

# Usefulness of Neuromelanin Sensitive MRI for En Plaque Meningeal Melanocytoma Involving the Cavernous Sinus: A Case Report

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Intracranial meningeal melanocytoma is a rare tumor. Here, we report a case of pathologically diagnosed en plaque meningeal melanocytoma involving the cavernous sinus along with a review of the pertinent literature. A 35-year-old female presented with progressing left oculomotor nerve palsy and melanosis oculi. Radiological examinations revealed a lesion spreading in an en plaque fashion and involving the left cavernous sinus, which was hyperintense on T<sub>1</sub>-weighted magnetic resonance imaging (MRI), and hypointense on T<sub>2</sub>-weighted MRI. The lesion was partially excised following a histopathological diagnosis of meningeal melanocytoma. For follow-up of the residual lesion, neuromelanin sensitive MRI was introduced, and it provided better contrast between the lesion and surrounding intracranial normal tissue than conventional T<sub>1</sub>-weighted MRI with or without gadolinium. The lesion remained stable without any growth for 3 years post-surgery. Neuromelanin sensitive MRI may be the method of choice for the follow-up of meningeal melanocytoma.

**Keywords:** cavernous sinus, en plaque, meningeal melanocytoma, neuromelanin

## Introduction

Intracranial meningeal melanocytoma is a rare tumor examined by neurosurgeons in clinical practice. It typically originates from melanocytic cells of leptomeninges, and is mainly found in the region of the foramen magnum, posterior fossa, Meckel's cave, or cervical spinal cord.<sup>1)</sup> Meningeal melanocytoma involving the cavernous sinus is even rarer, and to the best of our knowledge, no en plaque type lesion in this location has been previously reported. Here we report a case of an en plaque meningeal melanocytoma involving the cavernous sinus with focus on the usefulness of neuromelanin sensitive MRI, which can demonstrate neuromelanin-related contrast of the lesion for close follow-up of this tumor.

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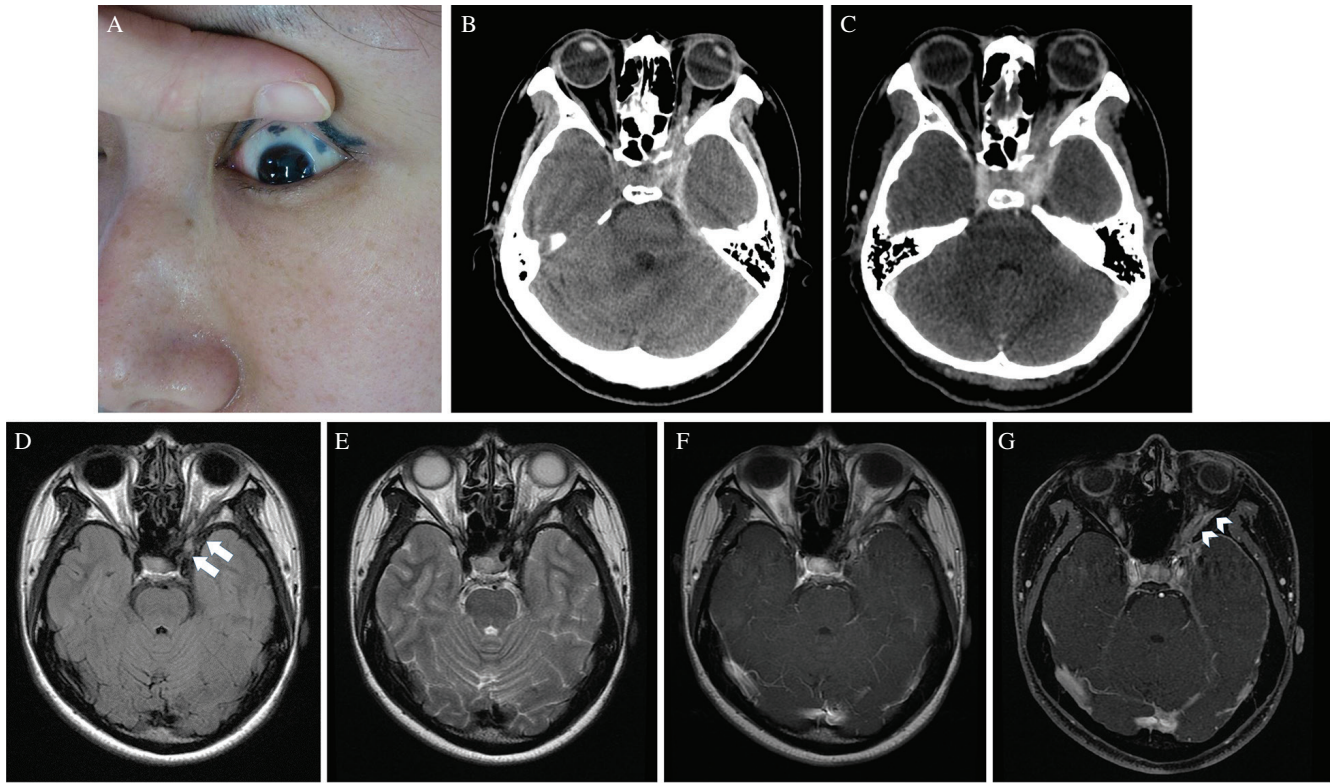
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## Case Report

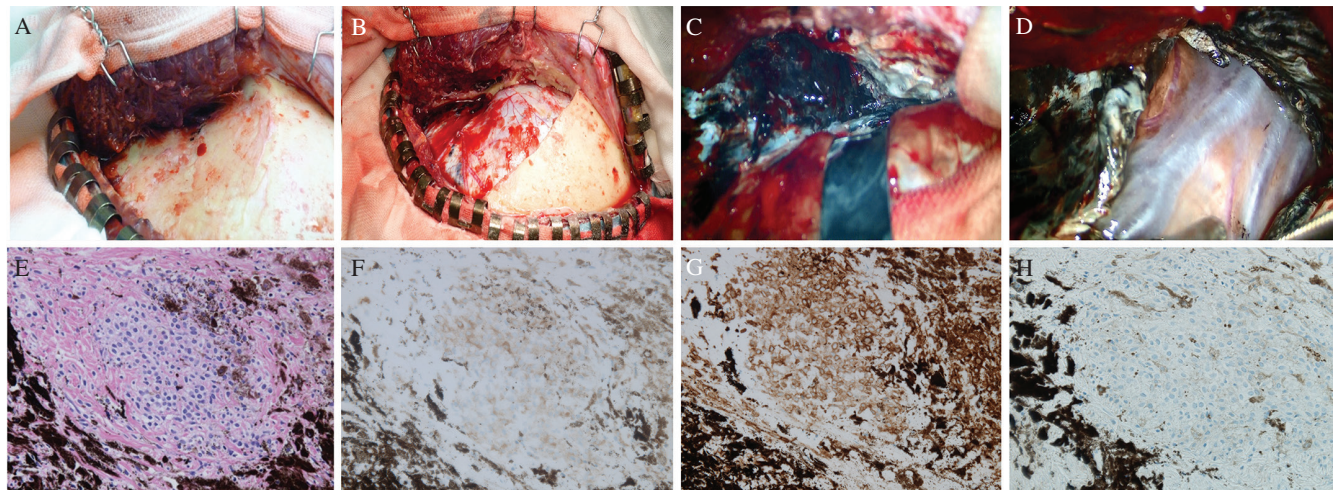
A 35-year-old female with a 6-year history of progressing left exotropia and diplopia was referred to our hospital. Neurological examination revealed left oculomotor nerve palsy including left mydriasis. Her facial sensation was normal. Physical examination revealed patches of black discoloration of the sclera on the left side of the face, which was reported to have been present since her childhood (Fig. 1A). There was no other pigmentation in her face or mouth. A plain computed tomography (CT) scan showed a hyperdense lesion around the left cavernous sinus extending to the orbital apex, with an enlarged superior orbital fissure on the left side. Contrast enhancement effect of the lesion was equivocal (Figs. 1B and 1C). The lesion appeared hyperintense on T<sub>1</sub>-weighted MRI sequence and hypointense on T<sub>2</sub>-weighted MRI sequence. It showed some enhancement effect after gadolinium administration, and the addition of a fat suppression technique clearly demonstrated intra-orbital extension of the lesion (Figs. 1D and 1G). Cerebral angiography was unremarkable. The association of patches of black discoloration of the sclera (nevus of Ota) to the lesion located on the ipsilateral cavernous sinus wall and the aforementioned radiological features suggested the lesion's nature of a melanotic disorder.

To obtain a pathological diagnosis and relieve the patient's symptoms, surgery was performed using the pterional approach. The anterior clinoid process and bones forming the lateral rim of the superior orbital fissure were removed for decompression of the oculomotor nerve. Intraoperative findings included some black staining of a part of the temporal bone, diffuse blackening of the dura with an increasing gradient toward the skull base and scattered pigmentation of the arachnoid membrane (Figs. 2A–2D). To avoid further disturbance of the cranial nerves in the cavernous sinus, the meningeal lesion in the temporal dura close to the superior orbital fissure was partially removed.

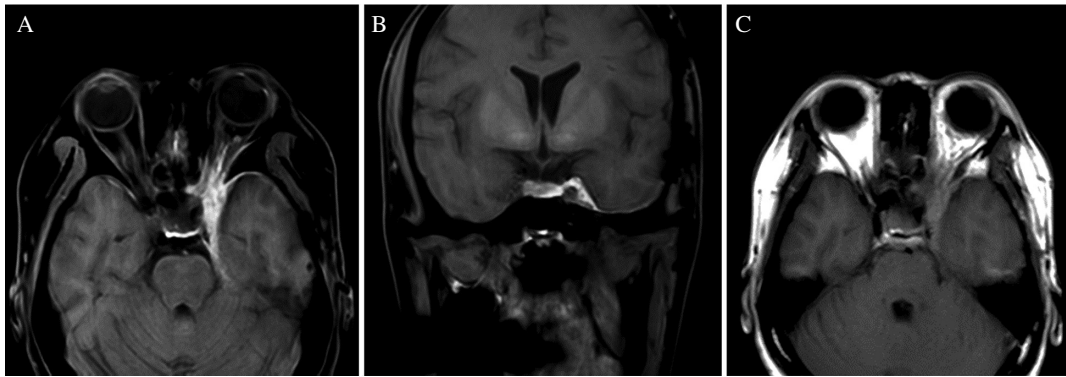
On hematoxylin and eosin staining, the tumor appeared as uniform sheets of polygon- or spindle-shaped cells, with regular nuclei and pigmentation of the cytoplasm. Mitotic activity was not observed and the MIB-1 labeling index was less than 1.0%. There was no necrosis or hemorrhage. Neoplastic cells were immunoreactive for human melanoma black-45, melanoma antigen recognized by T cells-1, c-kit, and vimentin, but not for epithelial membrane antigen, S100, and glia fibrillary acidic protein (Figs. 2E–2H). Histopathological diagnosis was meningeal melanocytoma.



**Fig. 1** A photograph of the patient's left eye showing congenital patches of black discoloration of the sclera (A). Pre-operative computed tomography scan showing hyperdense lesion around the left cavernous sinus, which shows an equivocal contrast enhancement effect (B, C). Pre-operative axial magnetic resonance images show the lesion around the left cavernous sinus as partly hyperintense on T<sub>1</sub>-weighted image (D, arrow) and hypointense on T<sub>2</sub>-weighted image (E). It shows some enhancement effect after gadolinium administration (F), and the fat suppression image clearly demonstrates intra-orbital extension of the lesion (G, arrow head).



**Fig. 2** Intraoperative photographs showing partly stained temporal muscle and bone (A), diffusely stained temporal dura (B, C), and partially infiltrated arachnoid membrane around the Sylvian fissure (D). Histologic features of the tumor showing uniform sheets of polygon- or spindle-shaped cells, with regular nuclei and pigmentation of the cytoplasm. Mitotic activity, necrosis, and hemorrhage are not observed (E). (Hematoxylin and eosin stain,  $\times 400$ , original magnification) The neoplastic cells are immunoreactive for HMB45 (a marker for melanocytic tumors) (F) and MART1 (G), but not for EMA (H). Intraoperative photographs showing partly stained temporal muscle and bone (A), diffusely stained temporal dura (B, C), and partially infiltrated arachnoid membrane around the Sylvian fissure (D). Histologic features of the tumor showing uniform sheets of polygon- or spindle-shaped cells, with regular nuclei and pigmentation of the cytoplasm. Mitotic activity, necrosis, and hemorrhage are not observed (E). (Hematoxylin and eosin stain,  $\times 400$ , original magnification) The neoplastic cells are immunoreactive for HMB45(a marker for melanocytic tumors) (F) and MART1 (G), but not for EMA (H).



**Fig. 3** Neuromelanin sensitive MRI with fat suppression (A: axial, and B: coronal) taken 3 years after surgery demonstrating the extension of the en plaque meningeal melanocytoma involving the left cavernous sinus more clearly than the T<sub>1</sub>-weighted image (C). There is no apparent growth of the lesion. Neuromelanin sensitive MRI with fat suppression (A: axial, and B: coronal) taken 3 years after surgery demonstrating the extension of the en plaque meningeal melanocytoma involving the left cavernous sinus more clearly than the T<sub>1</sub>-weighted image (C). There is no apparent growth of the lesion.

**Table 1** Characteristics of patients with meningeal melanocytoma involving the cavernous sinus

Author (Year)	Age	Gender	Presenting symptom	Growth pattern on MRI	Nevus of Ota	Surgical removal	Radiation	Regrowth and treatment	Outcome
Faro et al. (1996) <sup>7)</sup>	30	F	Headache	Nodular	–	Total	–	–	1 Month alive
Pan et al. (2011) <sup>8)</sup>	36	M	Ptosis with papilledema	Nodular	+ ipsi	Subtotal	+	–	1 Year alive
Doglietto et al. (2012) <sup>9)</sup>	20	M	Abducens nerve palsy	Nodular, multiple	+ ipsi	Partial	+	3 Years hemotherapy	7 Years died of metastasis
This case	35	F	Exotropia, oculomotor nerve palsy	En plaque	+ ipsi	Partial	–	–	3 Years alive

ipsi: ipsilateral to the meningeal melanocytoma.

Her oculomotor nerve palsy remained unchanged post-operatively. For radiological follow-up, neuromelanin sensitive MRI was introduced, which showed much better contrast between the lesion and the surrounding intracranial normal tissue, including the cavernous sinus, than conventional T<sub>1</sub>-weighted MRI with and without gadolinium (Fig. 3). The pulse sequence used was the fat suppression T<sub>1</sub>-weighted three-dimensional fast field-echo technique with a repetition time of 95 ms, echo time of 2.3 ms, field of view of 16 cm, matrix size of 160 × 159, slice thickness of 1.0 mm, and acquisition time of 6 min. No apparent growth of the lesion has been observed for 3 years after the surgery, and the patient had no other symptoms related to the cranial nerves in the left cavernous sinus. The patient provided informed consent for this case report.

## Discussion

Meningeal melanocytoma is a rare benign tumor accounting for less than 0.1% of brain tumors.<sup>2)</sup> Since the first case was reported by Limas and Tio,<sup>1)</sup> approximately 130 cases have been reported.<sup>3)</sup> In some cases, the association between intracranial meningeal melanocytoma and a nevus of Ota or a melanosis oculi has been reported suggesting the same embryonic origin (Table 1). Melanoblasts derived from the neural crest migrate to the skin, leptomeninges, ocular structures, and inner ear and differentiate into

melanocytes as early as the 10th week of fetal life. The nevus of Ota is considered to develop when migration of melanocytic cells is arrested at the dermis instead of at the dermo-epidermal union.<sup>4,5)</sup>

Meningeal melanocytoma is frequently found in the upper spinal cord and the posterior fossa, usually forming a mass lesion.<sup>1,3,6)</sup> In contrast, meningeal melanocytoma in and around the cavernous sinus is rare, and to the best of our knowledge, only three cases have been reported thus far<sup>7-9)</sup> (Table 1). Patients with this subset of the tumor usually become symptomatic in young adulthood. In contrast to nodular lesions found in the previous three cases, the lesion in the present case showed an en plaque growth pattern spreading not only along the dura but also into the orbital cavity, making total removal of the lesion without inducing post-operative neurological deficits difficult. Because of the potentially aggressive behavior of this tumor, the residual lesion needs to be followed up closely for regrowth that may occur after several years<sup>9)</sup> or for rare malignant transformation.<sup>10)</sup>

Meningeal melanocytoma is typically iso- to hyper-intense on T<sub>1</sub>-weighted MRI and hypo-intense on T<sub>2</sub>-weighted MRI owing to the presence of melanin in the tumor, which shorten the T<sub>1</sub> and T<sub>2</sub> relaxation time.<sup>11,12)</sup> The contrast enhancement effect with gadolinium appears variable depending on the case. Recently neuromelanin sensitive MRI, a modified T<sub>1</sub>-weighted

sequence with higher sensitivity to neuromelanin, has been introduced to examine structures with high neuromelanin content, such as the substantia nigra and locus ceruleus in patients with Parkinson's disease.<sup>13–15)</sup> Because the melanin-iron structure has an enhancing effect on T<sub>1</sub> shortening, neuromelanin-containing tissues appear as foci of high signal intensity on this image, with the intensity proportional to the neuromelanin concentration.<sup>13)</sup> In this case, this technique was performed for meningeal melanocytoma for the first time, and extension of the lesion was clearly demonstrated with a much better contrast than conventional T<sub>1</sub>-weighted MRI with and without gadolinium. The addition of the fat suppression technique would be useful for evaluating the intra-orbital extension of an en plaque lesion involving the cavernous sinus. However, the limitation of this method would be the difficulty in discriminating pure melanin pigmentation from residual melanocytoma after surgery. Thus, accumulation of cases is necessary to identify characteristic findings that will help discriminate between benign and malignant lesions.

In summary, this is the first report of an en plaque meningeal melanocytoma involving the cavernous sinus, and neuromelanin sensitive MRI may be the technique of choice for close follow-up of meningeal melanocytoma in general, especially when total resection of the lesion is difficult.

#### Informed Consent

The patient has consented to submission of this case report to the journal.

#### Conflicts of Interest Disclosure

All authors have no conflict of interest.

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