Case Report

Intramedullary Spinal Cord Metastatic Plasmacytoid Melanoma: Case Report and Literature Review

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Melanoma is melanocytic tumour that can be primary or metastatic. Though the metastatic melanoma is the third most common of the central nervous system metastases, the metastatic intramedullary melanoma is rare and diagnostic challenges. Melanoma is the tumour that closely mimics other tumour due to variations including morphology, architecture, stromal component and immunohistotype which are similar to those found in other tumours. The diverse variants of melanoma result potential diagnostic pitfalls in histological assessment. The authors reported a case of intramedullary spinal cord metastatic melanoma, plasmacytoid variant which had a diagnostic challenge to the pathologist involved.

Keywords: Spine intramedullary tumour, Melanoma, Plasmacytoid variant

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Melanoma is prone to early distant metastasis involving almost any organ or structure. Following the development of a distant metastasis, the prognosis is very poor. The most common form of melanoma of the spine usually bones such as vertebral metastasis, whereas intramedullary spinal cord metastasis is rare. Gokaslan et al reported 133 cases of vertebral metastatic melanoma over a period of 11 years and found this was a late event in the evolution of the disease[(1)]. Intramedullary spinal cord metastatic melanoma is very rare and Ishii et al found only 9 cases on reviewing the literatures[(6)]. Immunohistochemical studies are also important in diagnosis especially in unusal melanoma variants. The immunohistochemical studies may be used to distinguish spinal melanoma from other types of tumours.

Case Report

A right-handed-67 year old man presented with quadriparesis. The night before, he had developed severe neck pain and woke in the morning to tingling and mild heaviness of the right upper limb then rapidly spread to the left upper and lower extremities, and the right lower limb. Asymmetric weakness was greater on the right. He had no report of paresthesia. Patient did not provide history of any previous cancer. Physical examinations revealed a pleasant man with regular vital signs, no lymphadenopathy, no Horner's syndrome and no nystagmus. The cranial nerve examination was unremarkable with full ocular movements and no facial asymmetry or weakness. Neck movements were not restricted but there were possible signs of Lhermitte's. He had complete paralysis of the right upper limb except for shoulder shrugging. The motor strength of his lower extremities and left upper limb were grade II-III. There was global areflexia with bilateral Babinski signs. Pinprick sensation was impaired with graded sensory level to above the T2 level. Propioceptive senses were abnormal in the right upper and lower extremities.

The investigations included CT brain, MRI brain and spine scan, and lumbar puncture. The CT scan showed a lesion out of the right temporal horn at the lateral ventricle measuring 2.3 x 1.6 x 1.4 cm. with minimal mass effect, suspicious of solid mass or capsular lesion. The gadolinium-enhanced MRI of the brain and cervical spine revealed an extensive lesion to the upper cervical cord which appeared to be a solid lesion with central enhancement but did not appear to be inflammatory or vascular in etiology (Figure 1-2). The lesion to the right temporal horn of the lateral ventricle differed in imaging appearance from the spinal cord lesion. Lumbar puncture and CSF profiles combined with the MRI findings did not support an inflammatory process.

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At operation, there were two tumours, one at C3 and the other at C5 levels. The C3 was larger and compressed the cord and intramedullary, (Figure 1). The C5 lesion was extramedullary appeared at neural foramen, (Figure 2). A laminectomy was carried out at the level of C3 - C7 for spinal decompression and a biopsy was taken of the C3 intramedullary spinal tumour for pathological diagnosis. The microscopic sections of the biopsy prepared with H&E and immunostains were reviewed. The lesion from the intramedullary C3 biopsy revealed sheets of tumour cells composed of small to medium sized, round to polygonal-shaped cells, with moderate eosinophilic cytoplasm, irregular nuclear membrane, dense nuclear chromatin and inconspicuous nucleoli. Several cells had eccentric nuclei with perinuclear clearing. Scattered tumor cells contained brown granular pigments in their cytoplasm. Mitoses was rarely seen (0 - 1 mitotic figures per 100 tumor cells) and necrosis was absent. Both Perl's Prussian blue and Masson Fontana were performed and melanin pigments were identified. The PAS and reticulin highlighted the vascular channels. Although the possibility of a metastatic melanoma was entertained, but because of the plasmacytoid appearance of tumour cells, plasma cell neoplasm and plasmacytoid lymphoma were included in the differential diagnosis on the initial examination.

The CD3, CD20, and CD45 were positive for reactive lymphocytes. Tumour cells were negative for CD3, CD20, CD45, CD57, CD68, CD138, MPO, EMA, CKCAM5.2, synaptophysin, chromograninA and GFAP. The tumour cells were weakly positive for CD99, focally positive for MelanA (MART-1) and strongly positive for HMB45, S100 protein and vimentin. The Ki-67 was positive in 4% - 6% of tumor nuclei (Figure 3-8).

One week after spinal cord biopsy and decompress, a mass was discovered in the liver and needle core liver biopsy was performed. The sections from the liver biopsy showed tumour with similar histopathological features to those of the C3 intramedullary tumour. Mitotic figures were rare. Immunohistochemical study revealed that tumour cells were strongly positive for HMB45, S100 protein and MelanA (MART-1). Tumour cells were negative for CD45, C-kit (CD117), cytokeratin, TTF-1 and HSA. The Ki-67 was positive in about 9% of tumour nuclei and pHH3 showed 0 - 1 mitotic figure per 100 tumour cells.

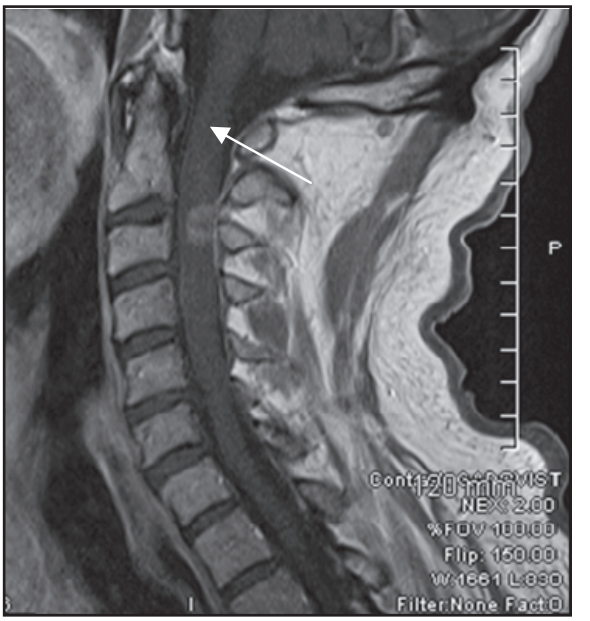

Figure 1. The Zone of enhancement measures 1.2 x 0.9 x 1.1 cm. at C2/C3 level and superior C3 Level. (Sagittal T1 weighted imaging with contrast).

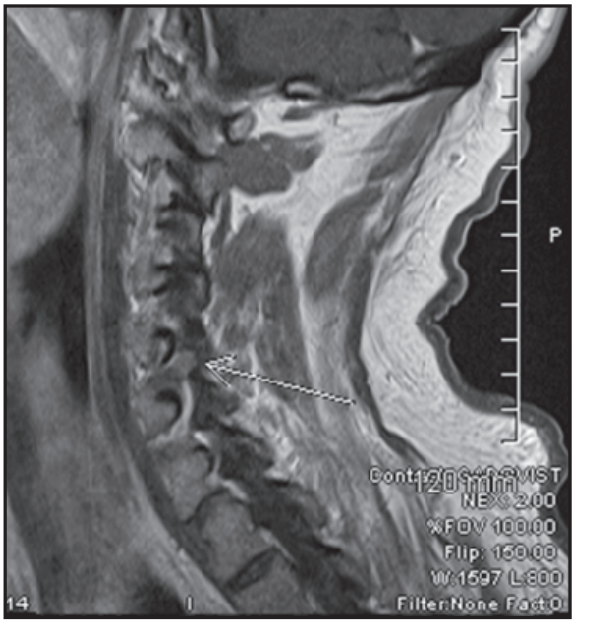

Figure 2. Mildly enhancing intermediate signal lesion noted at the origin of the right C5/C6 neural foramen measures 0.4 x 0.5 x 0.6 cm. (Sagittal T1 weighted imaging with contrast).

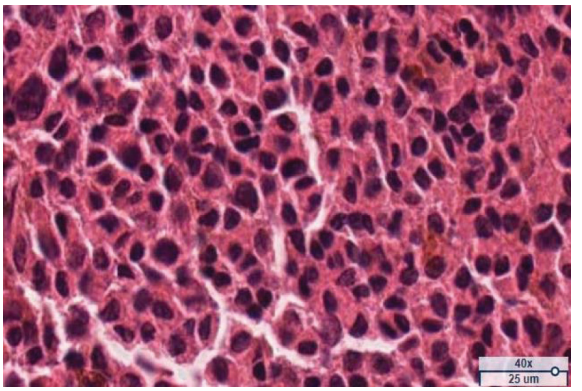

Figure 3. Tumour cells are small size with dense and eccentric nucleus (H&E 400X).

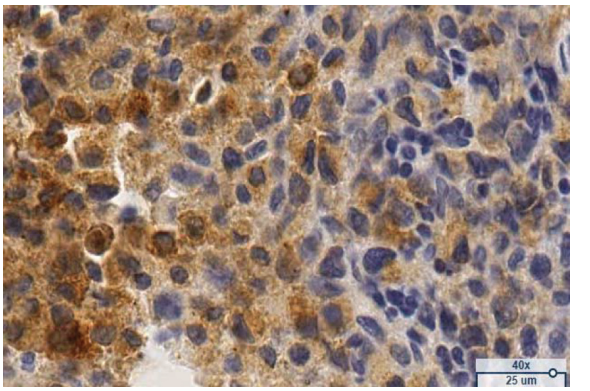

Figure 4. Tumour cells are scanty and focal cytoplasmic staining for Melan-A (400X).

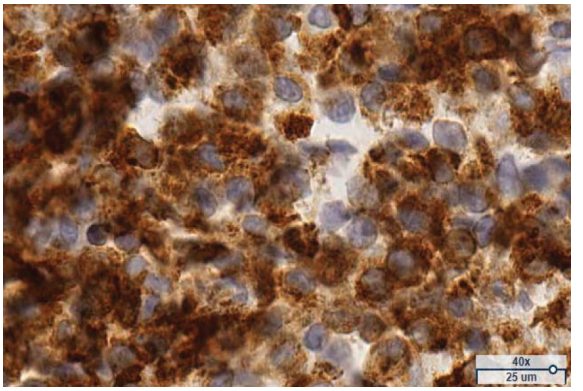

Figure 5. Tumour cells are strongly positive for HMB45 (400X).

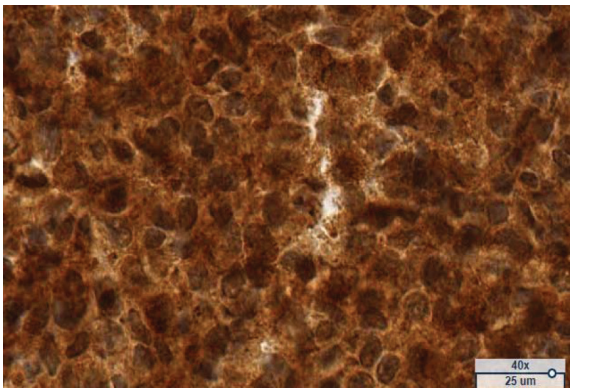

Figure 6. Tumour cells are strongly positive for S100 protein (400X).

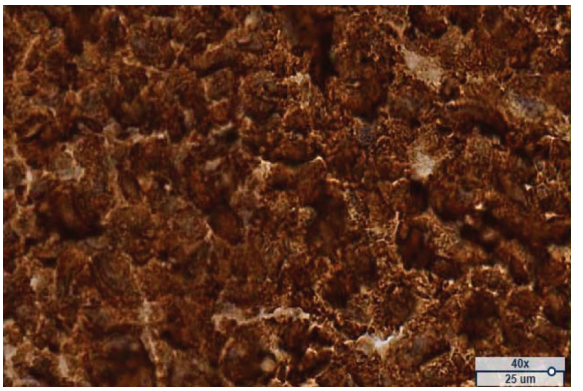

Figure 7. Tumour cells are strongly positive for vimentin (400X).

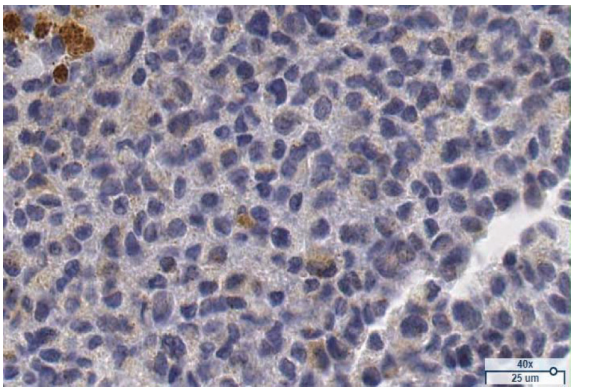

Figure 8. Few preexisting plasma cells are positive for CD138, iron pigments and melanin pigments are brown (400X).

Discussion

Intramedullary spinal cord metastasis is a rare complication of cancer and equally effected the cervical, thoracic and lumbar spine. Motor weakness is the most common symptom at presentation, followed by pain and sensory disturbance[7]. Intramedullary spinal cord melanoma metastasis is very rare and melanoma has many variations in morphology

subtype. An accurate diagnosis during intraoperative consultation and on routine light microscopy without immunohistochemistry can be difficult and challenging[9,10]. The plasmacytoid variant of melanoma is a rare finding which may mimic many other entities, especially plasma cell neoplasm. The use of immunohistochemistry is crucial to the initial diagnosis[4,8]. In our case, a positive staining for melanoma markers (MelanA, HMB45, S100 protein) distinguished the metastatic melanoma from a plasma cell neoplasm. The plasmacytoid variant of melanoma shows negative immunoreactivity for plasma cell marker (CD138) though the histopathological features by H&E look like plasma cells.

In conclusion, the authors have presented a rare case of intramedullary metastatic melanoma of spinal cord with no history of a known primary. The plasmacytoid features of this metastasis has further complicated the diagnosis on routine sections and only with awareness of the possibility of this plasmacytoid variant in melanoma that the correct immunohistochemical panel can be performed to confirm the diagnosis.

What is already know on this topic?

Metastatic melanoma to central nervous system is frequently found but the intraspinal cord metastatsis is very rare. Due to many variations of the melanoma, it mimics other tumours. The pathological diagnosis is challenged to pathologist.

What this study adds?

The plasmacytoid variant of melanoma may mimic plasma cell neoplasm especially in the case which no any previous history of cancer. The immunhistochemical panel is important for the accurate diagnosis.

Potential conflicts of interest

The authors declare no conflict of interest.

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