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Case Report

Primary intracranial melanoma

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ABSTRACT

Primary intracranial malignant melanomas are rarely reported tumors of the central nervous system that are highly malignant with a poor prognosis, accounting for 0.1% of intracranial neoplasms. Given the location of the tumor, preoperative diagnosis by imaging study is difficult. Typically, mainstream therapy involves total excision of the tumor followed by radiation therapy. We present a case of primary intracranial melanoma arising from convexity of the left frontal lobe in a 49-year-old male who presented with headache and progressive mentality change for one month. After total tumor excision and whole brain radiation therapy, the patient has experienced tumor-free survival extending through 47 months of follow-up. Herein we have reported the patient's MRI findings, pathological examination, treatment and outcome.

1. Introduction

Primary melanoma in the central nervous system (CNS) arises from melanocytes which have developed from precursor cells (melanoblasts) in the neural crest. Their distribution is correlated with the distribution of melanocytes, and routinely involve leptomeninges at the anterior and lateral surfaces of the spinal cord, the brain stem and the base of the brain. The incidence of primary intracranial melanomas is approximately 0.005/100,000, with male predominant.[1–3] Melanocytic lesions range from neurocutaneous melanosis to benign well-differentiated tumor (melanocytoma) to malignant melanoma. Of these different lesions, malignant intracranial melanomas account for approximately 1% of melanoma and 0.1% of all intracranial tumors.[4] Only 26 cases of primary malignant melanoma has been reported in the literature. Overall, these tumors are highly malignant, difficult to diagnose before pathological examination, and have a poor prognosis. Herein we have reported a case of primary intracranial malignant melanoma.

2. Case report

A 49-year-old female with an unremarkable medical history presented to our facility with a headache and suffering from mental decline for 1 month. There was no focal neurological deficit, but some psychological symptoms were noted such as social withdrawal. The patient denied any medical or malignant history, and no remarkable findings were found on neurological examination.

Contrast-enhanced brain MRI revealed a 3.7 × 3.8 × 3.2 cm intra-axial brain tumor at the left frontal subcortical area. This tumor is hyper-intense on T1 image and hypo-intense on T2 image, with heterogeneous, thick rim enhancement and non-enhanced central necrotic part. Significant peri-tumoral edema at the left front of the temporal lobes with mass effect was also noted (Fig. 1). However, no hemorrhage was detected from GRE series. Under the preliminary diagnosis of high grade glioma, frontal-temporal craniotomy was performed to excise the tumor.

During the operation, a large dark grayish tumor could be observed from the superficial parenchyma at the patient's frontal region. This tumor was about 3.5 cm and well-demarcated, without gross invasion of the surrounding tissue. Complete tumor excision was routinely accomplished by dissection along its surface.

Macroscopic findings included several pieces of irregular dark brownish tissue, and further included a solid neoplasm with infiltration of the surrounding tissue under H&E stain. Areas of necrosis and intracytoplasmic brown pigment were noted. Under a high power field, the tumor was composed of polygonal cells with prominent oval nuclei; these nuclei were pleomorphic, including some with mitotic figures. Upon immunohistochemistry examination, the diffused positivity for S100 and HMB-45 suggested these tumor cells were of melanocytic origin. Schwannoma and glioma was excluded by the tumor morphology and its obvious margin. The malignant tumor cells had melanin pigmentation and

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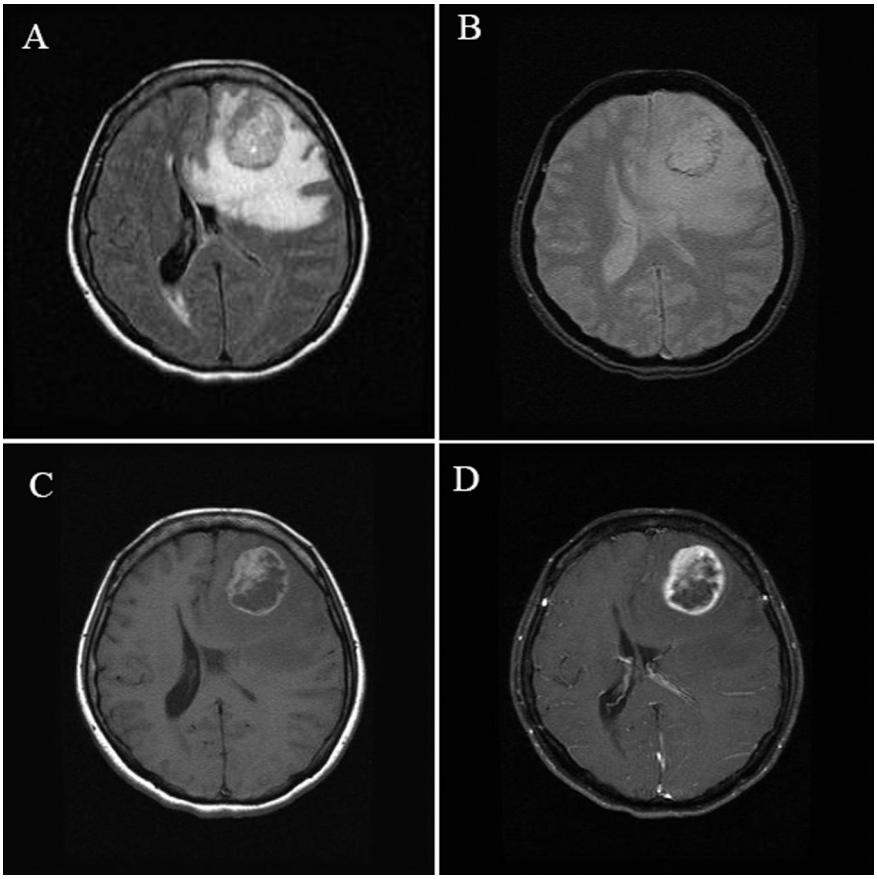

Fig. 1. Demonstrated imaging of tumor on MRI: (A) Tumor was hypointensive on T2-weighted image with significant perifocal edema. (B) GRE T2 weighted image displayed no evidence of hemorrhage. (C) T1 weighted image showed a hyperintense tumor arising from left frontal convexity. (D) Contrast image heterogeneous thick rim enhancement and non-enhanced central necrotic part.

manifested invasion of the surrounding brain tissue (Fig. 2). The tumor cells were positive for S-100 and human melanoma black 45 (HMB-45) (Fig. 3), but negative for cytokeratin. The above findings supported the diagnosis of malignant melanoma.

To facilitate a definite diagnosis, the patient also underwent positron emission tomography with 2-deoxy-2-[fluorine-18] fluoro- D-glucose integrated with computed tomography (^{18}F-FDG PET/CT) to detect any other primary melanoma site, but no FDG active lesion was found. Whole body skin examination showed a bluish papule over the right dorsal hand and a subcutaneous nodule over the anterior neck. Pathological diagnosis was blue nevus and granulomatous dermatitis, respectively.

However, uveal melanoma was found following funduscopic examination.

After excluding the possible of primary site, we believed the tumor was a primary cerebral melanoma. Thereafter, postoperative brain MRI showed residual hematoma with minimal enhancement.

No neurological deficit was found after operation. Following excision of the intracranial tumor, the patient received adjuvant radiotherapy 50 Gy in 20 fractions to the tumor bed smoothly. The patient's brain MRI examined one, two and four years later showed no tumor recurrence (Fig. 4). Since the operation, the patient has had tumor-free survival for more than 4 years.

3. Discussion

It is generally known that primary CNS melanomas are rare and difficult to diagnose prior to biopsy. According to tumor melanin content, four subtypes of malignant intracranial melanoma have been proposed, associated with different MRI image presentations. These subtypes are: 1) melanoma, characterized by hyper-intense T1 image and hypo- or iso-intense on T2 image; 2) non-melanoma, characterized by hypo- or iso-intense on T1 image and hyper- or iso-intense on T2 image; 3) uncertain or mixed, characterized by mixed MRI signal; and 4) hemorrhagic lesions, characterized by different stage of hemorrhage on MRI. The melanoma and hemorrhagic-type lesions account for approximately 70% of malignant intracranial melanomas.[5] In spite of characteristic differences in presentation on MRI imaging, differential diagnosis from meningioma or glioma is frequently difficult.

CNS melanocytic tumors include meningeal melanocytosis, melanomatosis, melanocytoma and melanoma. These lesions are thought to arise from leptomenigeal melanocytes, derived from the neural crest. Meningeal melanocytosis and melanomatosis involve the supra- and infra-tentorial leptomeninges and superficial brain parenchyma. Since this tumor is a solitary lesion and other intra-/extra-cranial lesions are excluded in the following image studies, melanocyotisis and melanomatosis are less likely to be the resulting diagnosis. Meningeal melanocytomas are rare, slow-growing and solitary tumors, sharing similar histological characters with melanomas. They both express melanocytic marker proteins such as S-100, melan A and HMB-45. In our case, the mitotic activity and nuclear pleomorphism of the tumor are characteristics that indicate melanoma rather than melanocytoma.

Due to the rarity of incidence of primary intracranial melanoma, optimal treatments had not yet been established until recently.