An uncommon intramedullary tumor: Primary medullary cone melanoma

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Received : 13 June 2020
Accepted : 25 June 2020
Published : 18 July 2020

DOI
10.25259/SNI_352_2020

Quick Response Code:

INTRODUCTION

Primary CNS melanoma is rare and accounts for only 1% of all melanomas according to the World Health Organization classification.1,2 Primary intramedullary melanoma are even less frequently encountered, and there are only a few such cases in the literature.3,4

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.1,4 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.

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].

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. Subsequent surveillance imaging is also recommended looking for recurrence of tumors and/or metastatic disease (e.g., MR, CT, and PET-CT)

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**Figure 1:** (a) Lumbosacral computed tomography showing a central hyperdense image at L1 level suggestive of an 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.

**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  |
| Bergdal & al, 1972      | M      | 45  | T6–T9         | ND                                      | Partial | 50 Gy                                       | No         | 3m. Dead  |
| Hayward, 1976           | M      | 69  | Thoracic      | ND                                      | Partial | Rt                                          | No         | NR        |
| Larson et al., 1987     | M      | 73  | T6–T8         | ND                                      | Partial | 50 Gy                                       | No         | 7y. Alive |
|                        | F      | 63  | T9            | ND                                      | Partial | 60 Gy                                       | No         | 13y. Dead |
|                        | F      | 67  | T9–T11        | ND                                      | Partial | 45 Gy                                       | No         | NR        |
|                        | F      | 57  | C1–C3         | ND                                      | Partial | 50 Gy                                       | No         | 30m. Dead |
| Magni et al., 1996      | M      | 64  | T8            | T1 hyperintense, T2 hypointense,        | Total   | No                                          | Recurrence at 18m | 18m. Alive |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Francois et al., 1998   | M      | 62  | T10           | T1 hyperintense, T2 hypointense,        | Total   | No                                          | Brain      | 15m. Dead |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Salpietro et al., 1998  | F      | 79  | T9–T10        | T1 hyperintense with irregular          | Parcial | 44 Gy                                       | Brain      | 15m. Alive |
|                        |        |     |               | enhancement Gd                         |         |                                             |            |           |
| Salpietro et al., 1998  | F      | 67  | C3            | T1 and T2 hyperintense with             | Parcial | Whole-brain and spine Rt                    | No         | 12m. Alive |
|                        |        |     |               | enhancement Gd                         |         |                                             |            |           |
| Vaquero et al., 1998    | F      | 69  | T10–T11       | T1 hyperintense, T2 hypointense,        | Parcial | 50 Gy                                       | No         | 9m. Alive |
|                        |        |     |               | with hyperintense areas with Gd         |         |                                             |            |           |
| Bidzinski et al., 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,        | Parcial | No                                          | No         | 9m. Alive |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Denaro et al., 2007     | M      | 68  | T8–T9         | T1 hyperintense, T2 hypointense,        | Total   | Interferon alpha-2                          | No         | 12m. Alive |
|                        |        |     |               | with hyperintense areas with Gd         |         |                                             |            |           |
| Nishihara et al., 2009  | M      | 31  | T6            | ND                                      | Parcial | Rt 50 Gy, interferon beta, intrathecal      | Brain      | 21y. Dead |
|                        |        |     |               |                                         |         | dacarbazine. Recurrence: Whole-brain Rt     |            |           |
|                        |        |     |               |                                         |         | 30 Gy+15 Gy with interferon beta            |            |           |
| Kim et al., 2010        | F      | 34  | T4            | T1 hyperintense, T2 hypointense,        | Total   | No                                          | 36m. Alive |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Kolasa et al., 2010     | F      | 57  | T10           | T1 hyperintense, Gd enhancement         | Total   | Chemotherapy                                | Recurrence at 13m | 13m. Alive |
| Perrini et al., 2010    | F      | 81  | T10–T11       | T1 hyperintense, T2 hypointense,        | Total   | No                                          | 6m. Alive  |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Liubinas et al, 2010    | F      | 59  | T11           | T1 hyperintense, T2 hypointense,        | Total   | 36 Gy                                       | 7m. Alive  |
| Fuld et al., 2011       | M      | 62  | C2–C3         | T1 hyperintense, T2 hypointense,        | Total   | 30 Gy                                       | 1m. Alive  |
|                        |        |     |               | with hyperintense areas with Gd         |         |                                             |            |           |
|                        |        |     |               | T2 isoointense with hyperintense        |         |                                             |            |           |
| Trinh et al., 2014      | F      | 75  | T11–L1        | T1 isoointense with Gd                  | NR      | Rt                                          | NR         | NR        |
| Getinalp et al., 2014   | F      | 47  | T9–L1         | T1 hyperintense, T2 iso-hypointense,    | Total   | No                                          | 9m. Alive  |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Wu y Xu, 2016           | M      | 51  | Medula-C2     | T1 hyperintense, T2 hypointense,        | Total   | Rt                                          | Brain and recurrence | 10m. Dead |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Yislenz Narvaez         | M      | 49  | T7–T8         | T1 hyperintense, T2 hypointese          | Parcial | 50 Gy                                       | No         | 20m. Alive |
| Martínez et al, 2017    | F      | 47  | T9–T10        | T1 hyperintense, T2 hypointense,        | Total   | No                                          | NR         | NR        |
|                        |        |     |               | with enhancement Gd                    |         |                                             |            |           |
| Current case            | M      | 68  | L1            | No                                        | Total   | No                                          | No         | 3m. Alive |
|                        | F      | 59  | L5            | T1 hyperintense, T2 hypointense,        | 11 partial | 18 Rt (0–60 gy) | No         | 8/28 | 3m. Alive |
|                        |        | Avg.|                | with enhancement Gd                    |         |                                             |            | Avg 34m   |

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

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

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**How to cite this article:** Nogueira RM, Cardoso LS, Fonseca L, Branco P, Correia M, Roque P, et al. An uncommon intramedullary tumor: Primary medullary cone melanoma. Surg Neurol Int 2020;11:200.