A case report of primary thoracolumbar intraspinal malignant melanoma

Huang JB et al. Thoracolumbar malignant melanoma: A case report

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Abstract

BACKGROUND
Primary intraspinal malignant melanoma is a very rare tumor that most often occurs in the cervical, thoracic, or thoracolumbar segment.

CASE SUMMARY
A rare case of primary thoracolumbar malignant melanoma is described. A 45-year-old female patient complained of low back pain with numbness and fatigue in both lower limbs. MR revealed an intradural space-occupying lesion at the Thoracic 12 to Lumbar 1 (T12-L1) level. The tumor was partially excised, and a malignant melanoma was confirmed by histopathology.

CONCLUSION
Primary intraspinal malignant melanoma has rarely been reported, and surgical resection and related characteristics and diagnoses have been discussed.

Key Words: Intraspinal canal; Malignant melanoma; Spinal cord; Thoracolumbar; Case report

**Core Tip:** We report a case of primary thoracolumbar intraspinal malignant melanoma and emphasize the diagnostic challenge.
INTRODUCTION
The incidence of primary intraspinal melanoma (intraspinal primary melanocyte neoplasm, ISPMN) is extremely low, and only a small number of cases have been reported in the domestic and international literature. In November 2022, a patient with primary T12-L1 intraspinal malignant melanoma was admitted to our hospital and treated with surgery. After more than 1 year of follow-up observation, the results are as follows.

CASE PRESENTATION

Chief complaints
A 45-year-old woman who presented with low back pain with numbness and weakness of both lower limbs for two months was admitted to our hospital.

History of present illness
Two months before admission, the patient developed back pain, numbness and fatigue in both lower limbs; obvious symptoms on the left side; no fever or night sweats; and normal urine and feces. She visited the outpatient clinic in our hospital and other hospitals many times. After analgesia, nutrition, and sealing, the symptoms did not resolve, and they became worse.

History of past illness
The patient had no history of acute or chronic infectious diseases, heart disease, hypertension or diabetes.

Personal and family history
The patient did not have any relevant family medical history.

Physical examination upon admission
There was no systemic skin pigmentation. The patient had a poor gait, no kyphotic spinal
scoliosis deformity, no bilateral intervertebral space, no spinous space tenderness, no bilateral hip tenderness, double lower limbs without deformity, and normal joint active flexion and extension activity. Her bilateral hip "4" character was negative, her double lower limb straight leg elevation test (70)° was negative, her double lower limb superficial sensibility decreased, and her left side was even worse. The left iliopsoas and quadriceps muscles were level 3, the right muscles were level 4, and the bilateral anterior tibialis, dorsal extensor halluc, and ankle plantar flexor muscles were level 5. Both patellar tendon reflexes and Achilles tendon reflexes were normal, and no pathological signs were found in either lower limb. The VAS score for lumbar pain was 6.

**Laboratory examinations**

No abnormalities were found by blood draw after admission.

**Imaging examinations**

Lumbar CT revealed that the spinal canal at T12-L1 had a slightly greater nodular density and unclear boundaries, approximately 11 mm\*13 mm\*32 mm. Diagnostic consideration: Considering the extramedullary space-occupying lesion at the T12-L1 level, the possibility of chomenioma should be considered (Figure 1).

Enhanced MR revealed spindle swelling in the T12-T1 region (approximately 13 mm\*13 mm\*34 mm). Compared with muscle signals, T1WI signals were slightly greater, T2WI signals were equal, and there was a clear boundary. The adjacent border of the left intervertebral foramen side was irregular, the lesion was significantly enhanced under the enhanced scan, and the spinal cord was displaced to the right and half-wrapped. The diagnosis was as follows: extramedullary intradural space-occupying lesion of the T12-L1 lesions, which was considered a benign tumor: meningioma or a neurogenic tumor (Figures 2 and 3).

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**FINAL DIAGNOSIS**

The patient had no history or clinical manifestations of the primary cutaneous or ocular
lesion. Based on the MRI analysis and histological examination, the final diagnosis was primary thoracolumbar intraspinal malignant melanoma.

**TREATMENT**

The patient underwent "posterior open T12 intraspinal mass resection + spinal decompression + internal fixation surgery". During the operation, the dural sac was slightly expanded, and the size of the spinal canal after the dural incision was approximately 25 mm*10 mm*10 mm (black brown mass) and clot-shaped. The mass had an unclear boundary and obvious adhesion to the cauda equina nerve. The severe adhesions between the mass and the cauda equina nerve could not be completely removed.

Postoperative pathology revealed that the T12 spinal mass was generally dark brown and clot-shaped (Figure 4), with microscopic tumor cells, rich pigment, heterogeneous tumor cells, and large nuclei.

Immunohistochemistry revealed the following: HMB 45(+), S-100(+), Milan-A(+), SOX-10(+), Bcl-2(+), P16(+), CK (partial +), CyclinD1 (partial +), BRAF (V600E) (partial +), and Ki-67 (hot zone 40% +). The results of immunohistochemistry were consistent with malignant melanoma (Figures 5 and 6).

**OUTCOME AND FOLLOW-UP**

Postoperative examination via brain enhanced MR, chest and whole abdomen enhanced CT revealed no suspicious tumor lesions. After surgery, the patient's lower limb numbness was reduced, and the patient still experienced mild numbness of the left lower limb and fatigue, especially in the left thigh. After discharge, the patient underwent regular outpatient follow-up and experienced numbness in the left thigh, occasionally with needle-like pain. One month, five months and one year after surgery, enhanced MR revealed that the signal was still abnormal in the spinal canal and was approximately 7 mm*12 mm*30 mm, indicating residual tumor tissue (Figure 7).
DISCUSSION

Epidemiology

Intracranial primary melanoma (primary melanocytic neoplasm, PMN) originates from leptominal melanocytes and is rare in clinical practice, accounting for 0.07%~0.17%\(^1\) of intracranial tumors. Primary intraspinal melanoma is even rarer and is mostly located in the extradural dural ridge or near the dural spinal nerve roots. Nericular pain often occurs first\(^2\), with the greatest occurrence occurring in individuals approximately 50 years old. Brat et al.\(^3\) reported 33 cases of central nervous system (CNS) melanoma, with 52% occurring in the spinal canal and 48% in the intracranial area. Among them, the age of onset of cerebrospinal membrane melanoma was 16~73 years, the mean age was 51 years, the female/male ratio was 1.43:1, while the age of onset of malignant melanoma was 15~71 years, the mean age was 43 years, and the female/male ratio was 1.6:1. Rahimi-Movaghar and Karimi\(^4\) summarized 95 cases of CNS cerebrospinal membrane melanoma (45 intracranial cases, 50 spinal cases); the mean age of intracranial patients was 40 years, the mean age of patients in the spinal canal was 49 years, and 57.9% of the patients were female.

CNS-PMN originates from the leptominal sheath of the vascular plexus or embryonic remnant melanoblasts, with a high degree of aggregation at the craniocervical junction, the ventral medulla, and the pontine cerebellum angle\(^5\). Although melanocytes are concentrated in the upper cervical segment, tumorigenesis is not limited to the cervical segment, occurs in the lower thoracic segment or thoracolumbar segment, and often originates near the cranial nerve and spinal nerve roots, which may be related to the extreme rarity of tumors, small sample sizes and significant differences\(^6,7\).

Pathology

Brat et al.\(^3\) classified PMNs into benign cerebrospinal membrane melanoma, malignant melanoma and the intervariable intermediate type, accounting for 52%, 39% and 9%, respectively. Benign cerebrospinal membrane melanoma primarily occurs in the spinal canal in 70.6% and 29.4% of malignant melanomas, respectively. Malignant melanoma
cells can be shed in the subarachnoid space and spread on the soft brain membrane, where they can form several black nodules of varying sizes, rich tumor blood transport and clear boundaries. Malignant melanoma occurs from the skin and mucosa and is a highly malignant tumor. Microscopy of malignant melanoma revealed that tumor cells are significantly pleomorphic, different sizes, polar processes and adhesive bands, are arranged into sheets and loose nests, or extend along blood vessels, invading the basement membrane layer; large nuclei, irregular shapes, coarse chromatin, inconspicuous nucleoli, dual or multinucleation, and abnormal nuclear phenomena are common; melanin granules are filled with cytoplasm, and melanosomes and premelanosomes coexist.

**Diagnosis**

ISPMN occurs near the spinal nerve root, mostly in extramedullary subdural or dural lesions, and nerve root pain caused by nerve root compression is often the initial manifestation. Compression of tumor development in the spinal cord can lead to sensory, motor or stool and urine dysfunction in the corresponding segments. Stimulation of the meninges by tumor cell metabolites may cause meningitis. Tumors can cause bleeding and recurrent episodes of spontaneous subarachnoid hemorrhage. The diagnostic conditions for spinal canal primary melanoma were as follows: ① only spinal canal lesion, no intracranial homologous lesion was found; ② except for skin, ocular pigment membrane melanin lesions, and parenchymal viscera melanoma; and ③ the tumor had a clear histopathological diagnosis. Due to the extremely low incidence of ISPMN and the lack of specificity of clinical symptoms and signs, comprehensive imaging, surgical and pathological data are needed to confirm the diagnosis.

**Imaging examination**

MRI of ISPMNs is mainly divided into type ④[8]: (1) melanin type, in which T1WI has a high signal and T2WI has a low signal; (2) nonpigmented type, in which the T1WI signal is low or equal, while the T2WI signal is high or equal; (3) mixed type, in which the signal
is different from that of melanin and nonpigmented type; and (4) blood type, in which the signal is only bleeding. In this case, MRI of an intraspinal malignant melanoma showed a slightly greater signal in T1WI and an equal signal in T2WI. The plain ISPMN CT scan was round or round, with a high-density shadow, and the degree of enhanced bleeding was high. Due to the very low incidence of intraspinal melanoma and the lack of specific clinical symptoms and signs for diagnosis, pathological and immunohistochemical examinations are needed. HMB-45, Melan-A, and S-100 are commonly used for melanoma immunohistochemistry. The S-100 protein is highly expressed in malignant melanoma but is less specific; HMB-45 is highly specific, and combining HMB-45 and Melan-A can prevent missed diagnoses.

Typical melanin imaging findings can improve the accuracy of preoperative diagnosis.

**Antidiastole**

Because intraspinal malignant melanoma is very rare and its imaging manifestations are variable, it mainly needs to be distinguished from intraspinal schwannoma and meningioma. In terms of location, morphology, and growth mode, ISPMNs are easily confused with meningioma and schwannoma. All three are common in the thoracolumbar intraspinal region. There are multiple predominant extramedullary subdural lesions. Melanomas mostly originate around the nerve root and can grow into the "dumbbell type" of the intervertebral foramen, leading to enlargement of the intervertebral foramen and destruction of bone, which are common imaging features of schwannomas. Some ISPMNs are closely related to the dura, grow by "creeping" along the dura and connecting to the broad base, exhibit an enhanced visible "dura" tail sign and show the characteristics of meningioma. The differences among the three imaging modalities mainly depend on the signal intensity characteristics of the three signals: ① Meningioma T1-weighted imaging (T1WI) signal is equal to or slightly greater than the spinal cord signal, reinforcement is obvious and uniform, and tumor calcification can exhibit a low signal. ② Schwannoma T1-weighted imaging (T1WI) signal is low, T2WI signal is slightly greater, and tumors are prone to cystic changes and uneven signals and
uneven mixed enhancement. ③ Typical melanin-containing granule tumors exhibit "short T1 high signal and short T2 low signal"[13].

Treatment
The best treatment may be total surgical resection of the lesion as much as possible, and radiotherapy is usually recommended after surgery[13-16]. Patients with intraspinal melanoma that causes neurological dysfunction should be removed as soon as possible to relieve spinal cord compression. However, the following factors limit the extent of resection: ① the onset is insidious, and the tumor volume is often large; ② the tumor is well located in the upper cervical spinal cord, inferior thoracic segment around the nerve root, and closely adhered to important nerve structures; ③ the tumor is highly vascular and prone to bleeding; and ④ few tumors have focal infiltration, which can invade the medulla. Whether there is radiotherapy for residual tumors has been controversial. With surrounding tissue infiltration in the spinal canal of malignant melanoma, total resection is difficult. Timely adjuvant chemoradiotherapy after surgery is important for prolonging survival and reducing local recurrence and implant metastasis. Metastasis of other extraspinal organs after surgery for intraspinal malignant melanoma is rare.

CONCLUSION
Finally, primary intraspinal malignant melanoma is extremely rare, with a high degree of malignancy, no clinical specificity, and large differences in imaging manifestations, which ultimately depend on pathological diagnosis[18]. At present, surgery supplemented with postoperative radiotherapy and chemotherapy is the first-line treatment.
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