Original Research Paper

Volume -10 | Issue - 5 | May - 2020 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Neurosurgery

# SINGLE STAGED COMPLETE LENGTH EXCISION OF LONG SEGMENT INTRAMEDULLARY EPENDYMOMA: AN INSTITUTIONAL EXPERIENCE

| | |
|---|---|
| **Dr Anand Sharma** | Assistant professor, Department of Neurosurgery, Gajra Raja medical College |
| **Dr Parth Lalcheta*** | Mch Neurosurgery, Resident, Department of Neurosurgery, Gajra Raja medical College *Corresponding Author |
| **Dr Avdhesh Shukla** | Associate professor, Department of Neurosurgery, Gajra Raja medical College |
| **Dr S N Iyengar** | Professor and Head, Department of Neurosurgery, Gajra Raja medical College |

**ABSTRACT** Intramedullary tumours are the rare tumours of spine constitute about 2 % of all spine tumours with 0.74 per 100,000 person-years. Ependymomas and astrocytoma represent most common intramedullary tumours. Long segment intramedullary tumors are usually myxopapillary ependymoma; involve more than three vertebral column lengths. Pain and motor weakness are the most common presentations in adults and children. The goals of surgery are total tumor removal with preservation of neurological function. We report case series of two long segment intramedullary tumours involving long segment with review of literature.

**KEYWORDS :** Long segment intramedullary tumour, ependymoma

## INTRODUCTION:

Intramedullary tumours are the rare tumours of spine constitute about 2 % of all spine tumours with 0.74 per 100,000 person-years. Ependymomas and astrocytoma represent most common intramedullary tumours. Ependymoma of spinal cord constitute about. 5% of all intramedullary tumours, usually common at conus and arises from filum terminale. Forty percent of spinal ependymomas occur in the filum terminale. The majority of these are myxopapillary, occurring with a slight male predilection (1.7: 1) with a mean age at diagnosis of 36.4 years. [1]Long segment intramedullary tumors are myxopapillary ependymoma; usually involve more than three vertebral column lengths. Standard treatment for these tumours is complete surgical excision, but long segment involvement requires challenging teamwork. We report case series of two long segment intramedullary tumours involving long segment with review of literature.

**Case report 1:** 35 year female reported to neurosurgery OPD with complain of progressive paraparesis with history of bladder-bowel dysfunction. Neurological examination revealed power 2/5 both lower limbs with bladder-bowel dysfunction. Sensory examination grossly normal below L1. Deep tendon reflex of both lower limbs were exaggerated while superficial reflex (abdominal) were absent. MRI dorsal spine revealed long segment intramedullary tumour extending from D6 to D12, it was hypointense on T1 image and hyper intense on T2 sequence. Two polar cysts also seen at both margin of tumours. After taking proper consent surgery was planned, D6-D12 laminectomy was done, dural were tense durotomy and myelotomy were done. Intraoperatively grayish white tumour was identified with clear plan of dissection from neural tissue. Intratumor decompression of tumour was done followed by complete excision of tumour done with proper hemostasis. Closure done in standard fashion. Neurological status of patient improves in postoperative period and there were no recurrence was found in two-year follow up with resolution of polar cyst. Histopathology report confirms ependymoma.

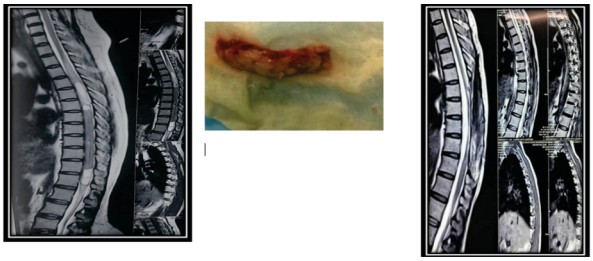
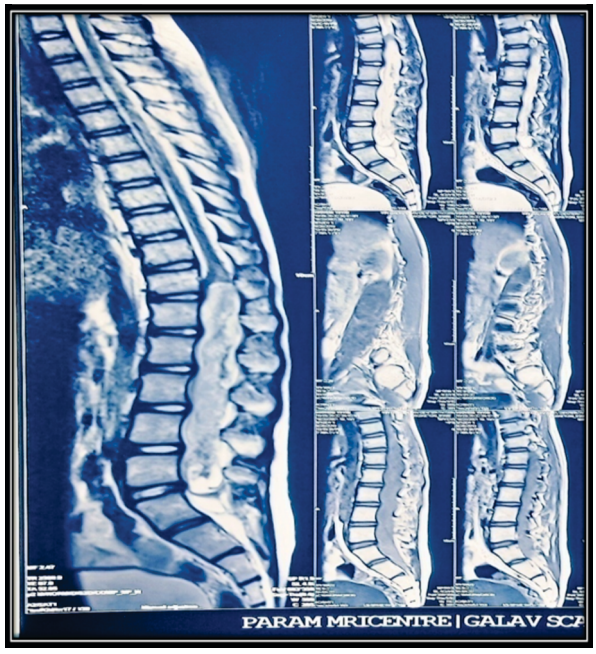


**Figure 1:A) MRI dorsal spine revealed long segment intramedullary tumour extending from D6 to D12 B) Postoperative MRI dorsal spine with complete excision with no recurrence on 2 year follow up C) Postoperative tumour specimen**

**Case Report 2:** 18 years old male presenting with complaints of low back pain of gradual onset, progressive, increased on standing and walking and not relived by taking medicines with bowel and bladder disturbances for last 2 months with no history of recent trauma and no known co-morbidities with no history of similar complaint in past. On examination power in both lower limb MRC grade was 5/5 with no sensory deficits. MRI lumbosacral spine with contrast done which showed expansile intramedullary soft tissue mass in thoracolumbar spine cord from D11 to L5 levels [17 cm length]. Posterior vertebral bodies at these levels show scalloping. The mass heterogeneously hyperintense on T2WI and hypointense on T1WI.Cystic area seen at the distal end of mass. Rest of the spinal cord is normal. Features suggestive of Myxopapillary ependymoma. In view of these findings patient planned for operation-D11 to L5 laminectomydone. Intraoperatively tumour was soft, grayish white attach to filum terminale and conus. Filum terminale were identified at both margin and complete en blocremoval of tumor with gentle nerve dissection wasdone. Post operatively, patient power on both lower limbs intact [5/5], with significant improvement in bladder and bowel function. Histopathologyreport confirmsmyxopapillary ependymoma.




**MRI LS Spine showed expansile intramedullary mass extending from D11 to L5 with hyperintense signal on T2 Weighted sequence**

## DISCUSSION:

Intramedullary spinal cord tumours account for 15−20%of all intradural tumours and only 2−4% of all intrinsic tumours of the central nervous system. Astrocytomas and ependymomas are the most common histological types of intramedullary tumours. Astrocytomas comprise 90% of paediatric (<10 years) and 60% of adolescent intramedullary tumours. [2] Ependymomas are the most common intramedullary tumours in adults[3,4]

Pain and motor weakness are the most common presentations in adults and children. Intramedullary tumours produce an ill-defined pain, which is seldom radicular (except in cauda-conus lesions) but localizes at the levelof the lesion. It is said to be more prominent at night in the supine position. Intramedullary lesions classically produce a long segment lower motor neuron type of weakness at the level of the lesion with atrophy, hypotonia, areflexia and fasciculations, along with features of upper motor neuron weakness below the lesion. Weakness is maximal at the level of the lesion. Patients often complain of numbness, which begins distally and then extends proximally. This often takes the form of “glove and stocking”, “cape distribution” or “dissociated anesthesia”. Scoliosis may be the earliest sign of an intramedullary tumour in a young child. Local lumps, spinal point tenderness and the presence of stigmata of spinal dysraphismare all indicators of underlying pathology. Patient may also present with features of urinary incontinence. [5]

Most intramedullary tumours expand the spinal cord, are hypointense on T1- and hyperintense on T2-weighted sequences and enhance with contrast. Ependymomas are more symmetrical, enhance with contrast uniformly and are more frequently associated with polar cysts.[6] Up to 65% of ependymomas may be associated with a syrinx.Astrocytomas tend to be poorly marginated, enhance heterogeneously with contrast and may be associated with intratumoural cysts and necrosis. Up to 20% of astrocytomas may be associated with a syrinx. Hemangioblastoma are intensely enhancing lesions located dorsally or dorsolaterally within the spinal parenchyma and may be associated with flow voids and large cysts or a syrinx out of proportion to the size of the tumour. The entire neuraxis should be screened, if this pathology is suspected. Metastases and lymphomas may be associated withleptomeningeal enhancement. This sign also indicates tubercular, bacterial or fungal myelitis. [6]

The goals of surgery are total tumor removal with preservation of neurological function. Patients need realistic guidance, especially those who are functionally intact, continent and have minimal neurological deficits. Ependymomas push the normal spinal parenchyma aside, are distinct from the surrounding spinal cord, usually with a well-defined plane of cleavage and may be dissected from it. They are firm and reddish-grey or yellow. Cysts may frequently be found at either end of the tumor. Myxopapillary ependymomas are well-encapsulated, lobulated solid fleshy tumors usually found adherent to the filum terminale generally confined to the lumbosacral area with good surgical plane amenable to en bloc resection. [7]The most common surgical approach -- the standard midline approach with laminectomy or laminoplastyadvantage being avoids encounter with major vessels orvisceral structures.Careful microdissection from adjacent cauda equina nerve roots is required to free the tumor of adhesions.[8,9]Patients with ependymomas should not be irradiated after documented complete excision. Recurrences should be dealt with by redo surgery and radiation if a residual tumour burden exists. When a residual tumour exists after surgery, radiation is an option if redo surgery is deemed risky.[10]

## REFERENCES :


1- Sonneland PR, Scheithauer BW, Onofrio BM. Myxopapillary ependymoma. A clinicopathologic and immunocytochemical study of 77 cases. Cancer. 1985;56: 883–893.
2- Shrivastava R, Epstein F, Perin N, et al. Intramedullary spinal cord tumors in patients older than 50 years of age: Management and outcome analysis. J Neurosurg Spine. 2005;2:249−55.
3- Helseth A, Mørk SJ. Primary intraspinal neoplasms in Norway, 1955 to 1986. A population-based survey of 467 patients. J Neurosurg. 1989;71:842−5.
4- Manzano G, Green BA, Vanni S, et al. Contemporary management of adult intramedullary spinal tumors-pathology and neurological outcomes related to surgical resection. Spinal Cord. 2008;46:540−6.
5- Schwartz TH, McCormick PC. Spinal cord tumors in adults. In: Winn HR (Ed). Youman’s Neurological Surgery.Philadelphia: Saunders; 2004.
6- Samii M, Klekamp J. Surgical results of 100 intramedullary tumors in relation to accompanying syringomyelia.Neurosurgery. 1994;35:865−73.
7- Lee M, Rezai AR, Freed D, et al. Intramedullary spinal cord tumors in neurofibromatosis. Neurosurgery. 1996;38:32−7.
8- Sonneland PR, Scheithauer BW, Lechago J, et al. Paraganglioma of the cauda equina region: clinicopathologic study of 31 cases with special reference to immunocytology and infrastructure. Cancer. 1986;58:1720–1735.
9- Pigott TJ, Lowe JS, Morrell K, et al. Paraganglioma of the cauda equina. Report of three cases. J Neurosurg. 1990;73:455–458.
10- Watson JC, Stratakis CA, Bryant-Greenwood PK, et al. Neurosurgical implications of Carney complex. J Neurosurg. 2000;92:413–418.