Yonsei Medical Journal
Vol. 45, No. 2, pp. 345 - 348, 2004

Primary Malignant Melanoma of the Cervical Spinal Nerve Root

Soon-Chan Kwon[1], Seung-Chul Rhim[1], Deok-Hee Lee[2], Sung-Woo Roh[1], and Shin-Kwang Kang[3]

Departments of [1]Neurosurgery, [2]Radiology, [3]Pathology, Asan Medical Center, University of Ulsan, College of Medicine, Seoul, Korea.

The authors report on a case of primary malignant melanoma of the 7th cervical spinal nerve root in a 45-year-old woman. Neuro-radiological features of this extra-dural mass were suggestive of a nerve sheath tumor. The lesion underwent total gross resection through the anterolateral approach. The patient's postoperative course was uneventful. Histopathological investigation confirmed malignant melanoma. There was no evidence of tumor recurrence or other melanotic lesions on regular follow-up examinations until the postoperative eighth month. When treating a common, benign-looking lesion of the cervical spinal nerve root, surgeons should be aware of the potential to encounter such a malignant tumor.

Key Words: Primary malignant melanoma, cervical spine, nerve root

INTRODUCTION

Primary malignant melanoma in the central nervous system (CNS) accounts for approximately 1% of all cases of melanoma.[1] Primary spinal melanoma was first reported by Hirschberg in 1906, and accounts for 38% of all CNS melanoma.[2] However, primary melanomas in the spinal nerve roots are extremely rare, especially in the cervical nerve roots. On review of the literature, only one case of melanoma arising in a cervical spinal nerve root was reported.[3] We present an additional case of primary melanoma arising at and limited to the cervical nerve root.

Received October 1, 2003
Accepted October 31, 2003

Reprint address: requests to Dr. Seung Chul Rhim, Department of Neurosurgery, Asan Medical Center, University of Ulsan, College of Medicine, 388-1 Pungnap-2dong, Songpa-gu, Seoul 138-736, Korea. Tel: 82-2-3010-3550, Fax: 82-2-476-6738, E-mail: scrhim@amc.seoul.kr

CASE REPORT

A 45-year-old woman presented with a 4 year history of severe pain and tingling sensation on the left side shoulder and arm. During the previous 5 months, her symptoms had worsened. On physical examination, a 3×2 cm sized, palpable soft mass was noted on her left, anterior neck. Neurologic examination revealed mild weakness on wrist extension and hand grasp on the left side, and diminished sensation on the C6 and C7 dermatomes. No motor weakness was detected.

Computed tomography (CT) of the cervical spine revealed an extra-foraminal, ovoid-shaped mass, which was slightly more hyperdense than the adjacent muscles. The left side neural foramen at the C6-7 level was widened by the mass. On magnetic resonance (MR) imaging, T2-weighted image (WI) showed a relatively well-demarked, purely extradural mass along the course of the left proximal brachial plexus. The main portion of the lesion showed decreased signal intensity on T2-WI and slightly increased signal intensity on T1-WI. There was a portion of different signal characteristics (bright in both T1-WI and T2-WI) at the posterolateral aspect of the lesion. Hemorrhage or necrosis of the tumor was suggested. Initial radiological diagnosis was complicated neurogenic tumor of the brachial plexus (Fig. 1).

An anterolateral approach was performed between the carotid artery and sternocleidomas-

toid muscle. Upon exposing the exit area of the C7 nerve root, a dark encapsulated tumor measuring 2 cm in its greatest dimension was detected at the ventral primary ramus of the C7 nerve root beneath the anterior scalene muscle. The tumor had darkly pigmented and muddy substance inside(Fig. 2). It did not appear to extend proximal to spinal ganglion of C7 nerve root. Mobilizing the vertebral artery medially, gross total removal of the tumor was accomplished.

Histopathological examination demonstrated a melanocytic tumor. The evidence of large nucleoli, considerable nuclear pleomorphism and the high Ki-67 labelling index (12%) suggested malignancy. The tumor cells were epithelioid and contain melanin pigments in their cytoplasm (Fig. 3). The tumor cells were immunoreactive for S-100 protein and HMB-45 (Fig. 4).

Postoperatively, her radicular pain and paresthesia disappeared. Post-operative medical, ophthalmological, and dermatological evaluations, including whole body Positron Emission Tomography (PET) scan (F-18-FDG) for primary melanotic origin, were all negative. Adjuvant radiation therapy was given postoperatively (60Gy). Regular follow-up examinations until the postoperative eighth month failed to find tumor recurrence or other melanotic lesions.

DISCUSSION

Melanoma is a malignant neoplasm of the melanocytes. Melanocytes are melanin-producing cells that arise from the neural crest during embryogenesis, and migrate to the skin, mucous membranes, and CNS. In general, melanocytic tumors in the CNS are well known as a result of metastasis. Compared to metastatic melanoma in the CNS, primary CNS melanomas are relatively uncommon lesions. Brat et al.[2] summarized 33 cases of primary melanocytic neoplasms of the CNS according to the primary site and pathological natures, and found these neoplasms presented in a spectrum ranging from well-differentiated melanocytoma to malignant melanoma.

Although a variety of melanotic neoplasms involving the spinal nerve root have been described,[4-6] reports of melanoma arising in and limited to the spinal nerve root are extremely rare. Schneider et al.[7] reported primary melanoma arising from the L3 spinal nerve root in a 68-year-old woman. They performed a wide en-bloc resection and adjuvant radiation therapy. This is first report of a melanoma arising in and limited to a spinal nerve root. Skarli et al.[3] reported a case of melanoma arising in a cervical spinal nerve root and involving the intra-dural portion. They performed a two-staged, posteriorly intra-dural and anteriorly extra-dural, surgical resection without

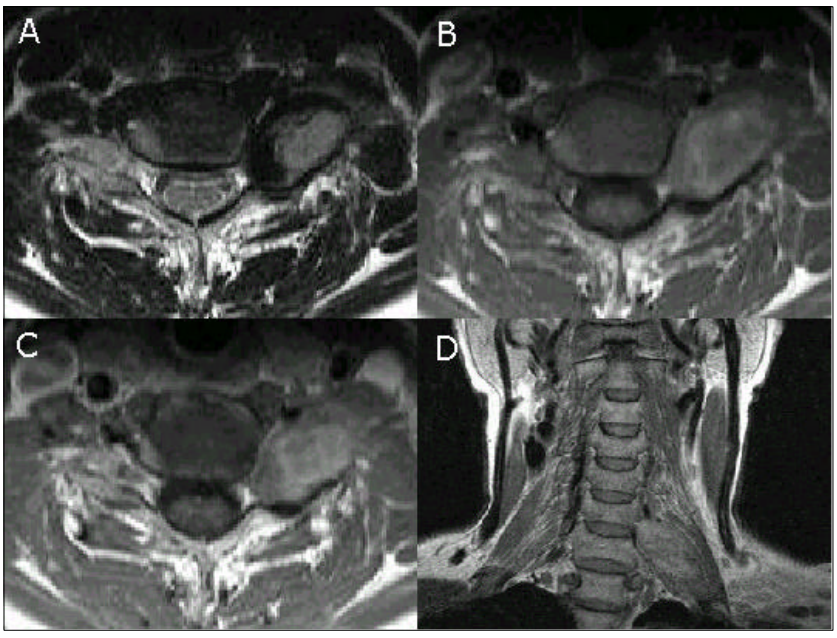

Fig. 1. Axial T2-weighted, T1-weighted, T1-contrast enhanced and coronal T1-contrast enhanced MRI scans (A, B, C & D, respectively) show a well-defined ovoid mass that is extended from the intervertebral foramen to the scalene muscles.

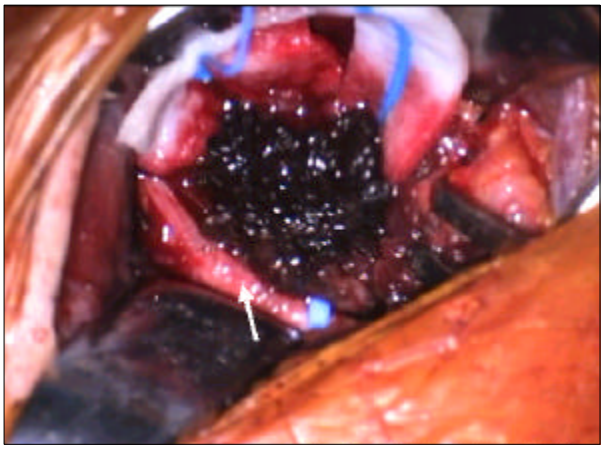

Fig. 2. Intra-operative microscopic photograph showing a darkly pigmented, muddy mass lesion. The mobilized vertebral artery is also seen in the medial side (arrow).

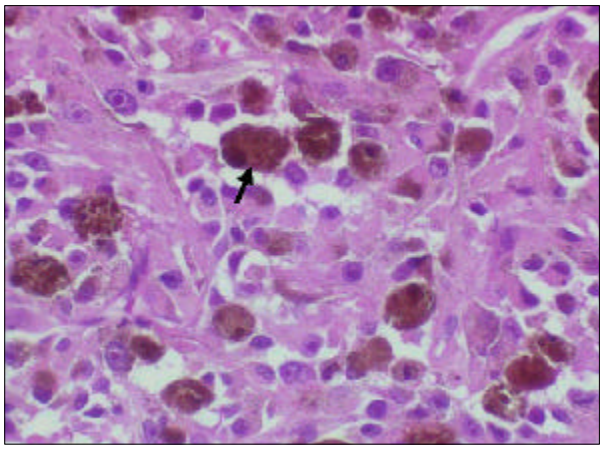

Fig. 3. The tumor cells have abundant eosinophilic cytoplasm and large macro-nucleoli. Some of them contain melanin pigments in their cytoplasm (black arrow). Hematoxylin & Eosin, × 200.

adjuvant radiation therapy. Histologically, the Schwann cell and the melanocyte share an origin in the neural crest tissues.[8] The primary melanotic neoplasms of the CNS may arise from either melanocytes or Schwann cells, and there is increased incidence of melanoma in patients with neurofibromatosis. The differential diagnosis of primary melanoma of the nerve root includes schwannoma, metastatic melanoma, melanocytoma, blue nevus, and melanotic clear-cell sarcoma. Especially, differential diagnosis from schwannoma is extremely difficult due to the rarity and the similarity in radiologic findings. On retrospective review of the CT and MR images,

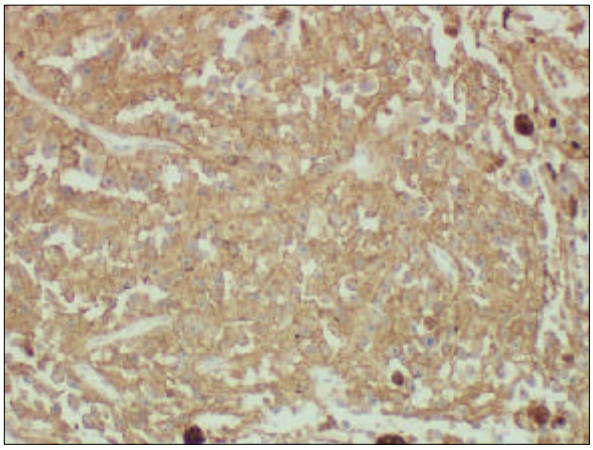

Fig. 4. The tumor cells are diffusely reactive for antibodies against HMB-45.

we realized the reason why the main mass showed unusually low signal intensity on T2-WI; it was due to the magnetic susceptibility effect of the melanin pigment. The posterolateral portion of the tumor with a different MR feature can be interpreted as necrotic focus. It is unusual for a schwannoma to be dark on T2-WI, as this mass was, although it can occasionally be hypointense to some degree. The bright signal on T1-WI can be one of the typical signal characteristics of malignant melanoma.

It is also important to differentiate whether the melanoma is primary or metastases. The prognosis for patients with metastatic melanoma to CNS is dismal, with a life expectancy of less than 1 year in most studies.[9,10] In contrast, although a number of patients with primary melanoma recur or eventually die due to melanoma, the overall long-term survival and prognosis are superior to those of metastatic lesion. In this case, metastatic melanoma was excluded because no primary pigmented lesion was found in spite of thorough physical and radiologic examination.

At the present, the prognosis of and optimal treatment for such a lesion have not been established. Neither has the role of radiation therapy in malignant primary melanoma been established. Because of the rarity of these lesions and the occasional paradoxical benign clinical course of primary CNS melanoma, further careful follow-up is needed in cases such as the one of this study.

In conclusion, the authors post-surgically con-

firmed an extremely rare case of primary malignant melanoma arising in a cervical spinal nerve root. Surgeons should be aware of the potential to encounter such a malignant tumor when treating a mass arising from the cervical spinal nerve root.

REFERENCES

1. Salpietro FM, Alafaci C, Gervasio O, La Rosa G, Baio A, Francolini DC, et al. Primary cervical melanoma with brain metastases. J Neurosurg 1998;89:659-66.
2. Brat DJ, Giannini C, Scheithauer BW, Burger PC. Primary melanocytic neoplasms of the central nervous systems. Am J Surg Pathol 1999;23:745-54.
3. Skarli SO, Wolf AL, Kristt DA, Numaguchi Y. Melanoma arising in a cervical spinal nerve root: report of a case with a benign course and malignant features. Neurosurgery 1994;34:533-7.
4. Mandybur TI. Melanotic nerve sheath tumors. J Neurosurg 1974;41:187-92.
5. McGavran WL III, Sypert GW, Ballinger WE. Melanotic schwannoma. Neurosurgery 1978;2:47-51.
6. Parker JB, Marcus PB, Martin JH. Spinal melanotic clear-cell sarcoma: a light and electron microscopic study. Cancer 1980;46:718-24.
7. Schneider SJ, Blacklock JB, Bruner JM. Melanoma arising in a spinal nerve root. J Neurosurg 1987;67: 923-7.
8. Myers JL, Bernreuter W, Dunham W. Melanotic schwannoma, Clinicopathologic, immunohistochemical, and ultrastructural features of a rare primary bone tumor. Am J Clin Pathol 1990;93:424-9.
9. Bullard DE, Cox EB, Seigler HF. Central nervous system metastases in malignant melanoma. Neurosurgery 1981;8:26-30.
10. Retsas S, Gershuny AR. Central nervous system involvement in malignant melanoma. Cancer 1988;61: 1926-34.