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# Intradural Extramedullary Ependymoma at Lumbar (L1-L4 Level) Spine: A Suspicious Case and Literature Review

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**Abstract**

Ependymomas constitute 4-6% of primary central nervous system tumors. Spinal ependymomas are most frequently found in intramedullary region but few cases of intradural extramedullary ependymoma have also been reported. We report a 24-year-old male patient with a suspected case of intradural extramedullary ependymoma. Magnetic resonance images of the lumbar spine depicted an intradural mass from L1-L4 level. The spinal lesion was isointense on T1-weighted images and hyperintense on T2-weighted images, relative to the spinal cord. Laminectomy L1-L4 with gross-total excision was performed. Histopathological examination was inconclusive but suggested the possibility of ependymoma. Neurological recovery was initially observed but after few months symptoms worsened.

**Keywords:** Suspicious; Ependymoma; Intramedullary; Extra medullary

## Introduction

Tumors of spinal cord constitute 15% of Central Nervous System (CNS) tumors [1,2] They can be categorized as intradural or extradural, the former being either intramedullary (involving the substance of spinal cord) or extramedullary (outside the spinal cord) depending on their location. Ependymomas are the most frequent glial cells derived tumors found in the spinal cord. Classically, spinal ependymomas are intradural intramedullary tumors with predominance in adults. Intradural extramedullary spinal ependymomas are rare. Current literature suggests that very few cases of such tumors have been reported (Table 1). Although these spinal tumors are rare and benign but compressive lesions secondary to ependymoma could lead to range of symptoms from lumbago (lower back pain), sensory and motor disturbances to acute paraplegia [3-7]. Herein, we report a rare and suspected case of intradural extramedullary ependymoma in a 24-year-old male.

## Case Report

A 24-year-old male presented with history of mid/lower lumbago (back pain) for 1 month, progressive weakness of lower limbs for the last 5-6 days, and fecal and urinary retention for the last 3 days. Past medical history was unremarkable for trauma. On comprehensive neurological assessment, there was decreased muscle tone in both lower limbs, with overall grade-2 and grade-3 power in left and right lower limb muscle groups, respectively. Deep tendon reflexes (DTR) were absent in all four limbs. Other spine examinations were inconclusive.

### MRI screening

Detailed MRI screening suggested evidence of a large abnormal lesion within spinal canal starting at the level of L1 vertebra and extending down to the lower border of L4 vertebral body (Figure 1a). The lesion appeared isointense to cord on T1-weighted image, while hyper intense on T2-weighted image. MRI features were consistent with neoplastic lesion, likely of nerve sheath origin. Intradural extramedullary tumor was suspected as the initial diagnosis.

### Surgery

Laminectomy L1-L4, durotomy and gross-total excision of spinal mass was performed under general anesthesia. Midline spinal incision was given from L1-L4 in order to remove the mass. Intra-operative findings were multiple irregular gray brown soft bodies collectively measuring 3 × 2.8 × 0.5 cm with hemorrhage. Piecemeal excision was carried out with Redivac drain placement. No post-operative complications were observed.

### Histopathology

Histopathological examination showed rounded nuclei with eosinophilic cytoplasm focally showing nesting pattern with interspersed thick-walled vessels. At places, neoplastic cells were arranged around vessels. A panel of immunohistochemical examination was performed for antibodies against CD99, GFAP, CKAE1/AE3, S100, Dermis, CD138. Immunohistochemical staining was negative for all except CD99. Final histology report demonstrated inconclusive result but also suggested that the possibility of ependymoma could not be entirely excluded.

## Discussion

Spinal cord tumors tumors account for 15% of all CNS tumors.1, 2 Most prevalent location of such tumors is found to be intradural intramedullary though cases of intradural extramedullary ependymoam have also been reported in literature along with this case [3] Intradural extramedullary ependymomas are more prevalent among females and in 5th decade of life.6 Hormonal factor had been indicted as the major reason for female predominance by Duffau et al. in their review paper; however, its definite involvement is not well appreciated by other studies [8]. Contrary to most of the previous case reports, our patient was male with age range almost similar to that reported by Iunes et al.; 24-69 years and 23-87 years in our review of literature (2000-2013), respectively [3,4,6-19].

Magnetic Resonance Imaging (MRI) was the choice of neuroimaging modality since it can well localize the lesion [7,8]. Thoracic spine has been found to be the most frequent location of intradural extramedullary ependymoma [3,4,6-9,11,12,16-19]. Compared with earlier cases, our case was among the few with the lumbar spine involvement (suggestive

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| Author | Age | Gender | Location | Symptoms | Preoperative diagnosis | Histologic diagnosis | Prognosis | F/U Period | Recurrence |
|---|---|---|---|---|---|---|---|---|---|
| Duffau et al. [8] | 43 | Female | Thoracic (T1-T8) | Paraplegia, sensory abnormality, bladder dysfunction | Not mentioned | Benign Ependymoma | Good neurological recovery | 24 months | No recurrence |
| Bavbek et al. [10] | 46 | Male | Lumbar (L1-L2) | Monoparesthesia, urinary, fecal incontinence | Neurofibroma | Myxopapillary ependymoma | Neurological improvement | 6 weeks | No recurrence |
| Robles et al. [11] | 47 | Female | D2-D3 | Not mentioned | Neurinoma, meningioma | Benign classic ependyoma | No complications were seen, good neurological recovery | 1 year | Recurrence with anaplastic transformation |
| Graca [12] | 67 | Female | Thoracic (T5-T6 to T8) | Sensory abnormality, GD | Arachnoid cyst with spinal cord compression | WHO grade II ependymoma | Worsening of symptoms | 3 months | Recurrent cystic lesion |
| Schuurmans et al. [13] | 29 | Female | Cervical (C3-C6) | Neck pain, muscular weakness, urinary dysfunction | Not mentioned | WHO grade III anaplastic ependymoma | Neurological improvement | 2 years | Intracranial extracerebral metastasis |
| Bonfield et al. [14] | 87 | Female | Lumbar (L3) | Hip, thigh pain, bladder dysfunction | Not mentioned | Extramedullary ependymoma near conus medullaris | Postoperative course was uneventful | Not mentioned | Not mentioned |
| Guppy et al. [4] | 50 | Male | Thoracic (T5-T6) | Progressive weakness, sensory abnormality | Not mentioned | WHO grade III anaplastic ependymoma | Neurological improvement observed | 6 months | No recurrence |
| Iunes et al. [9] | 32 | Male | Bulbomedullary juntion, cervical (C2-C5), thoracic (T5-T11), Lumbar (L2, L4-L5), and Sacrum | Lower limb paresthesia, GD, urinary retention | Not mentioned | WHO garde II ependymoma | Tumor Progression and death | 10 months | Tumor recurrence |
| Son et al. [15] | 57 | Female | Cervical (C2-C6) | Neck pain, muscular weakness | Neurinoma, neurofibroma or meningioma | WHO grade II ependymoma | Neurologic improvement | 5 years | No recurrence |
| Landriel et al. [6] | 30 | Male | D2 and D12-L1 | LBP, urinary disturbance, GD, sensory abnormality | Not mentioned | WHO grade I Myxopapillary ependymoma | Lower limb paresis and radicular pain improved | 10 years | No recurrence |
| | 32 | Male | D10 | LBP, sensory abnormality | Not mentioned | WHO grade I Myxopapillary ependymoma | No improvement in referred symptoms | 1 year | No recurrence |
| Ha et al. [7] | 36 | Female | Cervical (C6)-Thoracic(T4) | Pain, paraplegia | Not mentioned | WHO garde II ependymoma | Neurological improvement | 6 months | No recurrence |
| Gardener et al. [16] | 27 | Female | Thoracic (T2-T7) | Band-like sensation in chest and urinary symptoms | Not mentioned | Ependymoma | Neurological improvement | 8 months | No recurrence |
| Kim et al. [17] | 48 | Female | Thoracic (T7-T9) | Radiating pain, motor disturbance, urinary incontinence | Not mentioned | WHO grade III anaplastic ependymoma | Neurological deterioration | 14 months | Newly developed mass at lumbosacral region |
| Moriwaki et al. [3] | 23 | Female | Thoracic (T4-T6) | Pain, sensory and motor disturbances, GD | Schwannoma or Meningioma | WHO grade II ependymoma | Neurologic improvement with mild paresthesia and pain in the left abdominal region | 1.5 years | Recurring mass at T4-T5 level |
| Perez-Bovet et al. [18] | 36 | Female | Multiple locations | Headache, CNP, Hemiparesis | Not mentioned | WHO grade III anaplastic ependymoma | No neurological recovery, patient died after 7 weeks of diagnosis | Not mentioned | Not mentioned |
| Samanci et al. [19] | 34 | Male | Thoracic (T7) to Lumbar (L2) | Backache, weakness, bladder disturbance | Not mentioned | Myxopapillary ependymoma | Neurological improvement | 3 years | Not mentioned |
| Present Case | 24 | Male | Lumbar (L1-L4) | Backache, lower limb weakness, fecal and urinary retention | Intradural extramedullary tumor | Possible Ependymoma | Neurological Recovery | Not mentioned | Not mentioned |

GD: Gait disturbance; LBP: Low Back Pain; CNP: Cranial Nerve Palsy

**Table 1**: Literature review on Intradural Extramedullary Ependymoma.

of conus ependymoam) (Figure 1b and 1c), [9,10,14,18,19]. Tumor location directly correlates with the symptomatology.4 Pain, sensory and motor deficits and bladder dysfunction were the most commonly reported symptoms in previously published cases [3,4,6-10,12-17,19]. Similar clinical features were also found in our case.

Initial neuroimaging findings are usually non-specific in terms of firm diagnosis.15 MRI findings in our case were consistent with those of previous cases; the tumor appeared isointense to spinal cord on T1-weighted images, while hyperintense on T2-weighted images [4,6,9,12,15]. Our literature review depicted that initial diagnosis

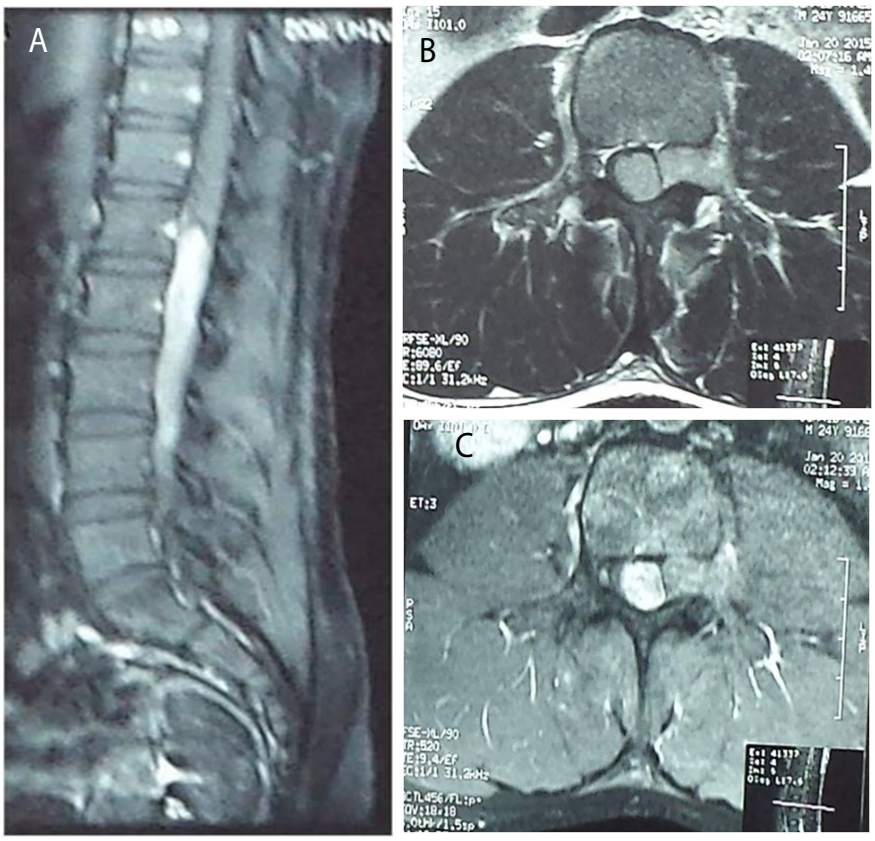


**Figure 1:** (a) MRI Image of the Lumbar Spine illustrating Intradural Extramedullary Lesion in the Lumbar Region, L1–L4. (b and c): MRI Images of the Lumbar Spine illustrating Intradural Extramedullary Lesion in the Lumbar Region, L1–L4.

was meningioma, neurinoma, neurofibroma, or schwannoma [3,10,11,15]. In the present case, pre-operative diagnosis was intradural extramedullary tumor.

Histopathologically, the case was suspicious which is striking and makes this case new in this entity. Despite extensive clinical and histopathological work up, the diagnosis remained elusive. The case is first of its kind in history with ambiguous histology result. On one side, MRI reports are clearly suggestive of conus ependymoma while on the other hand histology results are inconclusive but still suspection of ependymoma is open.

Surgically, gross total resection of the tumor has been regarded as the best approach for good prognosis as was done in the current case. Sonneland et al. also mentioned good survival results for patients who underwent gross total resection compared to those with partial resection of the tumor.20 Intradural extramedullary ependymomas have been described as benign tumors in the literature but countable cases have followed malignant sequelae [3,4,11,13,17,18]. No adjunctive radiotherapy was given to our patient since there was no evidence of residual tumor or any malignant transformation. Post-operative radiotherapy should be warranted in case of malignant transformation.7 Keeping in mind the possibility of malignant sequelae and recurrence, [3,4,9,11-13,17,18]. patient was guided for regular follow-up but no follow up was seen after few months.

## Conclusion

In toto, intradural extramedullary ependymomas are very rare in this part of the world. The present case of intradural extramedullary tumor is unique and should be subjected to scrutinize to identify what is going on at the molecular level.

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