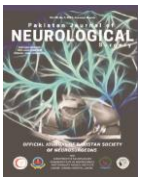

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Original Article (SPINE)

Multidisciplinary Management and Outcome of Intradural Extramedullary Spinal Tumors

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ABSTRACT

Introduction/Objective: About fifteen percent of the primary CNS tumors are intraspinal. About two-thirds of tumors are intradural extramedullary (IDEM). This study was conducted to review the outcome of operative management of intradural extramedullary tumors in correlation with the factors, both clinical & histopathological, influencing the neurology of patients & prognosis.

Materials and Methods: It was a multicenter study including 42 patients conducted from December 2018 to December 2020. All patients were diagnosed by MRI with and without contrast. Patients were surgically treated & analyzed for clinical features i.e., pain by visual analog scale (VAS) & neurology by modified McCormick scale both preoperatively & post-operatively. Clinical features & outcomes were correlated with tumor size & histopathology. p-value < 0.05 was considered significant.

Results: This study included 42 cases. The most common diagnosis was schwannoma (76.19%). The average intradural space occupied at presentation was 82%. The most common location was dorsal (90.4%). The visual analog score for pain (VAS) improved in all patients post-operatively from 7 ± 1.9 to 2 ± 0.8 (p = 0.003) & modified McCormick scale from 3.0 ± 1.3 to 2.0 ± 1.0 (p = 0.005). The preoperative symptoms were correlated with the only size of the tumor occupying the intradural space (VAS p = 0.021, modified McCormick scale p = 0.018).

Conclusion: All the tumors excised showed some improvement in neurological status. Therefore, all patients diagnosed with IDEM should be operated on even if present with prolonged symptoms or severe neurological compromise.

Keywords: Intradural Extramedullary, Meningioma, Schwannoma, Intraspinal.

Note: Neurofibroma & Schwannoma are used as a synonym in this study.

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INTRODUCTION

About 15% of the primary CNS tumors are intraspinal.[1-4] About 2/3rd of tumors are intradural extramedullary. Neurofibromas, meningiomas & myxopapillary ependymomas account for most extramedullary tumors.[5-8] Most common tumors are those derived from nerve sheath cells covering the spinal nerve roots known as schwannomas. Spinal meningiomas account for 25 – 46% of spine-related tumors. They occur most commonly in the dorsal area in middle-aged females & grow laterally in the subarachnoid space until they become symptomatic. These tumors mostly present due to compression rather than invasion.[9] 1% of nerve sheath tumors are intramedullary & are considered to arise from perivascular nerve sheath that goes along the penetrating spinal cord vessels.[10] Approximately 2.5% of intradural tumors are malignant.[11]

Intradural extramedullary tumors commonly present with dull pain, loss of sensations, motor weakness & sphincter loss. Although the clinical presentation & pathology of IDEM tumors remains the same, there has been a dramatic improvement in the diagnosis and treatment after the advancement of the radiological & surgical techniques, particularly CT & MRI brought marked improvement in early diagnosis and exact localization of tumor & its dural attachment. The operating microscope has significantly improved outcomes.[2-4] Surgical assistive devices like intraoperative neuromonitoring, ultrasonography, use of microscope & ultrasonic surgical aspirator have all played part in the improved outcome of excision of spinal tumors. It has aided in achieving total tumor resection.[5]

The objective of this study was to determine the surgical outcome of IDEM tumors & to identify factors both clinical & histopathological influencing patient outcomes in terms of functional improvement & pain relief. For early rehabilitation of patients' physiotherapists were involved and for rationale use of drugs opinion from pharmacist was obtained as per the need of patients. So, patients included in this study were managed with the help of a multidisciplinary team including a neurosurgeon, anesthetist, physiotherapist & pharmacist.

MATERIALS AND METHODS

Study Design and Setting:

This was a nonrandomized prospective study. We had 42 patients in this study, which was carried out in two centers i.e.; Services hospital Lahore& Surgimed hospital Lahore. The study period was between Dec. 2018 & Dec. 2020.

Inclusion criteria

All patients were diagnosed on MRI as new cases of intradural extramedullary tumors.

Exclusion Criteria

Recurrent cases and those not fit for general anesthesia were excluded from the study.

Data Collection

The study population was studied as patients& our patients. Each patient's demographic details symptomatology & signs as per questionnaire, radiological features, surgical procedure, the outcome at follow-up in terms of modified McCormick scale& VAS, and histopathological diagnosis were noted in proforma. The preoperative duration of symptoms was also carefully noted. Pre-operative pain was recorded using a visual analog scale (VAS). The patient's neurological status was graded into 5 levels as per the modified McCormick scale (Table 1).[5]

The %age of tumor occupying the intradural space was calculated on axial MRI images as:

Transverse + longitudinal diameter of tumor mass/transverse + longitudinal diameter of intradural space × 100.

Table 1: Modified McCormick Scale

1.	Neurologically intact, normal deambulation, minimal dysesthesia.
2.	Mild motor or sensory deficit but functional independence.
3.	The mild deficit, limitation of function, independent of support.
4.	Severe motor or sensory deficit, limited function, & dependent.
5.	Paraplegia or quadriplegia, even with flickering movements.

All patients were investigated with pre-op MRI. Follow-up of patients was done for clinical improvement & radiology at 3 months, 6 months & 1 year.

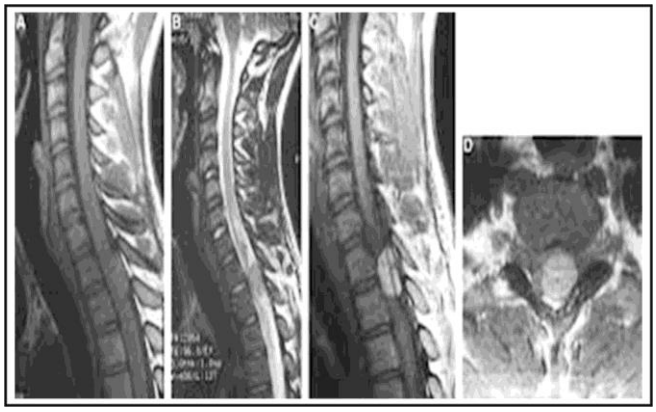

Figure 1: MRI cervicodorsal spine sagittal view T1, T2 weighted & contrast (A, B & C respectively) & axial view(D) showing meningioma at D1-2 level (Images included with the consent of the patient).

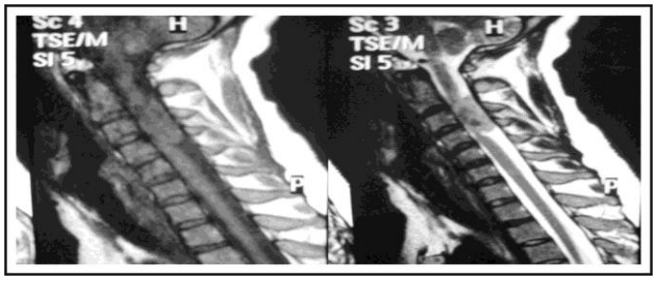

Figure 2: MRI cervical spine T1 weighted & T2 weighted images showing a neurofibroma at the C3-4 level. (Images included with the consent of the patient).

Surgical Procedure

All patients were approached posteriorly. Patients were positioned prone. Under strict aseptic measures, a skin incision was made. Skin, subcutaneous tissue cut, fascia incised, subperiosteal muscle dissection done. Laminectomy was performed after confirming level with C-Arm. Durotomy done. Tumor excised under the microscope. Nerve fibers passing through the tumor are sacrificed.34 marginal& 8 intralesional & piecemeal tumor excisions done. Dural attachment with tumor coagulated. Hemostasis secured. Dura closed water tight. Wound closed in layers. Tumor tissue was sent for histopathology.

Statistical Analysis

All the data was entered & analyzed by using SPSS 25. The Chi-square test was used to determine the significant or insignificant results. A p-value < 0.05 was considered significant.

RESULTS

Gender Distribution

Overall, 42 patients were included in the study. 7 (16.6%) patients were females &35(83.4%) were male.

Age Distribution

Maximum patients belonged to the age group of 30-40 years (Table 2).

Table 2: Age distribution: frequency & percentage.

Age	Frequency	Percentage
30-40 Years	24	57.1
41-50 Years	16	38.1
51-60 Years	2	4.8

Location of Tumor

As observed on MRI 38 (90.4%) cases were

located at the dorsal spine, 3 (7.1%) at the dorsolumbar junction & 1 (2.3%) was located at the cervical spine. The average % age of tumors occupying intradural space was 82.0 ± 8.6%.

Histopathological Diagnosis

Pathological data showed the most common diagnosis was schwannoma 32 (76.19%) followed by meningioma 8 (19.04%) followed by myxopapillary ependymoma 2 (4.76%).

Presentation

The mean duration of symptoms was 7.6 months ranging from one month to 16 months. Presenting complaints were limb weakness, sphincteric problems, dull pain & numbness.

Post-Operative Complications

Superficial wound infection was the most common complication (7.14%). See Table 3.

Table 3: Complications: Frequency & percentage.

Complications	Frequency	%age
CSF Leakage	2	4.76
Superficial wound infection	3	7.14
Total	5	11.9

Outcome at Final Follow-Up:

Symptoms improved in all cases postoperatively. The VAS improved in all subjects from 7 ± 1.9 to 2 ± 0.8 (p = 0.003). The Modified McCormick scale improved from 3.0 ± 1.3 to 2.0 ± 1.0 (p = 0.005) at the last follow-up. See Table 4.

Preoperative symptoms measured by VAS & modified McCormick scale were highly linked with tumor occupancy of intradural space (average pre-op space occupancy 82%) (VAS p = 0.021, modified McCormick scale p = 0.018). However, no statistically significant link was established between duration of symptoms preoperatively & outcome or location of tumor & prognosis/ outcome.

Table 4: Clinical Outcome Based on The Visual Analog Scale & Modified McCormick Scale.

	Pre-operative Average	Postoperative Average	P-value
Visual analog scale	7 ± 1.9	2 ± 0.8	0.003*
Modified McCormick scale	3 ± 1.3	2 ± 1.0	0.005*

*significant result

One case of neurofibroma & one case of meningioma reoccurred, at last, follow up. One patient lost to follow-up after the second one i.e., at 6 months.

DISCUSSION

Historically Sir Victor Horsley, succeeded in removing dorsal IDEM in 1988 & it was about 44 years before the invention of myelography.[3] Thereafter the advancements in radiological techniques & invention of the surgical microscope have shown remarkable improvement in identification & surgical cure, but basic operative rules remained unchanged.[4]

Previous reports documented that about 5 women & 3 men out of 1,000,000 people develop primary spinal tumors every year & only 2/3rd of these were IDEM (Intradural extramedullary).[6] So it is difficult to get studies with a large patient population, so most of the studies found are focused on analyzing previous studies.

Meningiomas account for 25-46% of intra-spinal tumors. Spinal meningiomas are only 7.5-12.5% of CNS tumors as mostly are intracranial.[7] Spinal meningiomas are mostly located in the

thoracic spine & are most common in women, may be due to hormonal influence.[7]

For dural adherence, three surgical methods were used: 1) Some part of the dura was removed with the tumor to achieve complete removal & then duroplasty was done to achieve water-tight closure. 2) Some part of the dura mater is peeled off & the rest is primarily closed. 3) Coagulate the dural attachment.[7] In this study, we only coagulated dural attachment & only one case of recurrence was noticed at follow-up.

Schwannomas form 55% of IDEM tumors. They grow by displacing nerve fibers & commonly do not invade fibers. Total excision of the neurofibroma is achieved when the involved nerve fiber is also excised. Most surgeons believe that without sacrificing nerve fiber complete excision cannot be accomplished.[10-12] As per Kim et al. only 23% of neurofibromas with functionally reserved nerve roots develop mild to moderate neurological symptoms because the involved nerve roots are already dysfunctional.[13] In this study the nerve roots with no identifiable distal ends were removed with the tumor in marginal excision, however, in the case of indefinable distal component intralesional excision was performed. However, the neurological status of patients was static (same as pre-operative) immediate postoperatively & was improved in all cases on the modified McCormick scale on follow-ups, it emphasized that involved nerve roots were already dysfunctional. As there was only one case of recurrence on follow-up MRI we believe that preserving the nerve root did not affect tumor recurrence. Our observation was also supported by a previous study.[19]

In this study, the tumor was approached posteriorly for removal irrespective of the location of the tumor concerning the spinal cord. As per literature, 31% of the tumors are ventral to the cord[14] & certain authors claimed an extreme lateral or an anterior approach as compulsory for tumor excision.[15] However, the extreme lateral approach requires spinal fixation & fusion and anterior approaches pose difficulty due to epidural venous bleeding, limited field & multiple corpectomies.[19] In the current study, we used the posterior approach for all cases preserving facet joints and avoiding spinal fusions. For excision of cervical tumor, paired denticulate ligament, superior & inferior to the tumor was cut to increase the mobility of the spinal cord.

To save the neurology of the patient eight tumors were removed intralesionally including two ependymomas, four schwannomas & two meningiomas. Ependymomas are not solid enough, so difficult to remove marginally.[16,19]

As per previous studies, neurological symptoms were better post-operatively but partial improvement and even deterioration were also reported. Considering factors influencing prognosis a longer duration of preoperative complaints, severe neurological deficit,[2] more proximal tumor location[3] & a tumor more ventral to the spinal cord showed a worse outcome. However, in our study & a few more studies[19,20] functional neurology & pain improvement were affected by the maximum %age of tumor engaging the intradural space rather than preoperative length of the period of symptomatology, as all patients had improvement on VAS & modified McCormick scale post-operatively irrespective of the length of duration of disease. But as these are rare tumors & the sample size is small in most of the available literature & even in our study so it may be a statistical error. But the improvement in signs & symptoms favors surgical intervention in all diagnosed cases regardless of the prognostic factors. Compared to the acute conditions with similar levels of neurological compromise IDEM show better outcome as these are slowly progressing & nerve tissues have enough margin to adjust to compression effects.[19] However, some studies describe early recognition of symptoms and signs leading to early diagnosis of disease as a good prognostic factor. As per these studies, early diagnosis reduces the risk of postoperative

morbidity and may improve surgical outcomes.[21,22]

According to Mehdy et al. recurrence rate of IDEM tumors was 16%.[3] Others reported a recurrence rate of intraspinal tumors at 7.2% & out of those 46% were IDEM.[10] However, they related this recurrence rate with the more ventral position of tumor, extradural invasion & ependymomas. Another study conducted by Klekamp and Samiion ependymomas described the recurrence rate as 29.5%.[17] In our study, the recurrence rate was 4.76% but our follow-up period was short i.e., up to one year. So studies with a longer follow-up period should be considered more reliable.

As two of our patients had a CSF leak post-operatively which prolonged their hospital stay, we recommend the use of fibrin glue/bio glue for a better recovery & to reduce this complication furthermore.

It is stated that clinical symptoms of the tumor may be the same as a herniated disc so conducting a thorough physical examination along with detailed history &MRI of the appropriate region is essential for better treatment of the patient.[18]

CONCLUSION

The most prevalent spinal IDEM tumor was neurofibroma. The severity of preoperative symptoms was linked with % the age of tumor occupancy within the intradural space. Postoperatively, significant betterment in the neurological deficit was attained irrespective of affecting factors, so it is highly recommended to surgically treat all cases of IDEM tumors even those with prolonged or severe neurological deficits.

LIMITATION AND RECOMMENDATIONS

As spinal tumors are not very common so the sample size of this study is small with a short follow-up of only one year. It is recommended to include more institutions in the study for a larger sample population. A study with a follow-up of up to 5 years should be conducted.

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Additional Information

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Human Subjects: Consent was obtained by all patients/participants in this study.
Conflicts of Interest:
In compliance with the ICMJE uniform disclosure form, all authors declare the following:
Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Samra Majeed	1. Study design and methodology.
2.	Samra Majeed, Fareeha Majid	2. Paper writing and data calculations.
3.	Samra Majeed, Waqar Azim	3. Data collection and calculations.
4.	Nayyara Tahir	4. Analysis of data and interpretation of results etc.
5.	Azam Niaz, Samra Majeed	5. Literature review and referencing.
6.	Prof. Anjum Habib Vohra	6. Analysis of data and quality insurer.