Primary Intracranial Melanoma
—Case Report—

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Abstract
We present a rare case of primary intracranial melanoma in the right occipital region of a 76-year-old male. Magnetic resonance imaging showed an isointensity mass with shorter T₁ and T₂ relaxation times than those of the opposite hemisphere. A well-defined, dark black tumor was totally removed and histologically diagnosed as malignant melanoma. Eight months postoperatively, however, the tumor recurred and was excised again. He was doing well 1 year after the second operation without additional treatment. In our case, 1) no systemic melanomas were found in close clinical examinations; 2) there was a single nodular tumor attached to the leptomeninges; and 3) a favorable outcome was obtained by surgical treatment alone. These results are consistent with the diagnosis of primary intracranial melanoma.

Key words: melanoma, primary brain tumor, magnetic resonance imaging

Introduction
Malignant melanoma arises either from melanocytes or their precursor cells, melanoblasts. Its incidence is relatively low, 0.38/100,000 in Japanese but 1.7–1.8/100,000 in Caucasians. Malignant melanomas commonly develop in the skin, mucosa, and eye regions. Since melanocytes are considered to originate from neural crest and are found in normal leptomeningeal tissue, it is not surprising that primary melanomas also grow within the central nervous system (CNS). However, primary CNS melanomas have rarely been reported, constituting only 16 of 22,898 cases of primary intracranial tumors in Japan.

Primary CNS melanoma represents two growth patterns, solid nodular mass lesions and diffuse meningeal infiltration. The former type melanomas grow in the spinal cord, cerebral hemispheres, cerebellopontine angle, cisterna magna, or pineal regions. We report a case of primary intracranial melanoma on the occipital cortical surface, which was diagnosed by operative, histological, and other clinical findings. Although a focal recurrence was encountered, better clinical results were obtained by surgical treatment alone. No systemic melanomas have been found so far by repeated meticulous examination. We also discuss the diagnostic value of magnetic resonance (MR) imaging for intracranial melanomas.

Case Report
A 76-year-old male with a 3-month history of progressive headache, visual disturbance, and left side muscle weakness was hospitalized in November, 1986. The neurological examination disclosed dysarthria, left homonymous hemianopsia, mild left hemiparesis, and left hyper-reflexia with extensor planter response. Physical examination detected no skin lesions or organomegalies. Laboratory data were normal. Computed tomographic (CT) scans demonstrated a large, irregularly shaped high-density tumor, with homogeneous contrast enhancement, surrounded by perifocal edema in the right occipital superficial region (Fig. 1A). Retrograde right brachial angiograms showed an avascular mass in the parieto-occipital area. Spin-echo (SE) MR images revealed an isointensity tumor on T₁ as well as T₂-weighted sequences (Fig. 1B). The mean T₁ and T₂ relaxation times measured at 5 regions of interest in the tumor were 650 and 84 msec, which were shorter than those of the opposite occipital gray matter
Through a right occipital craniotomy, a well-defined, dark black tumor attached to the pia-arachnoid was totally removed. Histological examination disclosed round or polygonal tumor cells containing dark brown cytoplasmic granules, which were positive to Fontana-Masson stain, indicating melanin (Fig. 2). The tumor cells had marked nuclear atypism and mitotic figures, and occasionally formed several nests with a honeycomb appearance or a reticular pattern. There were some amelanotic parts in the tumor as well. These findings were compatible with the histological diagnosis of malignant melanoma. Marked mononuclear cell infiltration and macrophages containing melanin were noted. There was no evidence of intratumoral bleeding.

The postoperative course was uneventful. No melanomas were detected by systemic clinical examination, including dermatological and ophthalmological examinations, $^{67}$Ga-radioisotope scintigraphy, and upper and lower gastrointestinal tract roentgenography. Postoperative chemotherapies were planned but could not be given because he and his family refused further treatment. He was discharged 1 month after the operation without obvious neurological deficit.

In July, 1987, 8 months postoperatively, follow-up CT and MR studies demonstrated tumor recurrence at the previous operation site (Fig. 3A, B). The radiological characteristics of the recurrent tumor were similar to those of the previous tumor, except for the longer $T_1$ and $T_2$ relaxation times (843 and 95 msec) than those of the opposite hemispheric gray matter (675 and 76 msec). On a $T_2$-weighted MR image with intravenous gadolinium-diethylene-

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Fig. 1 A: Precontrast CT scan (left), showing an irregularly shaped high-density mass with perifocal low density in the right occipital superficial cortical region. Postcontrast CT scan (right), demonstrating the mass to be homogeneously enhanced. B: $T_1$- (left) and $T_2$-weighted (right) SE MR images, showing an isointensity mass (arrowheads). Note the marked high-intensity area surrounding the tumor on the $T_2$-weighted image.

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triaminepenta-acetic acid (Gd-DTPA) injection, the tumor appeared as an enhanced high-intensity lesion (Fig. 3C).

At the second operation, a defined brown tumor on the occipital cortical surface, resembling the previously excised tumor except for the color, was successfully removed. Histological examination revealed more dominant amelanotic cells than in the previous specimen. Repeated systemic examination again indicated no melanomas. He was doing well 1 year after the second operation without evidence of tumor recurrence.

Discussion

Since systemic melanomas frequently metastasize to the CNS, a definitive diagnosis of primary CNS melanoma can be made if no systemic focus considered to be primary is found at autopsy. Terao et al., on the other hand, reported clinical differences between metastatic and primary melanomas in the CNS. Metastatic melanomas are characterized by 1) multiple intracerebral tumors, 2) a rapid, poor clinical course due to systemic metastases, and 3) development in older patients, while primary CNS melanomas unusually metastasize to systemic organs and develop in relatively younger patients (usually under 50 years old). Our patient was diagnosed as primary intracranial melanoma, because no systemic melanomas were found by repeated clinical examination, and there was a solitary mass located in the superficial cortical region and attached to the leptomeninges. In addition, the clinical course of our patient was better than that of metastatic intracranial melanoma.

The preoperative diagnosis of primary CNS melanoma is difficult, except in cases associated with neurocutaneous melanosis, or when melanin or melanin-containing cells are detected in the cerebrospinal fluid. The CT findings of intracranial melanomas, including high-density mass on precontrast scans, homogeneous enhancement, and marked peritumoral edema, are not specific. On the other hand, recent MR studies have demonstrated characteristic features of melanomas. Damadian et al. reported that the T₁ relaxation times of melanomas are remarkably shorter than those of other malignant tumors, and do not increase even compared with various normal tissues. They attributed these characteristics to the paramagnetic effects of the organic free radicals produced by melanoma cells in association with melanin synthesis. Recently, Gomori et al. supported this hypothesis, indicating that the relaxation times become shorter when the melanin content increases in the tumor tissue. In our case, the tumor showed isointensity to the normal cortex on T₁- and T₂-weighted MR images. These findings are different from those of other primary or metastatic malignant intracranial neoplasms, which usually show low intensity on T₁-weighted images and high intensity on T₂-weighted images. T₁ relaxation time of the initial tumor with

Fig. 3  A, B: Pre- (A, left) and postcontrast (A, right) CT scans and T₁- (B, left) and T₂-weighted (B, right) SE MR images, showing the recurrent tumor at the previous operation site, with similar features to the previous lesion.  C: Gd-DTPA positive enhancement is shown on the T₁-weighted image (left), but not on the T₂-weighted image (right).
many melanin-containing cells was shorter than that of the recurrent tumor with less melanin-containing cells, and was depressed even relative to the opposite gray matter. These observations indicate that MR images provide valuable information for the diagnosis of intracranial melanomas, either primary or metastatic.

Primary intracranial melanoma should be distinguished from other pigmented CNS tumors, particularly from meningeal melanocytoma which was introduced by Limas and Tio. The electron microscopic study by Winston et al. demonstrated that meningeal melanocytoma cells differ from meningothelial cells, suggesting that this tumor arises from the leptomeningeal melanocytes. Although meningeal melanocytoma seems to be a benign, surgically curable neoplasm, the histogenetic relationship between meningeal melanocytoma and primary CNS melanoma, both of which appear to have the same origin, i.e. leptomeningeal melanocytes, remains to be clarified. In our case, the tumor cells were similar to systemic melanoma cells and histological malignancy was apparent, indicating malignant melanoma.

Since melanomas do not appear to be radiosensitive, chemotherapy might be the most important treatment for melanomas. Combined chemotherapy with dimethyl-triazeno-imidazole carboxamide, vincristine, and ACNU (amino chloroethyl nitrosourea) seems to be the most effective currently for systemic malignant melanomas. In patients with a cutaneous melanoma detected in early stage, better outcomes have been obtained by such treatment following surgery. However, in cases harboring distant metastases, particularly involving CNS, the prognoses are usually poor. In contrast to these systemic melanoma cases, several patients with a primary CNS melanoma have achieved better outcomes by surgical intervention with or without additional treatment.

In our patient, the favorable outcome resulted from surgical treatment without additional chemotherapy, although a focal recurrence occurred. The biological behavior of tumor cells of primary CNS melanomas may differ from those of systemic melanomas.

References

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