Intracranial Meningeal Melanocytoma
—Case Report—

Osamu HAMASAKI, Toshinori NAKAHARA, Shigeyuki SAKAMOTO, Munenori KUTSUNA, and Katsuaki SAKODA

Department of Neurosurgery, Mazda Hospital, Hiroshima

Abstract

A 59-year-old man presented with a rare intracranial meningeal melanocytoma in the left cerebellopontine angle. The patient underwent partial surgical excision and radiosurgery for successful control of the tumor. Meningeal melanocytoma is an essentially benign melanotic tumor, derived from the melanocytes of the leptomeninges, and may occur anywhere in the cranial and spinal meninges. Preoperative differential diagnosis of intracranial meningeal melanocytoma from malignant melanoma is difficult based on magnetic resonance imaging. Ultrastructural findings are essential to establish the diagnosis. The prognosis of this tumor is not always favorable with occasional local recurrence. Total resection is the best treatment, but gamma knife radiosurgery is effective for the residual tumor following partial resection.

Key words: meningeal melanocytoma, cerebellopontine angle, magnetic resonance imaging, gamma knife radiosurgery

Introduction

Primary intracranial melanotic neoplasms are rare, and usually occur as malignant melanoma. Meningeal melanocytoma is a benign type of primary melanotic neoplasm and is less common than the malignant types. This benign tumor has also been named pigmented meningioma or melanotic meningioma because the light microscopy characteristics are almost identical to those of meningioma. Electron microscopy demonstration of the melanocytic ultrastructure of these tumors suggested the term “meningeal melanocytoma” based on the melanocytic origin. Melanocytes are derived from the neural crest during early embryonic development and occur in normal leptomeninges. Scattered melanocytes are most frequently found in the recesses of the sulci around the base of the brain and in the upper cervical spinal cord. Meningeal melanocytomas probably arise from these cells, and mainly occur in the posterior fossa and the spinal canal. We report a case of intracranial meningeal melanocytoma arising from the cerebellopontine angle and its imaging characteristics and clinical features.
portion of the tumor in the cerebellopontine angle was removed, but the portion in the jugular foramen was left unresected to preserve the lower cranial nerve. After treatment of postoperative cerebrospinal fluid leakage, the patient recovered without further complications.

Histological examination demonstrated cellular neoplasm consisting of densely packed cells with cytoplasm heavily pigmented by brown granules (Fig. 3 left). After bleaching with potassium permanganate, the neoplastic cells were found to have oval or elongated nuclei with a few prominent nuclei accompanied by moderate nuclear pleomorphism (Fig. 3 right). There were no mitotic figures, necrosis, or central nervous system (CNS) invasion. Immunoperoxidase staining for S-100 protein and HMB-45 was positive, whereas epithelial membrane antigen was not detected. Ultrastructural study showed the tumor cells were devoid of basal membrane structures and neither desmosomes nor

Fig. 1 Computed tomography scan without contrast enhancement showing a hyperdense mass in the left cerebellopontine angle.

Fig. 2 $T_1$-weighted magnetic resonance (MR) images showing a hyperintense extra-axial mass (left) but with no clear contrast enhancement (center). $T_2$-weighted MR image demonstrate a markedly hypointense area within the lesion (right).

Fig. 3 Photomicrographs of the tumor demonstrating the cellular neoplasm consisting of densely packed cells with cytoplasm heavily pigmented by brown granules (left), and with melanin bleaching showing tumor cells with moderate nuclear pleomorphism and no mitotic figures (right). HE stain, $\times 400$. 

Meningeal Melanocytoma
Fig. 4 Electron micrograph of the tumor showing many melanosomes in varying stages of maturation including premelanosomes within the cytoplasm. Bar = 2 μm.

Fig. 5 T₁-weighted magnetic resonance images with contrast enhancement 3 months after surgery demonstrating residual tumor (left), and 24 months after radiosurgery showing no regrowth of the tumor (right).

tonofilaments were seen (Fig. 4). Occasional clusters of melanosomes in various stages were observed corresponding to the compound melanosomes. The diagnosis was meningeal melanocytoma.

Three months after surgery, he appeared to be neurologically stable, but MR imaging with contrast enhancement suggested residual tumor (Fig. 5 left). The patient underwent radiosurgery with a ⁶⁰Co gamma unit. Stereotactic MR imaging-based multipole isocenter computer dose planning was performed with a complex source plugging pattern to limit the radiation dose to the brain stem to within 15 Gy. Irradiation of 13 Gy was delivered at the tumor margin. Following irradiation, no symptomatic radiation injury was observed. Follow-up MR imaging with contrast enhancement conducted 24 months after surgery showed no evidence of regrowth or metastasis (Fig. 5 right).

Discussion

The imaging and histological characteristics of this essentially benign tumor have been defined by recent cases. Ultrastructural examination is essential to establish the diagnosis of intracranial meningeal melanocytoma, as found in 14 cases of intracranial meningeal melanocytoma described previously and here (Table 1).

The patients were aged from 27 to 69 years (mean 53 years) with equal sex distribution. Eight cases were located in the posterior cranial fossa, especially ventral to the brain stem or posterior adjacent to the foramen magnum, and two cases in Meckel’s cave. Most patients showed signs and symptoms of intracranial expanding mass lesions with durations ranging from 1 month to 14 years. Histological findings showed the tumors were not aggressive with essentially slow progression in all cases.

Intracranial meningeal melanocytoma is a rare clinical entity and the differential diagnosis includes pigmented meningioma, melanotic schwannoma, and primary or secondary malignant melanoma. Intracerebral malignant melanoma has a similar MR signal pattern, but has a greater effect on MR imaging appearance than melanin, appearing as heterogeneous signal intensity. Most malignant melanomas include some hemorrhage, so the appearance varies depending on amount of hemorrhage. Non-hemorrhagic amelanotic melanoma appears as isointense or mildly hypointense on T₁-weighted MR imaging and isointense or mildly hypointense on T₂-weighted imaging. In addition, the MR imaging appearance of melanoma is not uniform but depends on the degree of melanization. Meningeal melanocytomas can be confused with meningiomas because their biological behavior is quite similar. Like meningiomas, melanocytomas tend to be solitary, are often attached to the underlying dura, and may be locally invasive. Intracranial meningeal melanocytomas tend to occur in the posterior fossa and cerebellopontine angle, so may also be difficult to differentiate from schwannomas.

The CT appearance of intracranial meningeal melanocytoma is characterized by well-circumscribed, iso- to slightly hyperdense, extra-axial tumors with homogeneous enhancement similar to meningiomas. Tumor calcification and hyperostosis of adjacent bone are rare in intracranial meningeal melanocytomas, but absence of these signs obviously does not rule out meningio-
### Table 1  Summary of reported cases of intracranial meningeal melanocytoma with ultrastructural diagnosis

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Author (Year)</th>
<th>Sex</th>
<th>Age</th>
<th>Location of tumor</th>
<th>Duration of symptoms</th>
<th>CT finding</th>
<th>MR imaging intensity</th>
<th>Operation</th>
<th>Radiation</th>
<th>Recurrence</th>
<th>Follow-up survival (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Botticelli et al. (1983)²</td>
<td>F</td>
<td>43</td>
<td>Meckel’s cave</td>
<td>10 yrs</td>
<td>hyperdense</td>
<td>n.d. n.d.</td>
<td>partial</td>
<td>yes, 55 Gy</td>
<td>yes</td>
<td>48</td>
</tr>
<tr>
<td>2</td>
<td>Jellinger et al. (1988)¹²</td>
<td>F</td>
<td>27</td>
<td>occipital</td>
<td>1 yr 10 mos</td>
<td>hyperdense with enhancement</td>
<td>n.d. n.d.</td>
<td>total</td>
<td>no</td>
<td>yes</td>
<td>60</td>
</tr>
<tr>
<td>3</td>
<td>Naul et al. (1991)¹⁰</td>
<td>F</td>
<td>68</td>
<td>foramen magnum</td>
<td>6 wks</td>
<td>hyperdense with enhancement</td>
<td>iso iso</td>
<td>subtotal</td>
<td>no</td>
<td>no</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>Litofsky et al. (1992)¹⁰</td>
<td>F</td>
<td>32</td>
<td>anterior to pons and medulla oblongata</td>
<td>2 yrs</td>
<td>isodense with enhancement</td>
<td>iso high</td>
<td>total</td>
<td>no</td>
<td>yes</td>
<td>60</td>
</tr>
<tr>
<td>5</td>
<td>Uematsu et al. (1992)²¹</td>
<td>M</td>
<td>62</td>
<td>foramen magnum</td>
<td>1 yr</td>
<td>iso–hyperdense with enhancement</td>
<td>high low</td>
<td>total</td>
<td>no</td>
<td>no</td>
<td>18</td>
</tr>
<tr>
<td>6</td>
<td>Prabhu et al. (1993)²³</td>
<td>F</td>
<td>67</td>
<td>CP angle</td>
<td>14 yrs</td>
<td>hyperdense with enhancement</td>
<td>n.d. n.d.</td>
<td>total</td>
<td>no</td>
<td>no</td>
<td>4</td>
</tr>
<tr>
<td>7</td>
<td>Maiuri et al. (1995)¹⁰</td>
<td>M</td>
<td>69</td>
<td>sphenoid wing</td>
<td>2 mos</td>
<td>hyperdense with enhancement</td>
<td>n.d. n.d.</td>
<td>subtotal</td>
<td>no</td>
<td>no</td>
<td>12</td>
</tr>
<tr>
<td>8</td>
<td>Takenaka et al. (1996)²⁸</td>
<td>F</td>
<td>64</td>
<td>medulla oblongata</td>
<td>3 mos</td>
<td>hyperdense with enhancement</td>
<td>high low</td>
<td>biopsy</td>
<td>no</td>
<td>no</td>
<td>12, died</td>
</tr>
<tr>
<td>9</td>
<td>Hirose et al. (1997)¹¹</td>
<td>M</td>
<td>66</td>
<td>foramen magnum</td>
<td>1 yr</td>
<td>n.d.</td>
<td>iso low</td>
<td>total</td>
<td>no</td>
<td>no</td>
<td>12, died</td>
</tr>
<tr>
<td>10</td>
<td>Kawaguchi et al. (1998)¹³</td>
<td>M</td>
<td>45</td>
<td>frontal</td>
<td>several mos</td>
<td>hyperdense</td>
<td>slightly high</td>
<td>low total</td>
<td>no</td>
<td>yes</td>
<td>36</td>
</tr>
<tr>
<td>11</td>
<td>Leonardi et al. (1998)¹⁰</td>
<td>M</td>
<td>67</td>
<td>Meckel’s cave</td>
<td>1 yr</td>
<td>hyperdense with enhancement</td>
<td>high iso–high</td>
<td>total</td>
<td>no</td>
<td>no</td>
<td>32</td>
</tr>
<tr>
<td>12</td>
<td>Clarke et al. (1998)⁵</td>
<td>F</td>
<td>30</td>
<td>retroclival to CP angle and foramen magnum</td>
<td>3 mos</td>
<td>hyperdense with enhancement</td>
<td>high n.d.</td>
<td>subtotal</td>
<td>yes, 54 Gy</td>
<td>yes</td>
<td>12, died</td>
</tr>
<tr>
<td>13</td>
<td>Czirjak et al. (2000)⁸</td>
<td>M</td>
<td>48</td>
<td>pineal region</td>
<td>2 yrs</td>
<td>hyperdense with enhancement</td>
<td>high low</td>
<td>total</td>
<td>no</td>
<td>no</td>
<td>3</td>
</tr>
<tr>
<td>14</td>
<td>Present case</td>
<td>M</td>
<td>59</td>
<td>CP angle</td>
<td>1 mo</td>
<td>hyperdense</td>
<td>high low</td>
<td>partial yes, GKR</td>
<td>no</td>
<td>24</td>
<td></td>
</tr>
</tbody>
</table>

Small schwannomas usually show uniform contrast enhancement, whereas larger schwannomas may be heterogeneous.\(^7\)

Nine cases of intracranial meningeal melanocytoma including ours have been reported with detailed MR imaging descriptions (Table 1). Five of the nine cases appeared as hyperintense on T\(_1\)-weighted images and hypointense on T\(_2\)-weighted images. The latter is attributable to content of the neoplasm or the paramagnetic effect of melanin. The reason for the different T\(_1\) signal from that usually seen in melanin-containing tumors cannot be explained. Histological examination showed the tumor cells contained scattered areas of cytoplasmic pigment deposition. The influence of hemorrhage was evident, but suggested that the MR imaging appearance of intracranial meningeal melanocytoma was not uniform but depends on the degree of melanization. Therefore, MR imaging of intracranial meningeal melanocytoma is not a very reliable or specific technique for the definition of this rare tumor.

Intracranial meningeal melanocytoma is usually characterized by a benign clinical course. The follow-up period of the 14 cases (Table 1) ranged from 3 to 60 months (mean 24.5 months) and recurrence was observed in five cases (3 cases after total resection). A study of the clinicopathological features of 33 cases of primary melanocytic neoplasm of the CNS found intermediate-grade lesions between melanocytoma and melanoma with some features of aggressive behavior such as relative hypercellularity and microscopic CNS invasion.\(^3\) The intermediate-grade lesions had a poor prognosis, so therapeutic options should be considered. Previous cases of intracranial meningeal melanocytoma indicate that the extent of surgical resection is the most important determinant of the outcome.\(^18,20,28,31,32\) Total resection of intracranial meningeal melanocytoma without complications is difficult because of the location and tight adhesion. Postoperative conventional radiotherapy cannot control recurrence.\(^2,5\) The prognosis of this tumor is not always good with occasional local recurrence, and whether postoperative radiotherapy is warranted remains controversial.\(^2,5,21,32\) Recurrence of meningeal melanocytoma of the CNS is significantly reduced by complete tumor resection at follow up between 1 and 4 years and by incomplete resection combined with radiotherapy at 2 years.\(^24\) Postoperative gamma knife radiosurgery after partial resection of intracranial meningeal melanocytoma can control tumor regrowth.\(^15\) Total resection is best, but gamma knife radiosurgery is effective for residual tumor after partial resection. More cases with a longer follow-up period will help to define the adequate treatment protocol for this rare tumor.

References

Meningeal Melanocytoma


25) Ray BS, Foot NC: Primary melanotic tumors of the meninges: resemblance to meningiomas. Report of two cases in which operation was performed. Arch Neurol Psychiat 44: 104–177, 1940


Address reprint requests to: O. Hamasaki, M.D., Department of Neurosurgery, Mazda Hospital, 2–15 Aosakiminami, Fuchu-cho, Aki-gun, Hiroshima 735-0017, Japan.
e-mail: ohamasa@orange.ocn.ne.jp.