CASE REPORT

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Primary Occipital Malignant Melanoma

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Primary intracranial melanoma is uncommon. These tumors most commonly occur at the temporal lobe, cerebellum and cerebellopontine angle. We report a case of intracranial malignant melanoma of the occipital lobe in a 60-year-old man who presented with headache and visual disturbance. The mass showed hyperintensity on T1-weighted images and hypointensity on T2-weighted magnetic resonance images. He underwent gross total removal of tumor and received radiotherapy. Follow-up imaging studies showed neither recurrence nor any signs of residual disease for 4 months.

KEY WORDS : Primary intracranial melanoma · Occipital lobe · Hyperintensity on T1WI · Hypointensity on T2WI.

Introduction

Primary intracranial melanomas are rare tumors, generally derived from the melanocytic elements, which normally exist in the leptomeninges[15].

Primary central nervous system(CNS) melanomas are reported to be found in the supratentorial area, in the posterior fossa, or in the spinal cord. Intracranial melanomas most commonly occur at the temporal lobe (20%), cerebellum (25%), or cerebellopontine angle (15%). Occipital lobe lesions are only 2% of all[19].

Radiological patterns of intracranial melanomas can mimic the presence of meningiomas. Long-term disease-free periods are often reported after gross total removal of such melanoma, despite their malignant behavior[2].

To our best knowledge, this case is the first primary occipital malignant melanoma originated from the tentorium. Its imaging finding and management are discussed.

Case Report

A 50-year-old man was admitted with a 1-month history of headache and visual disturbance. He had no specific past medical history. Upon neurological examination, the patient was found to have a right-sided homonymous hemianopsia. No nevi or swelling of the lymph nodes was observed upon physical examination.

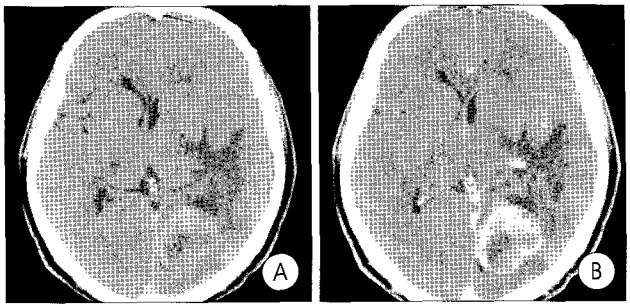

Fig. 1. Computerized tomography scan demonstrates a lobulated hyperdense mass in the left occipital area (A) with central necrosis, inhomogenous contrast enhancement and peritumoral edema (B).

Computerized tomography(CT) scan demonstrated a lobulated, inhomogenously enhancing hyperdense mass in the left occipital lobe with compression of the left occipital horn and marked peritumoral edema (Fig. 1A, B). On magnetic resonance images(MRI), the mass showed a mixture of iso- and hyperintense areas on T1-weighted images (Fig. 2A) and inhomogenous hypointensity on T2-weighted images (Fig. 2B). After administration of the contrast material, intense enhancement of the mass was observed (Fig. 2C). Sagittal and coronal views revealed that the tumor appeared to adhere to the tentorium and the falx. (Fig. 2D, E).

An occipital parasagittal craniotomy was performed. The dura was intact and no areas of pigmentation were evident

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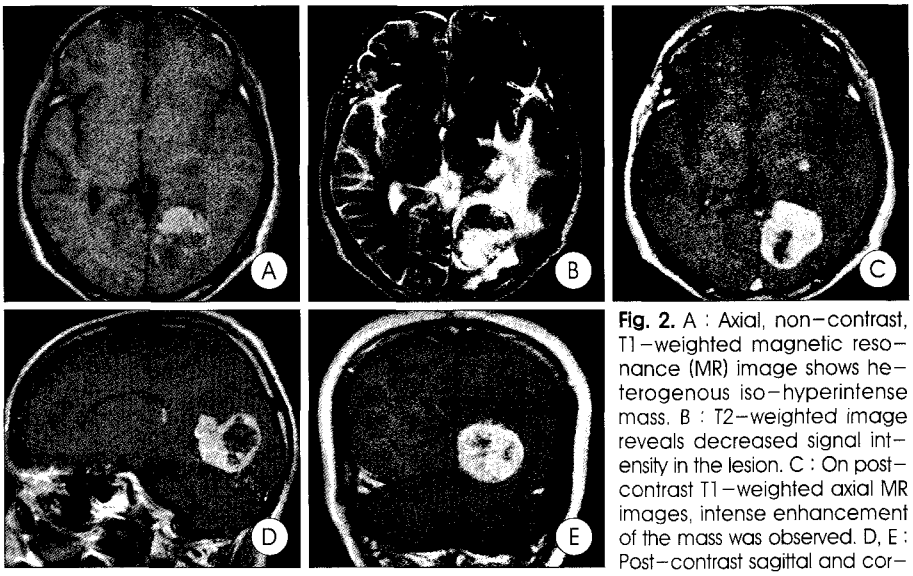

Fig. 2. A : Axial, non-contrast, T1-weighted magnetic resonance (MR) image shows heterogenous iso-hyperintense mass. B : T2-weighted image reveals decreased signal intensity in the lesion. C : On post-contrast T1-weighted axial MR images, intense enhancement of the mass was observed. D, E : Post-contrast sagittal and coronal view shows that the tumor was based on the floor of the tentorium.

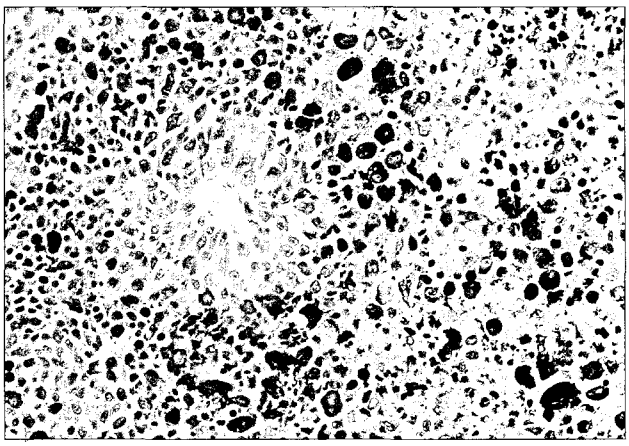

Fig. 3. Histopathologic section demonstrates either polygonal or ovoid cells with abundant melanin pigments. No evidence of hemorrhage or necrosis is observed (hematoxylin-eosin, original magnification X 200).

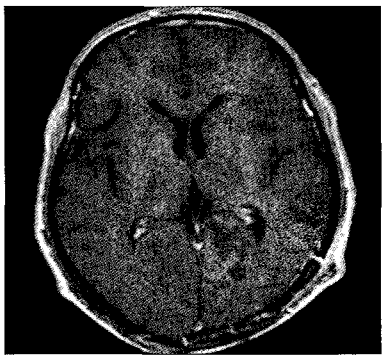

Fig. 4. Contrast-enhanced T1-weighted image 4 months after surgery, shows no evidence of tumor recurrence.

on the inner surface of the dura matter. After retraction of occipital lobe, a black tumor was observed. The tumor, 3.9×3.2×3.4 cm, was soft, rich in small blood vessels, and attached to the tentorium but not falx. Although the tumor was adherent to the arachnoid membrane and tended to bleed easily, it was successfully separated from the brain parenchyma and was gross-totally removed.

Histologic examination showed that the tumor was composed of large, atypical, polygonal or ovoid cells with large, ovoid nuclei. The tumor cells had abundant pigments that stained positively for melanin with hematoxylin-eosin stain (Fig. 3). Immunohistochemistry showed a positivity for HMB45 (anti-melanosomal antibody) and S-100 (anti S-100 protein antibody), but negativity for epithelial membrane antigen(EMA) or for glial fibrillary acid protein(GFAP). MIB-1 staining showed high proliferative index (30%). These findings are diagnostic for malignant melanoma.

Postoperative examinations including dermatological physical examination, ophthalmologic fundoscopic examination, endoscopy of the gastrointestinal tract, and positron emission tomography(PET) CT revealed no evidence of melanoma in other parts of the body. The patient had a normal postoperative course without any complications. The patient subsequently underwent radiotherapy of 5940cGy in 33 fractions (daily 180cGy). Four months after surgery, contrast-enhanced MRI demonstrated no recurrence or sign of residual disease (Fig. 4). He was healthy and independent, though he had residual right-sided hemianopsia.

Discussion

Although the majority of central nervous system(CNS) melanomas are metastatic in origin, primary melanomas occasionally arise from pigmented cells which are normally present in the leptomeninges[17]. Primary intracranial melanomas are rare and occur mainly in younger adults. The peak incidence of these tumors is in the fourth and fifth decades with predominance in males[9]. Metastatic melanomas are more commonly multifocal, located at the junction of gray and white matter, and are rarely accompanied by leptomeningeal dissemination[21]. In contrast, primary melanomas are usually present as either an extra-axial or superficially located mass with frequent leptomeningeal involvement. Primary intracranial melanomas are classified into two types : diffuse tumors of the leptomeninges and discrete tumors. Discrete melanoma with leptomeningeal involvement is more frequently reported in the literature. Patients with decrete melanoma without leptomeningeal involvement are reported to have longer survival[4,22].

The preoperative diagnosis of primary melanomas, parti-

cularly malignant melanomas, of the brain is possible in only 10% of cases and may be difficult due to the lack of specific clinical and radiologic findings[3,8,20]. Typical CT findings of primary discrete intracranial melanomas include a superficially located, well enhancing hyperdense mass. The increased attenuation on the precontrast CT scan is thought to be caused by the presence of melanin. A well enhancing hyperdense mass at a superficial location may often mimic meningioma. Although variable, MRI shows the tumor to be hyperintense on T1-weighted images and hypointense on T2-weighted images. Some cases of previously reported tumors have had low signal intensity on T1-weighted images and iso-, or high signal intensity on T2-weighted images[1,6,10,20-22]. These differences in MR signal intensities are related to the degree of the paramagnetic effects of stable free radicals in melanin and/or hemorrhagic products[12]. The differential diagnoses include meningeal melanocytoma, melanotic meningioma, plasma cell granuloma, melanocytic schwannoma, and other melanin-containing glial tumors with exophytic growth.

Light microscopic examination usually allows for definite diagnosis of melanotic tumors, but there can be some overlap among the various tumor types. Immunohistochemical analysis sometimes be indispensable for the diagnosis of malignant melanoma. In our case, histopathological examination revealed large amounts of melanin pigment and high proliferative index (30%) but no evidence of hemorrhage or necrosis.

The poorest prognostic factor in intracranial primary solitary melanoma is leptomeningeal dissemination. A leptomeningeal enhancement in the melanoma suggests a malignant tumor rather than a benign lesion. Although primary CNS melanomas are aggressive with higher rates of local recurrence and mortality, the prognosis of patients with surgically treated primary solitary melanomas varies considerably, likely reflecting numerous factors such as tumor site, extent of resection, and response to adjuvant therapy. In contrast, the prognosis of the patients with metastatic melanoma to the CNS remains dismal, with a reported life expectancy of less than one year in most studies[4,14].

To prevent sudden spread after surgery, it is important to prevent dissemination by not touching the tumor directly and removing additional surrounding tissue when resecting tumor with leptomeningeal enhancement. Radiotherapy, chemotherapy, immunotherapy, and other adjuvant therapies may be judged necessary to reduce the chances of post-surgical dissemination, but this has not been definitely established. Malignant melanoma was once considered to be one of the most radio-resistant tumors, but radiotherapy, using fractionized irradiation in high doses, has improved response rates[11,16,18]. In chemotherapy, the imidazole carboxamide-derivative, dimethyl triazeno imidazole carboxamide(DTIC), is the most common agent in use and is being researched as a single agent for melanoma[13]. Chiba et al.[7] reported the complete disappearance of metastatic brain lesions by chemo-immunotherapy using DTIC, 1-(4-amino-2-methyl-5-pyrimidinyl) methyl-3-(2-chloroethyl)-3-nitrosourea hydrochloride(ACNU), and vincristine with OK 432(picibanil). In addition, viral oncolysate, an immunotherapeutic agent against neoplasms, is reportedly effective against malignant melanoma[5].

Conclusion

Although primary solitary intracranial melanomas are rare, this tumor type should be suspected if a superficially located mass displays hyperintensity on T1WI, hypointensity on T2WI, and intense enhancement after contrast administration, particularly in conjunction with leptomeningeal enhancement.

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