

CASE REPORT OF HUGE CERVICAL DUMBBELL TUMOR IN NEUROFIBROMATOSIS. Give up surgery?

Joehalimy, J, Ahmad Sabri.O, Mohammad Anwar Hau Department of Orthopaedic ,Hospital Raja Perempuan Zainab 2, Kota Bharu, Kelantan

Mohd Ariff Sharifuddin Department of orthopaedic , Universiti Sains Malaysia Kubang Kerian, Kelantan

Introduction

A dumbbell-shaped lesion is a solitary tumor that is constricted as it exits the neural foramen. The constriction gives the neurofibroma the appearance of a dumbbell that is used by weight lifters. Neurofibromas that arise from the spinal canal may be intradural or extradural and are most commonly seen at the cervical and thoracic level.

The intraspinal portion of the tumor

may cause spinal-cord compression and nerve root failure. The dumbbell tumors are seen most commonly in the cervical spine (44%), followed by the thoracic spine (27%), and the lumbar spine (21%).

The resection of huge dumbbell tumors raises several problems, including preservation of the cervical nerve root, control of the vertebral artery, and maintenance of spine motion-curvature.

Case report

14 year old boy , known case of neurofibromatosis type 1. Patient presented with progressive weakness of upper and lower limbs (quadripareisis) for 3 months prior to admission. No history of constitutional symptoms.

On examination noted patient to have upper motor neuron lesion at level C3 downward.

Right neck swelling of 5.0cm in size at submandibular region.

An initial clinical evaluation showed a severe cervical compressive myelopathy and also a large 7cm diameter palpable mass on the right side of neck. MRI of Cervical Spine and Neck showed an intraspinal Extradural and extraspinal (paraspinal) tumour on the right side of the neck pushing the spinal cord to the right side. The hourglass shaped tumour also extended outwards through the Intervertebral Foramen (canal through which the nerve exits to supply the limb), reaching in front of his neck.



Discussion

Tumor removal by a lateral approach still carries a risk of injuring not only VA but also the phrenic, vagus, accessory, or hypoglossal nerves.

The posterior approach is a classic standard technique for intraspinal lesions.² Advantages of a combined posterior and anterior approach for resection of a cervical

spinal cord tumor extending into or through the foramen have been described by several authors.¹⁵⁻¹⁷ McCormick¹⁸ reported 12 patients with cervical spine

Dumbbell tumors who underwent resection via a posterior midline approach including partial laminectomy and complete unilateral facetectomy.

He considered these procedures more familiar to surgeons than the anterolateral approach and also emphasized that posterior exposure provided extensive intraspinal access for adequate exposure of large intradural tumor components.

We have chosen to use combined anterior and posterior surgical approach

Conclusion

Surgical enucleation of Dumbbell cervical tumor in neurofibromatosis give good prognosis of the recovery despite of the severity of neurology of initial clinical presentation.

2 stages operation provide safe surgery and minimal morbidity to patient.

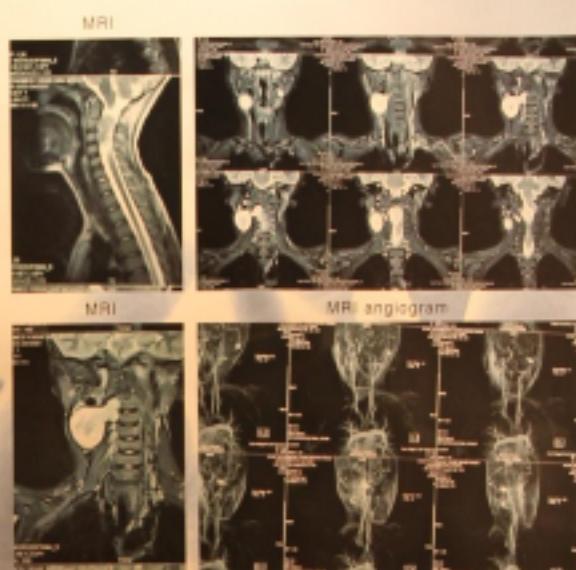
References

- Aszkenasy T, Yossefai T, Hirshberg M, et al. Surgical strategy for cervical dumbbell tumors based on a three-dimensional classification. *Spine* 2003;28:E10-4.
- O'Toole JE, McCormick PC. Malignant intradural schwannoma of the cervical spinal cord resected via anterior approach with reconstruction: Technical case report and review of the literature. *Neurosurgery* 2003;52:1482-8. discussion 1489-8.
- George B, Leff B. Surgical treatment of dumbbell neurinomas of the cervical spine. *Crit Rev Neurosurg* 1998;8:156-80.
- Gutmann DH, Drury JD, Scheithauer MH. Surgical approaches to spinal schwannomas. *Contemp Neurosurg* 2002;27:1-8.
- Ketkamp J, Samii M. Surgery of spinal nerve sheath tumors with special reference to neurofibromatosis. *Neurosurgery* 2000;42:279-89. discussion 290-290.
- Leff B, George B. Cervical neurinomas with extradural component: surgical management in a series of 87 patients. *Neurosurgery* 1997;41:813-22.
- Bilbao A, Karkas S, Canalejo E, et al. Review of 36 cases of spinal cord meningioma. *Spine* 2003;28:727-31.
- McCormick C. Management of dumbbell tumors of the cervical spine. *Neurosurgery* 1985;38:294-300.
- el-Mahdy W, Kass PJ, Pecetti MP, et al. Spinal intradural tumors: Part I—Extramedullary. *Br J Neurosurg* 1999;13:590-7.
- Guzan F, Karakas S, Canalejo E, et al. Review of 36 cases of spinal cord meningioma. *Spine* 2003;28:727-31.
- Borrelli J, Baudier S, Biagioli R, et al. Chordoma at the mobile spine: 15 years of experience. *Spine* 2005;31:499-503.

1st stage surgery (posterior approach)

Surgery was done combined posterior and anterior approach with 2 stages surgery.

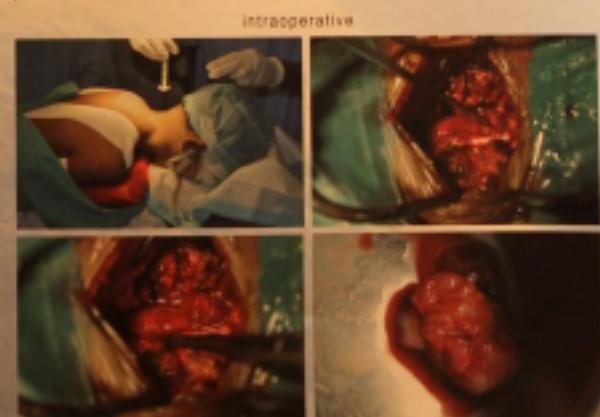
He showed dramatic recovery immediately following the Stage 1 operation involving the spinal cord decompression from the back of neck. He was able to walk with one person's support and feed himself with a spoon within 4-5days after the stage 1 surgery.



2nd stage surgery (anterior approach)

2nd stage surgery performed after 1 month apart for anterior removal of the neck mass.

After the stage 2 surgery his neurological recovery improved further and by end of two weeks he was able to manage himself independently, and also perform fine motor functions like writing.



Neurofibromatosis in patient's mother

