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Primary Spinal Meningeal Melanocytoma[1]

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Primary meningeal melanocytic neoplasms are rare lesions that originate from leptomeningeal melanocytes. An intradural meningeal melanocytoma in the thoracic spine is less common than a malignant melanoma, which is its malignant counterpart. We report a case of a histopathologically confirmed primary intradural meningeal melanocytoma in the thoracic spine along with a literature.

Index words : Spine
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Primary meningeal melanocytic neoplasms are rare lesions that arise from leptomeninges, which in certain regions normally contain melanocytic cells. A primary meningeal melanocytoma is the less common benign counterpart of a malignant melanoma (1-4). The MR imaging findings of meningeal melanocytoma have a relatively typical appearance, owing to the paramagnetic effect of melanin and hemorrhage. However, the differential diagnosis for a meningeal melanocytoma is often confusing due to its similar appearance with other melanotic neoplasms or tumoral hemorrhage. Since the biological behavior, treatment, and prognosis of these lesions are different, it is important to make a correct pathological diagnosis.

We describe here the MR findings of a primary intradural melanocytoma case located in the thoracic spine with accompanying pathological and immunohistochemical analysis features.

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Case Report

A 58-year-old male presented with a several week history of weakness in both lower legs and voiding difficulty. The neurologic examinations indicated slight paraparesis without sensory disturbance.

MR imaging was performed at a 1.5T MR imaging unit (Magnetom Vision, Siemens Medical Systems, Erlangen, Germany), and indicated the presence of a well-demarcated, nodular intradural extramedullary mass at the thoracic level. The lesion was attached to the underlying dura and showed increased signal intensity compared to the intervertebral disc on T1-weighted images (repetition time msec/echo time msec, 580/12) and decreased signal intensity with dark margin on T2-weighted images (repetition time msec/echo time msec, 3600/110) (Figs. 1A, B). Marked spinal cord compression was also noted on an axial image (Fig. 1C).

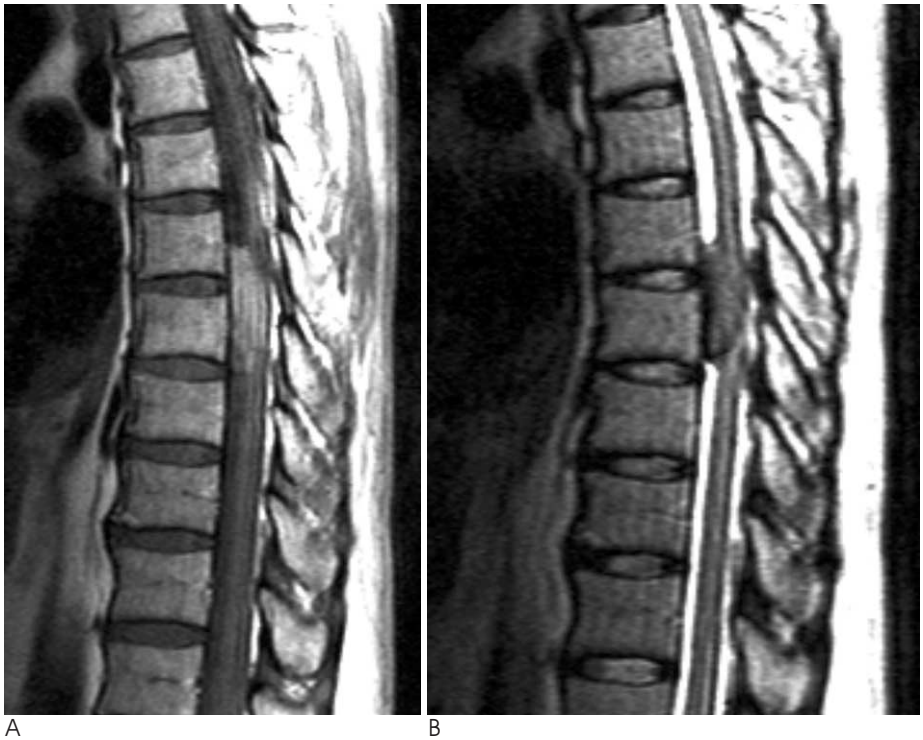
The initial differential diagnosis included meningioma and schwannoma, based on the tumor involvement site and MR imaging signal intensity. Unfortunately, the possibility of a meningeal melanocytoma was not considered.

An elective bilateral total laminectomy with tumor re-

section was performed at the T6–7 level. The surgeon observed a soft dark brown colored mass that was firmly attached to the underlying arachnoid membrane. The mass was also noted to have severe adhesion with the spinal cord. A few small pieces of tissue from the mass were submitted as frozen sections. The diagnosis of the frozen section was a neoplasm with melanin pigment, suggestive of a melanocytoma; however, a malignant melanoma could not be ruled out.

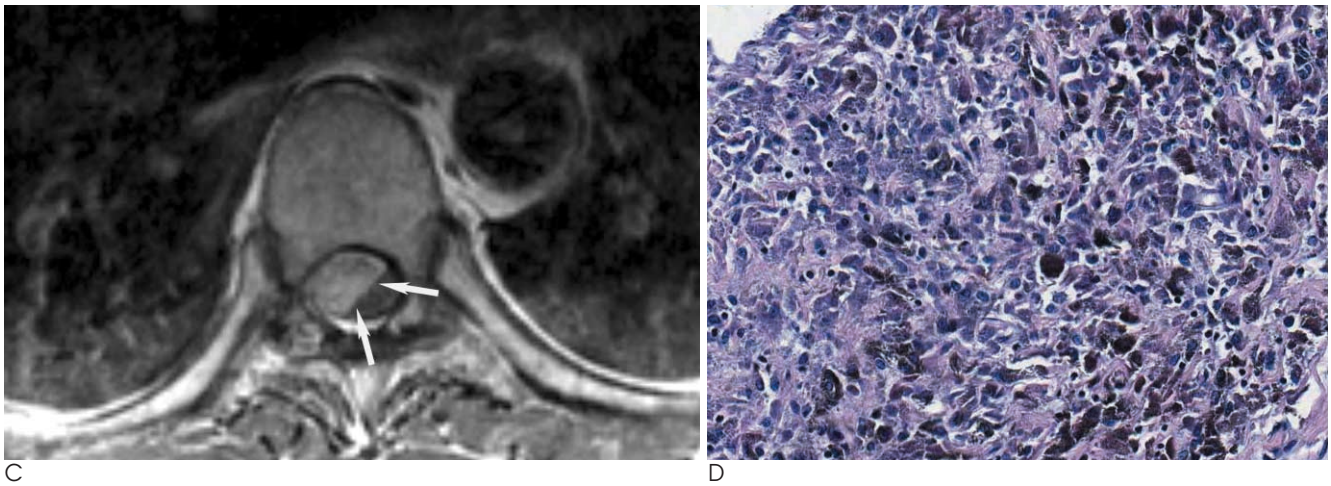
A histological analysis of the mass demonstrated the presence of a relatively cellular neoplasm composed of spindle or cells with an epitheloid appearance and cytoplasmic granular dark brown pigment, which is suspicious for the presence of melanin (Fig. 1D). The results of immunohistochemical staining indicated that the tumor cells were positive for S-100 protein and HMB-45. This finding suggests that the neoplasm is of melanocytic origin, an example being a meningeal melanocytoma or a malignant melanoma (Figs. 1E, F). The tumor showed no necrosis, no hemorrhage, no mitotic activity, no significant cellular atypia, and a very low Ki-67 labeling index (data not shown). As a result, the tumor was diagnosed as a melanocytoma rather than a malignant melanoma suggested by the histological findings and immunohistochemaical staining.

After surgery, the patient underwent additional radiological studies including PET-CT as well as a dermatological and ophthalmological examination, which did

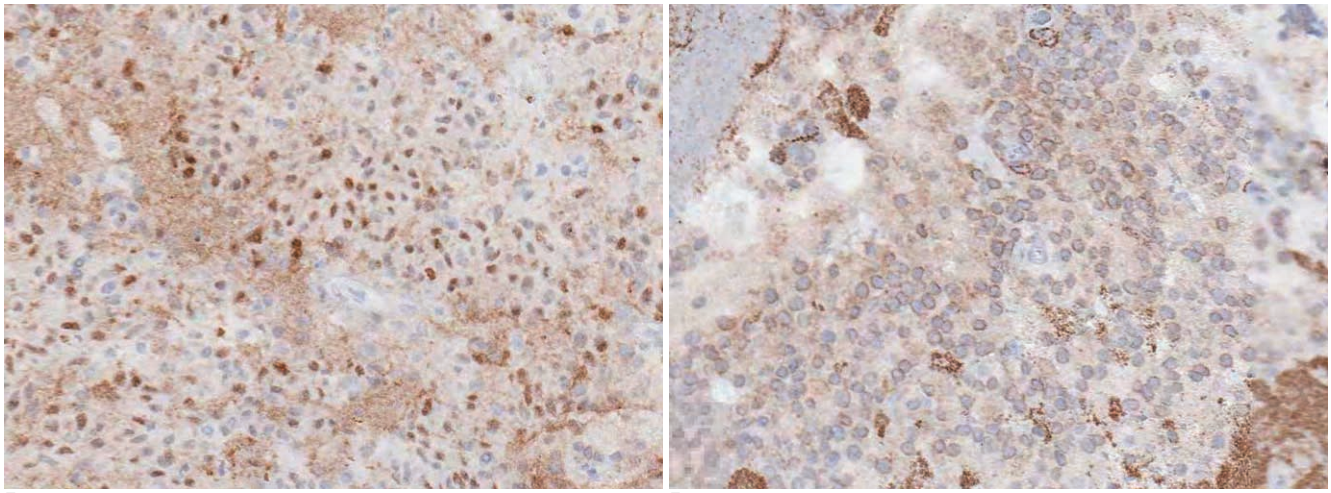


A B

Fig. 1. Imaging findings of a 58-year-old male with weakness in both lower legs and voiding difficulty.
A. A sagittal non-contrast T1-weighted image shows the presence of a well-marginated, high signal intensity, intradural mass at the T6–7 level.
B. A T2-weighted image depicts a low signal intensity mass with a darkened margin. The mass shows close contact with the dura and the spinal cord.
C. An axial T1-weighted image reveals marked spinal cord compression caused by the mass (arrows).
D. The tumor is composed of spindle to ovoid cells without significant cellular atypia or cytoplasmic granular dark brown pigment, which is suspicious for melanin (H & E staining after bleaching of melanin pigment, ×200). The tumor does not show a focal hemorrhage foci.



C D



E F

Fig. 1. E. The tumor cells show nuclear staining for S-100 based on immunohistochemical staining (×200).
F. The tumor cells show cytoplasmic reactivity with HMB45 based on immunohistochemical staining (×200).

not reveal any other melanoma foci.

Discussion

The majority of melanotic neoplasms in the central nervous system (CNS) are metastatic in origin (1). In rare cases, primary melanocytic neoplasms are derived from leptomeningeal melanocytes, which have a neuroectodermal origin as do cutaneous melanocytes. Leptomeningeal melanocytes are most commonly found in and around the ventral portion of the brain stem and spinal cord (5). The majority of reported cases of melonocytic neoplasms are located at the posterior cranial fossa or the spinal canal (6).

Primary melanocytic neoplasms are generally divided into three main types: 1) diffuse melanosis, 2) meningeal melanocytoma and, 3) malignant melanoma. Meningeal melanocytoma and primary malingnant melanoma of the leptomeninges are similar in origin to leptomeningeal melanocytes, but actually represent both ends of the spectrum in terms of lesions that are benign in appearance and behavior, to lesions that are malignant (7). The prognosis of meningeal melanocytoma is quite good in most cases. However, local recurrence has been observed (4). These entities should not be associated with other pigmented lesions in different locations, including benign congenital pigmented nevi or frank cutaneous malignant melanoma (7, 8).

Jellinger et al. (2) have summarized 16 cases of meningeal melanocytoma, including 15 previously reported cases and one novel case. The study indicates that meningeal melanocytoma could occur at any age (age range, 9 to 71 years), with no predilection for sex. Spinal canal melanocytomas could occur at any level with intradural localization and could arise in both intradural and extradural compartments.

The gross appearance of meningeal melanocytoma as seen on surgery or during an autopsy is that of a single, well-encapsulated, nodular, dark brown or black lesion, which is firmly attached to the underlying leptomeninges (7). The lesions have a tendency to compress the adjacent tissue, but there has been no evidence of CNS tissue invasion at the time of the initial diagnosis or surgery (2).

Microscopically, the tumors are cellular, composed of fusiform or polygonal cells arranged in fascicles, but have a tendency for a whorl formation (2). Tumor cells do not have anaplasia, necrosis, or significant mitotic activity. The cells show varying degrees of melanization (7).

Immunohistochemical staining has been extensively used to differentiate melanomas from lesions that mimic melanomas in conventionally stained sections. Meningeal melanocytoma is characterized by a positive immunohistochemical reaction to S-100 protein, to antibody HMB-45, and to vimentin and by way of a negative reaction to epithelial membrane antigen (EMA). A melanocytic meningioma is characterized by having a positive reaction to EMA and vimentin, and by having a negative reaction to S-100 protein and antibody HMB-45 (2, 3, 7).

The histological differentiation between melanocytoma and malignant melanoma can be very difficult. The features which differentiate melanocytoma versus

malignant melanoma include a lack of mitotic activity, nuclear pleomorphism, and hyperchromaticity, as well as the indolent growth of a mass that spans more than four years (7). Ki-67 staining has been commonly used as an adjunct to distinguish benign from malignant melanocytic tumors. Ki-67 staining has been reported as being positive in 13–30% of cells, for malignant melanoma, although individual cases can show almost 100% nuclear positivity (9).

MR imaging of meningeal melanocytoma characteristically reflects the paramagnetic effect of melanin, which causes shortening of the T1 and T2 relaxation times. Therefore, the MR appearance of these lesions generally includes an increase in signal intensity on T1-weighted images and a decrease in signal intensity on T2-weighted images (7). This signal pattern is unique, but the imaging pattern may correspond to that of other pigmented tumors such as malignant melanoma, melanotic meningioma and melanotic schwannoma, as well as a tumoral hemorrhagic lesion (4, 7, 8). Furthermore, pigmented tumors may show a variable appearance on MR images due to the presence of different degrees of melanin or hemorrhagic foci (7, 8). A nonhemorrhagic amelanotic melanocytoma has a iso-signal intensity or mildly decreased signal intensity on T1-weighted images, and an iso-signal intensity or mildly increased signal intensity on T2 weighted images (4). Thus, the MR imaging appearance is of limited value in the differential diagnosis.

The MR imaging appearance of a primary melanocytoma also may be similar to that of metastatic melanomas; therefore, careful examinations of skin, squamous mucosa, and eyes must always be performed to exclude the possibility of metastatic melanomas in these sites (8).

Although unusual, when an intradural extramedullary tumor with increased signal intensity on T1-weighted images and decreased signal intensity on T2-weighted images is detected, meningeal melanocytoma should be considered in the differential diagnosis.

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하 동 호

일차성 수막성멜라닌세포종은 연수막에 정상적으로 분포하는 수막성 멜라닌 세포로부터 발생하는 아주 드문 양성 종양이다. 저자들은 흉추의 경막 내 척수 외 공간에서 발생한 일차성 수막성멜라닌세포종을 경험하였기에, 문헌고찰과 함께 보고하는 바이다.