left upper and lower limb for 3 moths. He became bed ridden at the time of admission.

Pain was mild to moderate in upper back, used to increase in supine position. Patient had history of constipation, urgency and hesitancy for 3 months. There was no history of tingling, parasthesia, trauma and tuberculosis.

Motor examination revealed muscle atrophy in right hand with no fasciculation. Power was 3/5 in proximal joints in upper limb bilaterally. He was not able to hold objects bilaterally. There was significant spasticity in both lower limbs with normal bulk. He was not able to move digs and feet. Sensory examination revealed diminution of pain and touch from C_3 dermatome downward. Posterior column sensations were absent. All superficial reflexes were absent and plantar reflexes were extensor bilaterally. Deep tendon reflexes were brisk in lower limbs and decreased in upper limbs. There was no spinal tenderness or deformity. There was no subcutaneous nodules or café-au-lat spots.

Magnetic resonance imaging of cervical spine showed C_5-C_6 extradural and C_2-T_1 intradural extramedullary space occupying lesions which was isointance in T1 weighted image (fig. 1) and hyperintence on T2 weighted image (Fig. 2) with homogenous contrast enhancement (Fig. 3).

Figure 1

Figure 1: T weighted image of MRI cervical spine sagittal view showing isointense intradural intramedullary mass lesion extending from C-T and pushing the cord laterally.

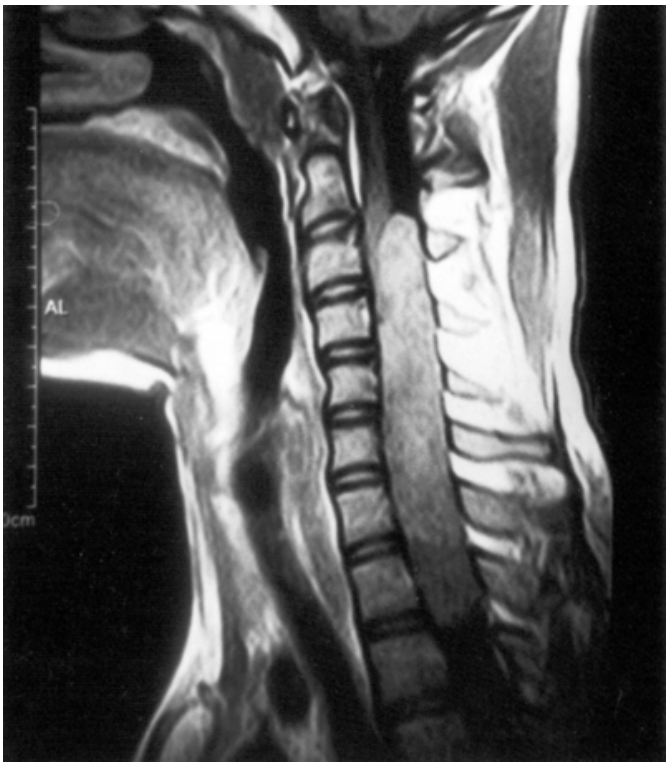


Figure 2

Figure 2: T weighted image of MRI cervical spine sagittal view showing hyperintense intradural intramedullary mass lesion extending from C-T and pushing the cord laterally.

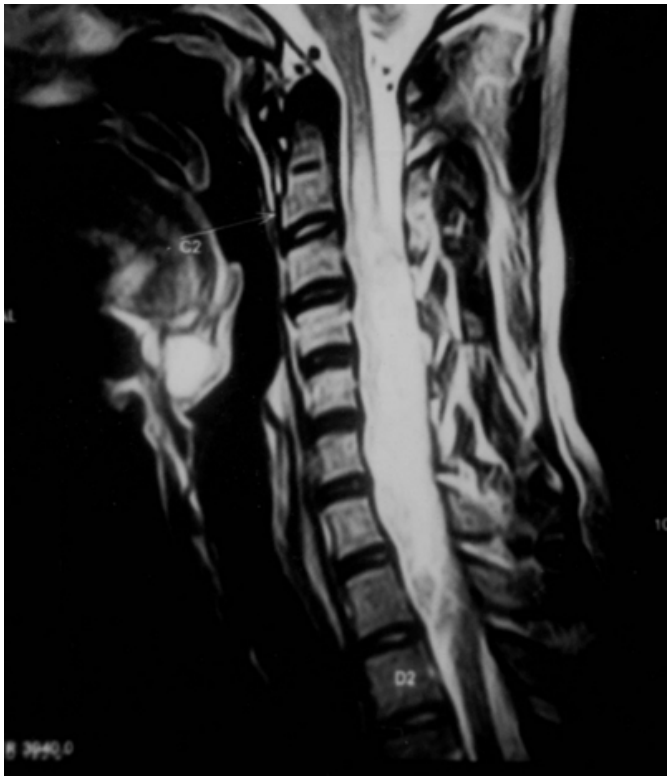
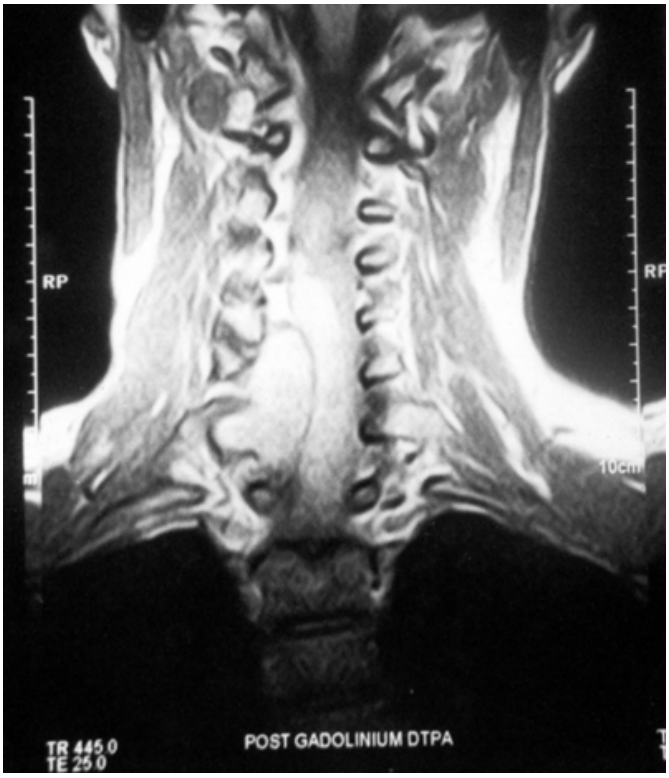


Figure 3

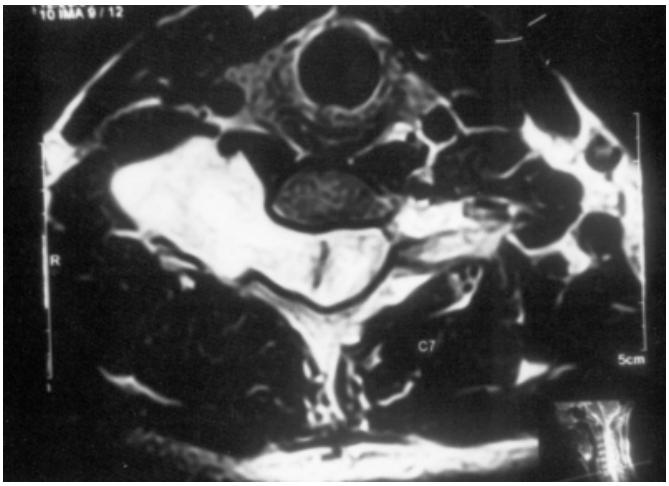
Figure 3: GdDTPA enhanced coronal T weighted MR image of cervical spine showing enhancing mass lesion extending from C-T and going into extraspinal space.



The lesion was also extending extraspinal through enlarged right C_5-C_6 intervertebral foramen (Fig. 4).

Figure 4

Figure 4: Axial T weighted MR image of C vertebra shows a hyper intense dumbbell lesion pushing the cord to opposite side.



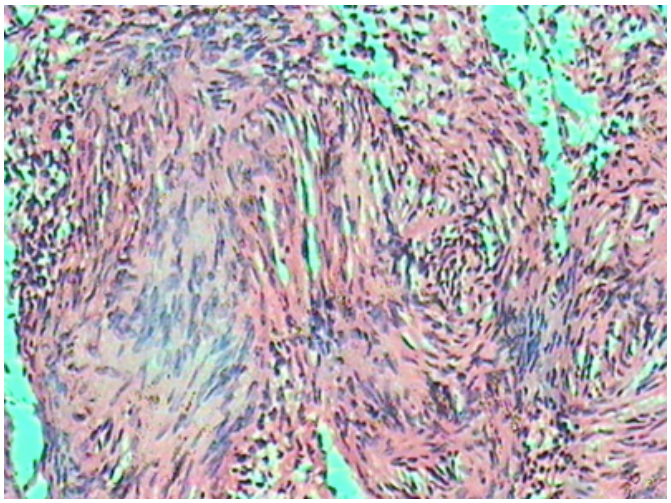
Total excision of tumour through C_2-T_1 lamminectomy was done in prone position. Tumour was capsulated, arising from right C_6 root extending extardural and extraspinal with 12 x

2.5 cm intradural component on right side displacing the cord laterally. Patient showed neurological improvement on first postoperative day and started walking with support at discharge. Postoperative period was uneventful. He regained near total power in first follow up checkup after 3 weeks.

Histopathological examination was suggestive of Schwannoma Antoni type A with typical varrocay body (Fig. 5).

Figure 5

Figure 5: Schwannoma showing antoni type A area with typical varocay body (H&E x 100).



DISCUSSION

Giant intraspinal nerve sheath tumors are rarely reported in literature. Ejelhoff et al in 1992 reported large intraspinal neurofibroma in lumbar region [2]. In 2003 Garg S et al reported multiple cervical neurofibroma extending from occiput to C_6 in a 10 year old boy who presented with quadriplegia [3]. Extensive laminectomy was done to relieve compression.

Our patient had quadreparesis with bladder bowel involvement due to large extradural and intradural extramedullary neurofibroma in cervical canal extending from $C_2 – T_1$. C_2-T_1 laminectomy was required to excise the tumor and for adequate decompression.

Nevertheless, spinal nerve sheath tumor is relatively common in neurofibromatosis. Thakkar et al (1999) found spinal tumor in 65% of 54 neurofibromatosis 1 patients examined with MRI [4]. The tumor was more common in symptomatic than asymptomatic patients and different level of spine were similarly effected. Tatagiba et al (1994) described that cervical area is most commonly affected followed by thoracic and lumbar area [5].

Neurofibroma are under one of the two main category of nerve sheath tumor the second one being schwannoma. Neurofibroma is a separate entity derived from Schwan cells and fibroblast, in contrast schwannoma are composed solely of schwan cells and tends to appear lobulated rather than fusiform tumors as in neurofibroma [1].

Krandsdorf and Murphy found MR imaging differentiation between schwannoma and neurofibroma to be difficult except in large tumors [6].

On MR imaging schwannoma are isointense in 75% cases on T_1 weighted image (25% are hypointense) and hyperintense on T_2 weighted image. Intense homogenous enhancement is seen with contrast. Schwannoma is usually solitary but may be multiple in neurofibromatosis 2. Most commonly seen in forth decade of life, sex distribution is equal and distributed uniformly among the cervical, thoracic and lumbar region. The majority of lesions are intradural extramedullary (70-75%) followed by extradural (15%) and dumbbell shaped with both extradural and intradural component (15%) [1].

Metastasis is other differential diagnosis of an intradural extramedullary lesion. They may arise from primary tumor of CNS (germinoma, penial tumor, ependymoma and medulloblastoma) or from outside the CNS (carcinoma breast, lung, melanoma and leukemia). Metastasis may be differentiated from nerve sheath tumor and meningioma on the basis of their location, appearance and number. The lumbosacral subarachnoid space is the most frequent site and/or multiple in number. Furthermore, the presence of dumbbell appearance would be against metastasis [7,8].

Other intradural extramedullary tumors are rare. Paraganglioma may present as intraspinal tumor that are most commonly localized in cauda equina and filum terminale. These lesions are also isointense on T_1 and hyperintense on T_2-weighted image. However, other factors may be taken into account for correct diagnosis [1,8].

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