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1    **Multiple melanocytoma of the thoracic spine: a case report and literature review**

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16

17    **Abstract**

18    **BACKGROUND CONTEXT:** Primary melanocytic neoplasms of the central  
19    nervous system (CNS) are rare tumors, and multiple localized melanocytoma are even  
20    more rare.

21    **PURPOSE:** To report **an unusual case of** multiple melanocytoma of the thoracic  
22    spine and discuss the diagnosis and treatment of primary CNS melanocytoma

1   **STUDY DESIGN:** A clinical case report and review of the literature.

2   **METHODS:** Chart review and analysis of **the** relevant literature.

3   **RESULTS:** A 60-year-old man who presented at our neurosurgery department with

4   progressive truncal numbness spanning three months. Magnetic resonance imaging of

5   the thoracic spine revealed two intradural extramedullary masses at T1 and T3–4,

6   respectively. Gross total resection of the lesions was performed. Postoperative

7   positron emission tomography/computed tomography (PET-CT) and **magnetic**

8   **resonance imaging** revealed no residual tumor or recurrence.

9   **CONCLUSIONS:** **We report a case of multiple primary spinal cord melanocytoma at**

10   **the T1 and T3–4 level.**

11

12

13   **Keywords:** Melanoma, Thoracic spine, Melanocytoma, Spinal cord

14

15   **Introduction**

16   Primary melanocytic neoplasms of the central nervous system (CNS) are rare tumors

17   that present in diffuse or localized form; multiple localized lesions are even rarer[1].

18   Here, we report a case of multiple primary thoracic spine melanocytoma and discuss

19   the preoperative diagnosis and treatment of primary CNS melanocytoma.

20

21   **Case report**

1 A 60-year-old man was admitted for progressive truncal numbness spanning three  
2 months. Physical examination revealed no evidence of cutaneous melanoma.  
3 Magnetic resonance imaging (MRI) of the thoracic spine revealed two intradural  
4 extramedullary lesions at the T1 and T3–4 level, respectively. Both lesions were  
5 hyperintense on T1 and hypointense on T2, and showed homogeneous enhancement  
6 following the administration of contrast agent (Fig. 1). The patient underwent T1–4  
7 thoracic laminectomy for symptomatic improvement. Intraoperatively, the dural,  
8 leptomeningeal, and arachnoid layers were partly dark-stained. The neoplasms were  
9 dark and closely adhered to the dura. Following gross total resection of both masses  
10 (Fig. 2), we coagulated the dura closely adhered to the masses. Some of the  
11 dark-stained dura and the leptomeningeal layer that could be exposed adequately were  
12 coagulated using bipolar coagulation to prevent recurrence. Histopathological  
13 examination suggested the diagnosis of intermediate-grade melanocytoma (Fig. 3).  
14 Light microscopic examination revealed tumor cells with intracytoplasmic brownish  
15 pigment and well-delineated nuclei. Mitotic count = 2/10 high-powered fields.  
16 Immunohistochemical staining revealed that the lesion was positive for S-100 and  
17 Ki-67 but negative for epithelial membrane antigen (EMA) and glial fibrillary acidic  
18 protein (GFAP). Postoperatively, the truncal numbness was markedly relieved, but the  
19 patient refused radiotherapy. He was discharged home a few days later and he  
20 resumed his normal activities. Positron emission tomography/computed tomography  
21 (PET-CT) conducted four months later confirmed the diagnosis of primary CNS

1 neoplasms and that the lesions had been completely resected. Follow-up MRI  
2 revealed no residual tumor or recurrence 19 months after surgery (Fig. 4).

3

4 **Discussion**

5 Primary melanocytic neoplasms of the CNS that arise from leptomeningeal  
6 melanocytes can be diffuse or localized. This group consists of a spectrum ranging  
7 from well-differentiated melanocytoma to its overtly malignant counterpart,  
8 melanoma[2]. In 2010, Liubinas *et al.* reviewed the English literature on primary  
9 melanocytic neoplasms of the CNS[1], and found that multiple lesions are very rare.

10 To the best of our knowledge, this is the fourth report in English of multiple localized  
11 melanocytoma; Table 1 lists the previous reports. Ali *et al.*[3] reported a case of  
12 multifocal meningeal melanocytoma; they had assumed it was a new pathological  
13 entity or the result of leptomeningeal seeding. Huang *et al.* reported a 24-year-old  
14 man with multiple spinal cord melanocytoma at the cervical and upper thoracic spine:  
15 the histological findings of the tumor were consistent with intermediate-grade  
16 melanocytoma[4]. In the present case, we confirmed the diagnosis of  
17 intermediate-grade melanocytoma according to the World Health Organization  
18 classification[5]. Thus, we believe that primary multifocal form, intraspinal metastasis,  
19 and leptomeningeal seeding could be the reason for the multiple lesions. We  
20 conducted PET-CT four months after the surgery, and detected no other lesions and  
21 no metastasis. **We speculate that preoperative PET-CT is helpful in the diagnosis of**  
22 **primary CNS neoplasm.**

1 Meningeal melanocytoma presents with focal neurological signs that are  
2 associated with their location, intracranial hypertension or hemorrhage,  
3 neuropsychiatric symptoms, spinal cord compression, or seizures. The lesions most  
4 commonly occur in the cervical and thoracic spine as intradural extramedullary  
5 lesions[1].

6 Somers *et al.*[6] reported a case of primary CNS melanoma and discussed the  
7 diagnosis, reporting that primary melanocytic lesions have characteristic findings  
8 upon neuroimaging: hyperintense on CT, hyperintense on T1, and hypointense on T2,  
9 which may be attributable to the paramagnetic effects of free radicals within the  
10 melanoma. In the present case, both lesions were hyperintense on T1 and hypointense  
11 on T2, and enhanced by contrast agent. Therefore, we believe that physicians who  
12 encounter a neoplasm that is hyperintense on T1 and hypointense on T2, whether  
13 solitary or multiple, should not discount primary CNS melanocytic lesions.

14 Melanocytomas are low-grade neoplasms curable by primary resection;  
15 melanomas are aggressive tumors with variable prognoses that depend on the tumor  
16 site and extent of resection[2]. Thus, whether benign or malignant, complete resection  
17 is advised for localized lesions. Even the benign melanocytomas may follow an  
18 aggressive course, with recurrence possible even after apparently complete excision.  
19 Therefore, adjuvant radiation therapy is advised in both cases of complete and  
20 incomplete resection[1]. In the present case, we performed gross total resection for  
21 both neoplasms. The dura was closely adhered to the masses, and parts of the  
22 dark-stained dura were coagulated, yet the patient refused radiotherapy for personal

1 reasons. Nineteen months after gross total resection, no sign of recurrence was found.  
2 We believe that, whether lesions are solitary or multiple, complete resection should be  
3 the first choice of treatment. However, we are uncertain of the best approach for  
4 addressing the dark-stained dural and leptomeningeal layers; we chose to coagulate as  
5 extensively as possible.

6

7 **Conclusions**

8 Multiple primary CNS melanocytoma is very rare and has not been well documented.  
9 We report a case of multiple primary spinal cord melanocytoma located at the T1 and  
10 T3–4 level.

11

12 **Conflict of interest**

13 None.

14

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1   **Figure legends**

2   Fig. 1. Preoperative MRI images. Preoperative MRI of the thoracic spine revealed two  
3   intradural extramedullary lesions at the T1 and T3–4 level. Both lesions were  
4   hyperintense on T1 (A) and hypointense on T2 (B); following the administration of  
5   contrast agent, they showed homogeneous enhancement (C).

6

7   Fig. 2. (A) The dark-stained dural, leptomeningeal, and arachnoid layers. (B)  
8   Macroscopic characteristics of the resected tumor tissue.

9

10   Fig. 3. Histopathological findings. (A) Hematoxylin–eosin, (B) S-100, (C) Ki-67, (D)  
11   EMA, (E) GFAP (all,  $\times 20$  magnification).

12

13   Fig. 4. (A) Postoperative PET-CT imaging (coronal view) revealed no recurrence or  
14   metastasis. (B, C, D) Follow-up MRI revealed no residual tumor or recurrence (B,  
15   T1-weighted image; C, T2-weighted image; D, T1-weighted contrast-enhanced  
16   images).

17

1    **Tables**2    **Table 1 The English literature on multiple localized melanocytoma**

Authors	Year	Age	Sex	Location	Resection	Follow-u	Status
O'Brien <i>et al.</i> [7]	1995	13	M	Thoracic spine	Incomplet e	3.5 years	Well
Ali <i>et al.</i> [3]				Bilateral cerebellop ontine angle and thoracic spine	Incomplet e	A few weeks	Death
	2009	31	M				
Huang <i>et al.</i> [4]	2011	24	M	Cervical and thoracic spine	Incomplet e	2 weeks	Well
Current case	2014	60	M	Thoracic spine	Complete	19 months	Well