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*Case Report*

# An uncommon intramedullary tumor: Primary medullary cone melanoma

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## ABSTRACT

**Background:** Melanoma is the third most common primary tumor to metastasize to the central nervous system (CNS). However, primary CNS melanoma is very rare, and primary intramedullary melanoma is even less frequently encountered, with only 27 cases published in the literature. There are no pathognomonic imaging characteristics, therefore, the diagnosis must be confirmed immunohistologically and the preferred treatment is the gross total resection.

**Case Description:** A 68-year-old male presented with low back pain of 2 months duration, and 1 week of urinary retention/anal sphincter incontinence. The neurologic examination revealed bilateral paraparesis (3/5 level) with bilateral Babinski signs, and a T10–T11 pin level. The lumbar CT-Scan showed a hyperdense intramedullary tumor arising from the conus medullaris. The patient underwent a D12–L2 laminectomy with myelotomy for gross-total tumor resection. Postoperatively, he regained motor function but the urinary incontinence remained unchanged. The diagnosis of a primary malignant melanoma was confirmed both histopathologically and immunohistochemically (e.g., staining revealed positive immunoreactivity for S100 protein and Melan A).

**Conclusions:** Primary intramedullary spinal melanoma is very rare, and the diagnosis must be biopsy/operatively confirmed. Whether gross total resection is feasible depends on the extent of tumor infiltration of the cord/adherence as well as the potential for clinical deterioration with overly aggressive removal.

**Keywords:** Intramedullary tumor, Medullary cone, Melanoma

## INTRODUCTION

Primary CNS melanoma is rare and accounts for only 1% of all melanomas according to the World Health Organization classification.[1,8] Primary intramedullary melanoma are even less frequently encountered, and there are only a few such cases in the literature.[3,8]

Patients present with symptoms reflecting the level of the intramedullary lesion. Complaints typically include somatic pain, myelopathy/motor deficits, sensory changes/pin levels, and sphincter dysfunction.[5,4] They occur most frequently in the thoracic followed by the cervical cord.[3] Since they are so rare, the recommended treatment is difficult to define, and the prognosis remains unclear.[4]

Here, we present a case of an intramedullary primary melanoma in a 68-year-old male who presented with paraparesis and a T10/T11 sensory level who did well following gross total tumor excision.

## CASE REPORT

A 68-year-old man presented with lumbar pain of 2 months duration that progressed to paraparesis with sphincter incontinence over the last week.

The neurologic examination confirmed the lower extremity motor strength of 3/5, bilateral Babinski signs, and a relative pin level at T10.

The lumbar CT-scan [Figure 1] showed a hyperdense intramedullary tumor arising from the conus medullaris at the L1 vertebrae level. No MR was done due to the patient's pacemaker device.

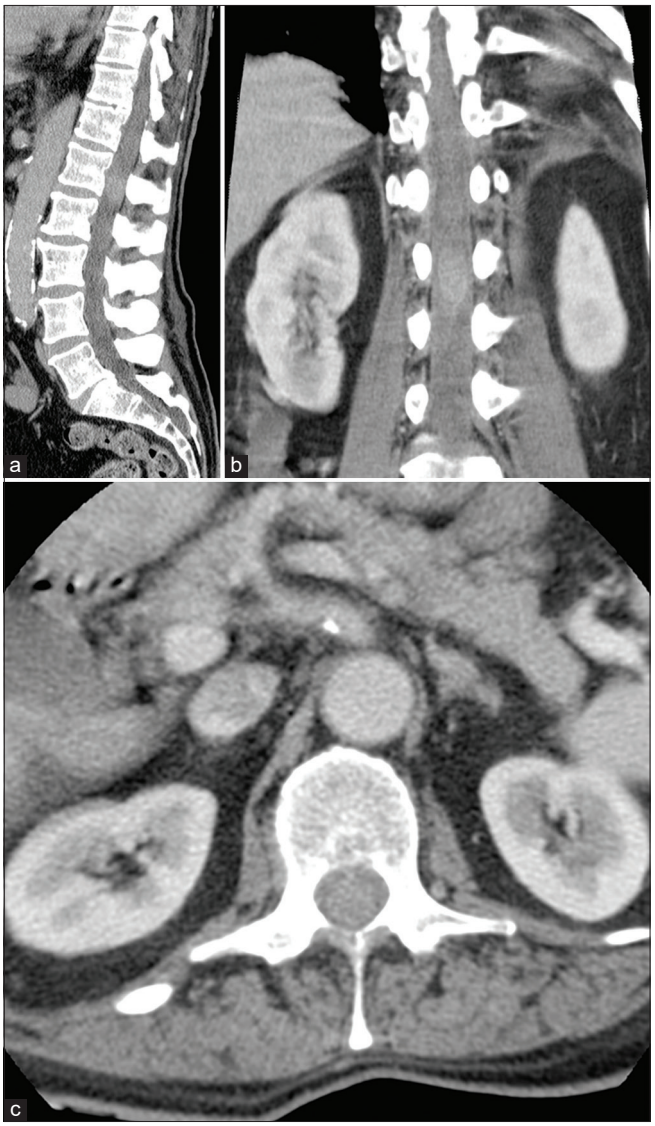


**Figure 1:** (a) Lumbosacral computed tomography showing a central hyperdense image at L1 level suggestive of a intradural lesion occupying almost all the canal with no extension to the foramen and with no bone lesion associated; sagittal plane, (b) pre-operative, coronal plane (c) pre-operative, axial plane.

Through a D12–L2 laminectomy and midline myelotomy, tumor was excised at the conus medullaris level; it was readily dissected and full resected exhibiting an excellent cleavage plane. It was soft, black, and had an intratumoral hematoma. Intraoperatively, there were no changes in the in the motor evoked potentials or somatosensory evoked potential. Watertight dural closure was achieved, and a laminoplasty was performed.

Postoperatively, the patient partially recovered muscular strength, the pain disappeared and he started a rehabilitation program, but did not regain urinary continence [Figure 2].

Notably, a positron emission tomography (PET)-SCAN, with tumor markers, ophthalmological, and dermatological examinations were performed, but no other primary tumor was identified. Further, primary malignant melanoma was confirmed with histopathology and immunohistochemical (e.g., positive immunoreactivity for S100 protein and Melan A) [Figure 3].

The patient refused radiation therapy and chemotherapy and was lost to follow-up at 3 months.

## DISCUSSION

The case presented was remarkable because the melanoma presented as an intramedullary lesion in the conus. Only, 27 similar intramedullary melanoma cases can be found in the literature [Table 1].[8]

Although MR is study of choice, here it could not be done due to the patient's pacemaker. The CT however showed a hyperdense lesion in the conus, suggestive of hemorrhage.[1] Subsequent surveillance imaging is also recommended looking for recurrence of tumors and/or metastatic disease (e.g., MR, CT, and PET-CT)

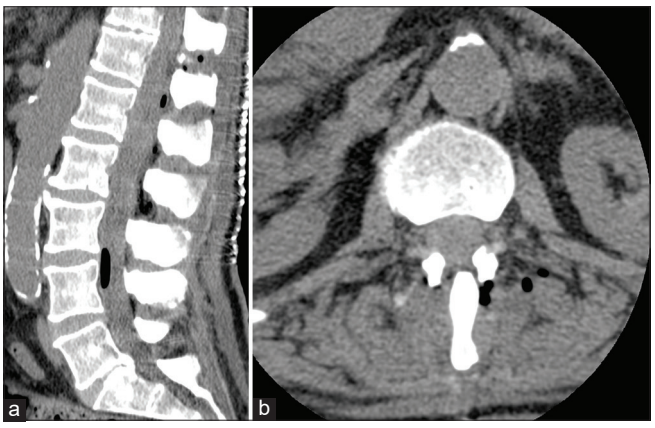


**Figure 2:** (a) Lumbosacral computed tomography showing a post-operative laminoplasty and excision of the previous lesion; sagittal plane (b) post-operative, axial plane.

**Table 1:** Summary of published cases of intramedullary primary melanoma.

| Author | Gender | Age | Level | MR | Removal | Adjuvant Treatment | Metastasis | Follow-up |
|---|---|---|---|---|---|---|---|---|
| Hirano y Carton, 1960 | M | 42 | T8–T9 | ND | Total | 60 Gy | No | 6m. Dead |
| Bergdahl *et al.*, 1972 | M | 45 | T6–T9 | ND | Parcial | 50 Gy | Brain | 33m. Dead |
| Hayward, 1976 | M | 69 | Thoracic | ND | Parcial | Rt | No | NR |
| | F | 52 | L1 | ND | Parcial | Rt | No | NR |
| Larson *et al.*, 1987 | M | 73 | T6–T8 | ND | Parcial | 50 Gy | No | 7y. Alive |
| | M | 63 | T9 | ND | Parcial | 60 Gy | Recurrent | 13y. Dead |
| | F | 67 | T9–T11 | ND | Parcial | 45 Gy | No | NR |
| | F | 57 | C1–C3 | ND | Parcial | 50 Gy | No | 30m. Dead |
| | F | 69 | T9–T10 | ND | Parcial | No | No | 45m. Dead |
| Magni *et al.*, 1996 | M | 64 | T8 | T1 hyperintense, T2 hypointense, without enhancement Gd | Total | No | Recurrence at 18m | 18m. Alive |
| François *et al.*, 1998 | M | 62 | T10 | T1 hyperintense, T2 hypointense, with enhancement Gd | Total | No | No | 28m.Alive |
| Salame *et al.*, 1998 | F | 79 | T9–T10 | T1 hyperintense with irregular enhancement Gd | Parcial | 30 Gy | No | 21m. Alive |
| Salpietro *et al.*, 1998 | M | 69 | C3 | T1 and T2 hyperintense with enhancement Gd | Parcial | 44 Gy | Brain | 15m. Dead |
| Vaquero *et al.*, 1998 | F | 50 | T10–T11 | T1 hyperintense, T2 hypointense with hyperintense areas with Gd enhancement | Parcial | Whole-brain and spine Rt | No | 15m. Alive |
| Bidzinskietal.,2000 | M | 36 | C6–C7 | T1 and T2 hyperintense | Total | 30 Gy | No | 48m. Alive |
| Farrokh *et al.*, 2001 | F | 80 | T12–L1 | T1 hyperintense, T2 hypointense, with enhancement Gd | Parcial | No | No | 9m. Alive |
| Denaro *et al.*, 2007 | M | 68 | T8–T9 | T1 hyperintense, T2 hypointense with hyperintense areas with Gd enhancement | Total | Interferon alpha-2 | No | 12m. Alive |
| Nishihara *et al.*, 2009 | M | 31 | T6 | ND | Parcial | Rt 50 Gy, interferon beta, intrathecal dacarbazine. Recurrence: Whole-brain Rt 30Gy+15Gy with interferon beta | Brain | 21y. Dead |
| Kim *et al.*, 2010 | F | 34 | T4 | T1 hyperintense, T2 hypointense, with enhancement Gd | Total | No | No | 36m. Alive |
| Kolasa *et al.*, 2010 | F | 57 | T10 | Gd enhancement | Total | Chemotherapy | Recurrence | 13m .Alive |
| Perrini *et al.*, 2010 | F | 81 | T10–T11 | T1 hyperintense, T2 hypointense, with enhancement Gd | Total | No | No | 6m. Alive |
| Liubinas *et al*, 2010 | F | 59 | T11 | T1 hyperintense, T2 hypointense | Parcial | 36 Gy | Recurrence | 71m. Alive |
| Fuld *et al.*, 2011 | M | 62 | C2–C3 | T2 Isointense with hyperintense areas with Gd enhancement | Parcial | 30 Gy | No | 11m. Alive |
| Trinh *et al.*, 2014 | F | 75 | T11–L1 | T1 Isointense with Gd enhancement | NR | Rt | NR | NR |
| Cetinalp *et al.*, 2014 | F | 47 | T9–L1 | T1 hyperintense, T2 Iso-hypointense, with enhancement Gd | Total | No | NR | 9m. Alive |
| Wu y Xu, 2016 | M | 51 | Medula-C2 | T1 hyperintense, T2 hypointense, with enhancement Gd | Total | Rt | Brain and recurrence | 10m. Dead |
| Yislenz Narváez-Martínez *et al*, 2017 | M | 49 | T7–T8 | T1 hyperintense, T2 hypointense | Parcial | 50 Gy | No | 20m. Alive |
| Current case | M | 68 | L1 | No | Total | No | No | 3m. Alive |
| Summary | 15 M | Avg. | C5 | T1 hyperintense, T2 hypointense, | 11 total | 18 Rt (0–60 gy) | 8/28 | Avg. 34m |
| | 13 F | 59 | T21 | with enhancement Gd | 16 partial | 2 Chemo | | |
| | | | L5 | | 1 NR | 8 No | | |

C: Cervical, F: Female, Gy: Greys, L: Lumbar, M:Male, m: Months, ND: No descried, NR: No related, MR: Magnetic resonance, Rt: Radiotherapy, T: Thoracic

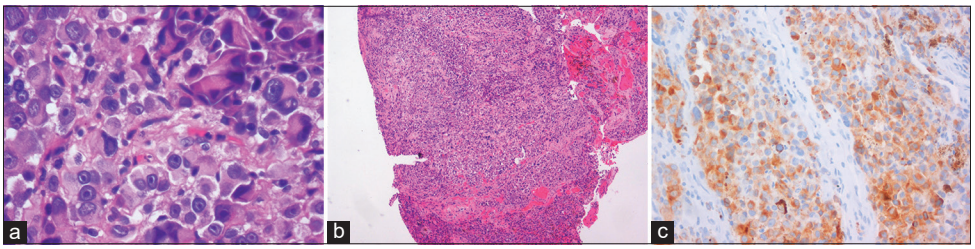


**Figure 3:** (a) Hematoxylin and eosin stain ×40 - large, irregular cells with moderate pleomorphism, with hyper-chromatic nucleus and prominent nucleoli visible. Some binucleated cells are also identified, (b) Hematoxylin and eosin stain ×4 - small magnification of the biopsy fragments, (c) Melan-A 20x - neoplasic cells with cytoplasmic marking.

As the diagnosis must be pathologically and immunohistologically confirmed, the patient in this study underwent gross total excision; this is preferred over biopsy or partial excision where feasible.[3,7]

Postoperatively, it is imperative to carry out dermatological, ophthalmological, and gastrointestinal examinations, along with a PET scanning to determine whether the intramedullary mass was primary or metastatic.[1,2]

Postoperative radiotherapy and/or chemotherapy are often recommended but there is no, clear evidence regard their efficacy.[7,6]

## CONCLUSION

Primary intramedullary spinal melanoma is very rare and unpredictable pathology and surgery remains the first choice of treatment with gross total resection utilizing microsurgical techniques and intraoperative monitoring.[3,8]

### Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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