C 4 Nerve Root Meningeal Melanocytoma:
Confusing for Spine Surgeons?

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ABSTRACT
Melanocytic lesions of the nervous system are to be distinguished from metastatic malignant melanoma and from histologically different nervous system tumors undergoing melanization, such as schwannoma, medullablastoma, paraganglioma and various gliomas. Melanocytomas are of neural crest origin during early embryonic development and occur in normal leptomeninges. They are rare lesions arising from normally occurring leptomeningeal melanocytes. The terminology of meningeal melanocytoma was first described by Limas and Tio in 1972. Melanocytomas account for 0.06-0.1% of all brain tumors. They are most frequently found in sulci around the brain and in the upper cervical spinal cord. Complete surgical resection can be curative and there is no need for further adjuvant therapy. Our presented case is the 12th cervical spinal melanocytoma among spinal melanocytomas in a literature review.

KEY WORDS: Cervical spine, Immunohistochemical study, Melanocytoma, Spinal tumor

Primary pigmented tumors of the central nervous system that arise from leptomeningeal melanocytes are an uncommon and can be diffuse or circumscribed. Melanocytomas are of neural crest origin during early embryonic development and occur in normal leptomeninges. They are usually malignant. This unusual entity includes pigmented meningioma or melanotic meningioma, meningeal melanocytoma, diffuse melanocytosis, malignant meningioma, melanoblastosis and melanotic schwannoma (11,12,13). The terminology of meningeal melanocytoma was first described by Limas and Tio in 1972 (3,6,11). Melanocytomas account for 0.06-0.1% of all brain tumors. They occur in all ages, but are most frequent in the fifth decade with a slight female predominance (2). They are most frequently found in sulci around the brain and in the upper cervical spinal cord. We present a case of a melanocytoma involving the right C4 nerve root.

CASE REPORT
A 41-year-old woman presented with neck and right upper extremity pain. The Visual Analogue Score was six. There was no neurological deficit and no melanotic pigmentation of the skin, mucous membranes or eyes on physical examination. Cervical MRI showed a 20x21x10 mm right C4 foraminal intradural extramedullary lesion. There was increased signal intensity relative to the spinal cord on T1 WI while the lesion was hypointense on T2 WI (Figure 1A,B,C,D). The lesion showed diffuse homogeneous enhancement after gadolinium administration.

The patient underwent surgery in the prone position and C3-4 hemilaminectomy was performed. Brown-colored dura was seen after hemilaminectomy. The right C4 root dura was opened in an oblique fashion precisely through the midline. The intradural extramedullary dumbbell-shaped red-brown colored tumor was totally removed (Figure 2A,B). Postoperative neurological
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Figure 1: A) T2 WI showed 20x21x10 mm right C4 foraminal intradural extramedullary lesion. B) The lesion showed diffuse homogeneous enhancement on T1WI after gadolinium administration. C) Axial T1 WI showed hypointense lesion at the level of C4. D) Diffuse contrast enhanced lesion at the level of C 4 foramen showed on T 1 WI after gadolinium administration.

Figure 2: Intraoperative photograph showed dark brown ovoid-shaped lesion 1 cm in diameter (A, B).

Figure 3: Postoperative MRI showed no enhanced lesion. A) T2WI sagittal; B) T1WI sagittal contrast administration; C) T2WI axial D) T1 WI axial contrast administration.
examination was otherwise normal. She appeared to be neurologically stable with no residual or recurrent tumor on follow up MRI three months after surgery (Figure 3 A,B,C,D).

**Histopathology**

Histologic examination showed the typical appearance of a pigmented lesion. The tumor was composed of spindle and epithelioid cells with heavy brown-black pigmentation. Melanin decoloration was performed histochemically. The tumor cells showed no pleomorphism, mitosis or necrosis. The tumor cells stained positive with Human Melanoma Black 45 (HMB-45) and focally positive with S 100 protein but negative with epithelial membrane antigen (EMA). The Cluster of Differentiation 68 (CD 68), CD 34 and Ki 67 index was less than 1%. (Figure 4 A,B,C).

**DISCUSSION**

The differential diagnosis of primary pigmented lesions of the leptomeninges includes pigmented meningioma or melanotic meningioma, meningeal melanocytoma, diffuse melanocytosis, malignant meningioma, melanoblastosis and melanotic schwannoma. They are rare lesions arising from normally occurring leptomeningeal melanocytes. Melanocytomas are of neural crest origin. Current embryological evidence suggests a common origin of melanocytes originating from the neural crest elements normally found in the basal layer of the epidermis and leptomeninges. Consequently, the areas commonly involved are the base of the brain and upper cervical spine. These tumors are divided into three main types as diffuse melanosis, meningeal melanocytoma, and primary malignant melanoma (1,2,6,12). Diffuse melanosis occurs more frequently in children and is associated with congenital neurocutaneous syndromes and Ota’s nevus (6). Melanocytomas are of benign nature and cured after total excision. Meningeal melanocytomas and primary malignant melanomas are similar in their origin from leptomeningeal melanocytes and range from a lesion that is benign in appearance and behavior to one that is malignant. Primary malignant melanomas are aggressive tumors and the prognosis is related to tumor localization and respectability (8,11,13).

The preoperative diagnosis of meningeal melanocytoma is often difficult as the histopathological and radiological features of the various meningeal lesions are not definitive (1,4,11,14). Meningeal melanocytomas are radiologically similar to meningiomas and neurinomas. They have a characteristic MRI appearance due to the paramagnetic properties of melanin except for malignant melanomas. The MRI appearance may often suggest a diagnosis of a melanocytic leptomeningeal process with the characteristic shortening of T1 and T2 relaxation times. The MRI also shows homogeneous enhancement on post-contrast images (5,9,10,14). The ultrastructural and IHC characteristics of these varied lesions are also unique (1,2,4).

Most melanocytomas arise in the extramedullary, intradural compartment at the upper cervical and thoracic spinal levels. They can be dura-based or associated with nerve roots or the spinal foramina and present with symptoms related to compression of the spinal cord or nerve root (1,2,6,11,13). Our presented case is the 12th cervical spinal melanocytoma among spinal melanocytomas in a literature review (Table 1).

We evaluated the patient’s neurological and neuroradiological findings to exclude other lesions considered in the differential diagnosis such as

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**Figure 4:** A) HMB-45 100x; B) Hematoxylin and Eosin (HE) 100x; C) Melanin Decoloration
Melanocytic lesions of the nervous system are to be distinguished from metastatic malignant melanoma and from histologically different nervous system tumors undergoing melanization, such as schwannoma, medullablastoma, paraganglioma and various gliomas. Malignant leptomeningeal melanoma is histologically similar to melanomas. Anaplastic spindled or epithelioid cells, arranged in loose nests, fascicles or sheets, display variable cytoplasmic melanin. Melanocytoma is characterized by a positive immunohistochemical reaction to antimelanoma antibody (HMB-45), S-100 protein, and vimentin antibodies, and by a negative reaction to epithelial membrane antigen (EMA). Ki 67 protein is a cellular marker for proliferation. Ki 67 index was less than 1% in our case that means its benign (1,2,4,12).

Schwannomas may occur at the lumbar spinal levels. Neural foraminal widening, eroded peduncles and an intradural extramedullary localization are seen radiologically. It is similar to melanocytoma after contrast enhancement. Radicular symptoms are more frequent than in melanocytomas. Meningiomas are the second most frequent spinal tumor. Radiological features and clinical findings are similar to melanocytomas. Ependymomas, epidermoid and dermoid tumors and metastases are well-discussed tumors of conus and filum localization (1,6,7,11,12,13).

The gross appearance of melanocytoma as seen during surgery or at autopsy is that of a well-encapsulated, nodular, dark brown or black lesion that is firmly attached to the underlying leptomeninges as likely in our case. A meningioma may mimic this gross appearance if large amounts of hemosiderin are present within the lesion from previous episodes of hemorrhage (6,11,12,13).

In conclusion, patients with meningeal melanocytoma do much better than with primary or metastatic melanoma of the leptomeninges. Complete surgical resection can be curative and there is no need for further adjuvant therapy. Ocular and physical examinations are recommended to search for malignant formations of phacomatosis. Malignant melanoma, schwannoma, meningioma, ependymoma, epidermoid and dermoid tumors need to be considered in the differential diagnosis. The differential diagnosis will allow the surgeon to make decisions regarding aggressive surgical management and inform the pathologist regarding whether the diagnosis is benign or malignant.

### Table 1: Literature review of cervical spinal melanocytomas

<table>
<thead>
<tr>
<th>Serial-reference no</th>
<th>Age-sex</th>
<th>localization</th>
<th>operation</th>
<th>radiotherapy</th>
<th>recurrence</th>
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<tr>
<td>Lindbom, 1912(16)</td>
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<td>C2</td>
<td>Autopsy</td>
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<td>(-)</td>
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<td>Limas and Tio, 1972(15)</td>
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<td>C1</td>
<td>Autopsy</td>
<td>(-)</td>
<td>(-)</td>
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<td>Graham et al, 1876(9)</td>
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<td>C7</td>
<td>Subtotal</td>
<td>(-)</td>
<td>(-)</td>
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<td>Lach et al, 1988(14)</td>
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<td>C3-5</td>
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<td>(-)</td>
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<td>C1-5</td>
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<td>Tatagiba et al, 1992(26)</td>
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<td>C8</td>
<td>Total</td>
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<td>?</td>
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<tr>
<td>Sankhia et al, 1996(23)</td>
<td>?</td>
<td>C5</td>
<td>Total</td>
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<td>Rasmussen et al, 2000(21)</td>
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REFERENCES


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