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J Cancer Allied Spec 2024;10(1):1

Case Report

A Rare Case of Primary Malignant Melanoma of Cervical Spine Having Extramedullary Intradural Origin: A Single Case Report and Literature Review

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Received: 09 December 2022/Accepted: 29 November 2023

OPEN ACCESS

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Citation: Haider I, Khan S, Ul Qamar F, Inam Y, Gul K, Rana IA. A Rare Case of Primary Malignant Melanoma of Cervical Spine Having Extramedullary Intradural Origin: A Single Case Report and Literature Review. J Cancer Allied Spec [Internet]. 2024;10(1):1-5. https://doi.org/10.37029/jcas.v10i1.513

Funding: None.

Competing interest: Nil.

Abstract

Introduction: Primary spinal malignant melanoma (PSMM) of extramedullary intradural origin is a rare malignant condition with limited current literature regarding its clinical course, magnetic resonance imaging (MRI) findings, treatment strategies, and outcomes. **Case Discussion:** This is a case report of a patient with PSMM who was treated with surgery followed by radiotherapy for his residual disease in Shaukat Khanum Memorial Trust, Pakistan. The clinical and radiological findings of this case were retrospectively analyzed using the Hospital Information System. **Practical implementations:** PSMM of extramedullary intradural origin is a rare malignant tumor that shows characteristic findings on MRI. Surgical resection is the preferred treatment, and radiotherapy is useful for residual disease.

Key words: Cervical spine, extramedullary, intradural, malignant melanoma

Introduction

Primary melanomas of the central nervous system (CNS) are one of the least found melanomas, attributing to only 1% of all cases of melanomas. The spinal location is even less frequent.[1,2] The first-ever report of primary spinal malignant melanoma (PSMM) was by Hirschberg in 1906.[3] The thoracic segment (42.3%) is the most found site, followed by the cervical (34.6%), thoracolumbar

(11.5%), cervicothoracic (7.7%), and lumbar (3.8%) segments.[4]

The primary melanoma of the cervical spine is a rare type. In the English literature, only two cases of primary extramedullary intradural malignant melanoma of the cervical spine and three cases of primary malignant melanoma arising from cervical nerve roots have been reported.[5] This type of cancer originates from melanotic cells of the leptomeninges, imitating the general appearance of other nerve sheath tumors or meningiomas.[5] On magnetic resonance imaging (MRI) scans with contrast, the cancer usually gives hyper-intense signals in T1 and hypointense in T2.[5-7] Most encountered symptoms are pain and progressive weakness for weeks to months with or without asymmetric myelopathic symptoms.[7]

This case report presents a unique PMSS instance observed and diagnosed at a tertiary care cancer center. The diagnosis was established with the help of a thorough clinical assessment, advanced imaging techniques, and detailed histopathological analysis.

Case Description

A 21-year-old man visited the radiation oncology outpatient clinic after undergoing surgical excision of an intradural extramedullary mass in his cervical region. Before his surgery, he had been experiencing neck pain and right-sided body weakness for the past 2 years. His symptoms had gradually worsened over time. He underwent an MRI scan of his spine, which showed the presence of an intradural extramedullary mass in the cervical region outside of the hospital.

Diagnosis and Management

Histopathological examination revealed a spindle cell tumor with moderate atypia, hyperchromatic nuclei, occasional prominent nucleoli, and scattered mitoses. In the background, melanin pigment was also seen [Figure 1a]. We performed three immunohistochemical stains, including

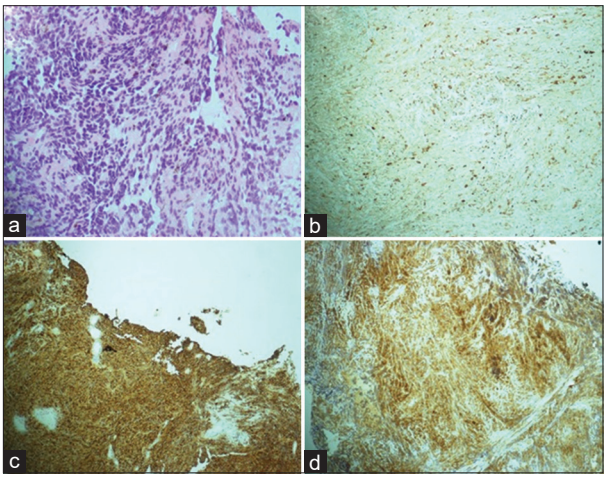

Figure 1: (a) Photomicrograph shows oval to spindle-shaped tumor cells with hyperchromatic nuclei. A few areas of melanin pigmentation are also visible. (H&E, ×200). (b-d) Positive immunoreactivity for S100 protein (×200), HMB-45 (×200), and Melan-A (×200)

S100, HMB-45, and Melan-A [Figure 1b-d]. All these stains were positive in tumor cells. Based on immunophenotype, the differential diagnosis was malignant melanoma and clear cell sarcoma. To rule out clear cell sarcoma, fluorescence *in situ* hybridization for EWSR1 gene rearrangement was performed, which was not detected, and a diagnosis of malignant melanoma was made.

At the time of presentation, the patient reported marked improvement in all his symptoms except pain behind the neck. General physical examination was unremarkable, and CNS examination showed no motor or sensory deficit. He underwent a post-operative MRI brain with the whole spine, which showed a small area of signal void measuring approximately 0.8 cm, projecting in the cervical canal posteriorly, almost in the midline at the C3 level [Figure 2a and b]. His positron emission tomography-computed tomography (PET-CT) scan and dermatological and ophthalmic examinations were also negative for any suspected lesion. His case was discussed in the multidisciplinary neuro-oncology meeting, and all members agreed upon the diagnosis of primary malignant melanoma of the spinal cord because there was no other melanotic lesion found.

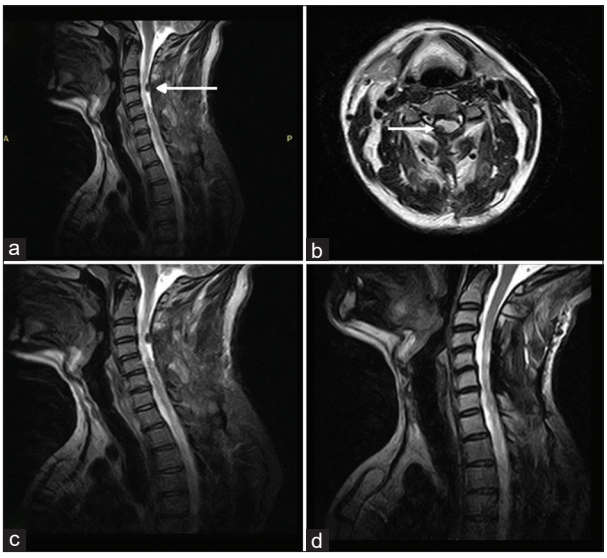

Figure 2: (a) is the sagittal view of the magnetic resonance imaging (MRI) scan of the cervical spine, while (b) is the axial view of the same. Both images display a small 8-mm area of signal void in the cervical canal, located posteriorly and almost at the midline of the C3 level (indicated by a white arrow). (c and d) show the pre- and post-radiotherapy MRI scans of the cervical spine in sagittal view

Since this patient had already undergone surgery, it was deemed unfit to plan him for such an invasive procedure again for a single focus of 8 mm. Risks of operative and post-operative complications were evaluated against the benefits, and it was decided to proceed with post-operative radiotherapy. The patient was treated with 45 Gy (Gray) in 25 fractions at 180 cGy per fraction. Radiotherapy was completed uneventfully, and it was tolerated well with no significant toxicities.

Post-treatment MRI of the brain and spine of the patient showed complete resolution of previously seen focus, and there were only post-intervention changes [Figure 2c and d]. After 1 1/2 years of his treatment, the patient had no concerning symptoms. For future follow-up, he will be seen every 3 months for clinical examination and 6 months with an MRI brain and spine.

Discussion

Primary melanomas of the CNS are rare, accounting for only 1% of all melanomas. Similarly, a spinal site is the least expected location.[2] The World Health Organization classifies primary melanocytic lesions of the CNS into diffuse melanocytoma, melanocytosis, meningeal melanomatosis, and malignant melanoma. Hirschberg first published primary spinal melanoma in 1906, accounting for 38% of all primary CNS melanomas. Merely 40 cases of spinal cord primary melanoma have been identified in the literature since 1906. This very rare malignancy, the primary malignant melanoma of the spinal cord, usually occurs in the middle or lower thoracic portion of the spinal cord.[1-5] Previously, only nine tumors have been reported in the cervical spine region.[6] As per the Hayward classification, primary spinal cord melanoma is diagnosed when there is the absence of melanoma outside the CNS or in other sites in the CNS; histological features also confirm it. In our case, no evidence of melanoma was found elsewhere, as clinical examination, ophthalmological examination, and PET-CT scan were negative for any other lesions.

Primary pigmented tumors of the CNS are rare and include melanotic schwannoma, meningeal melanocytoma, blue nevus of the CNS, and primary melanoma.[3,6,7] Primary melanoma of the CNS may arise from melanoblasts accompanying the pial sheaths of vascular bundles or from neuroectodermal rest cells during embryogenesis.[1,2,7] The tumor is equally seen both in males and females. The most common age groups affected are the fifth decade. However, they may present between the ages of 20 and 80 years.[4] This case was a 21-year-old male.

Clinical symptoms are usually non-specific.[8,9] Symptoms exhibited are predominantly those of spinal cord compression.[2,3,6] Pain is the most common complaint, at times accompanied by progressive weakness for weeks or months or asymmetric myelopathic symptoms. Our patient, before his surgery, had neck pain and right-sided body weakness for 2 years. It was gradual in onset and progressive in nature. However post-surgery and at the time of presentation to us, he had marked improvement in his symptoms. MRI is considered the gold standard for diagnosing

spinal cord tumors. Nevertheless, it is somewhat difficult to discriminate between tumors based on their morphologic or signal intensity characteristics at MR imaging.[3]

MRI of PSMM of extramedullary intradural origin usually shows typical changes. Usually, it presents with hyper-intense T1W signals, hypo-intense T2W signals, and varying enhancements. The characteristic MRI findings may be associated with melanin concentration and hemorrhages. In CNS melanoma, this typical sign was observed in less than half of the patients.[10] However, the typical features were absent in our review of PSMM of extramedullary intradural origin. The main reason was post-operative presentation.

Histopathological diagnosis of malignant melanoma is usually straightforward. However, this may present a diagnostic dilemma at odd locations like in this case. First, dermatological, ophthalmic, and mucosal examinations should be carried out to rule out primary lesions in these locations. Second, other entities, including clear cell sarcoma, should also be considered in the differential diagnosis due to differences in the treatment options. To rule out/rule in clear cell sarcoma, fluorescence *in situ* hybridization for EWSR1 gene rearrangement is a better diagnostic modality.

No cases have been reported from Pakistan at all; however, in the available literature, we did not find a case with extramedullary intra- and extradural primary cervical spinal melanoma with residual disease post-surgery treatment with radiotherapy. The most common site for metastasis of these tumors is the primary site and/or within the CNS.[8] PMS usually exhibits less aggression and slow progression compared to other common melanomas of the skin with metastases to the CNS. Treatment options for spinal cord malignant melanoma are limited. Surgical excision of the lesion is considered a treatment of choice. Radiation therapy is often recommended after surgery.[2,6,7] Chemotherapy may be used with radiotherapy during the post-operative period, but its role is controversial.[3,6,7] The median survival after surgical treatment +/- radiotherapy is 6 years.[7,8] As seen in our case, the patient exhibited residual disease post-surgery. However, after successful radiotherapy, the patient could return to his routine life with no disease recurrence or neurological deficits. His post-radiotherapy imaging showed complete resolution of residual disease. He was last seen after 1½ years of his treatment and reportedly had no concerning symptoms.

In conclusion, extramedullary-origin PSMM is very rare. Pre-operative diagnosis is usually based on standard MRI presentations. Surgery is considered the primary therapeutic modality for these tumors. Nonetheless, additional post-operative radiotherapy may be helpful depending on the case presentation and disease status.

Acknowledgment

None.

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Author contributions

Conceived and designed the analysis: IH. Collected the data: YI, KG, and SK. Contributed data or analysis tools: FQ, YI, and IAR. Performed the analysis: IH, FQ, and IAR. Wrote the paper: IH, FQ, SK, IAR, and KG.