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CASE REPORT

# Meningeal melanocytoma in the cerebellopontine angle: A rare case report and review of the literature

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## Abstract

Primary meningeal melanocytoma (MM) in the cerebellopontine angle (CPA) region is an extremely rare neoplasm that originates from the melanocytes in the leptomeninges. These lesions are usually misdiagnosed as they mimic other common CPA lesions through their nonspecific presenting symptoms, signs, and radiological characteristics. Here, we report a 47-year-old Chinese female patient who presented with a 1-month history of the right-sided tongue numbness and 1-week history of the right-sided face numbness that had been worsening for 2 days. The tumor, in the right CPA region, showed a slight isointensity on T1-weighted image and mixed signal intensity on T2-weighted image. The clinical presentation, surgical treatment, and pathologic characteristics were determined. The tumor was microsurgically resected and gross-total resection was achieved. The tumor revealed a solid, capsulated, brown-black lesion. Immunohistochemistry showed that the tumor cells were positive for human melanoma black-45 (HMB-45), melanoma antigen (MelanA), S100, SOX10, and BRAF, confirming the final diagnosis of meningeal melanocytoma. Ultimately, no signs of radiological local recurrence were observed during the two-year follow-up. Collectively, meningeal melanocytoma is difficult to distinguish from common tumors in the CPA region before operation due to the lack of specificity in imaging and symptoms. Complete surgical resection is the best therapeutic option for this tumor. Although the tumor is commonly considered as a benign lesion, recurrence and metastasis are common, and pathogenesis remains unclear.

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Primary meningeal melanocytomas (MMs), first reported by Limas and Tio in 1972 [1] are extremely rare primary brain tumors, accounting for only about 0.07% of all tumors in the central nervous system (CNS), with an annual incidence of 1 case per 10 million individuals [2–3]. MMs are benign lesions originating from the melanocytes of the leptomeninges, and they often present in the leptomeninges adjacent to the medulla oblongata and anterolateral spinal cord because of the abundance of melanocytes here [1]. MMs can occur in different areas of the brain but are commonly seen in the region of the foramen magnum [1], the posterior fossa [4], cervical spinal region [5–7], anterior cranial fossa [8], and the sellar region [9]. However, they rarely occur in the cerebellopontine angle (CPA) region [10–11]. To the best of our knowledge, only ten studies of MMs in the CPA have been published.

Here, we elucidate a case of a pathologically diagnosed MM in the CPA region, which had been treated at our department. We focus on the clinical presentation, surgical treatment, and pathologic characteristics, and review the related literature.

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## Clinical summary

### History and physical examination

A 47-year-old Chinese woman presented with right-sided tongue numbness for one month and right-sided face numbness for one week, which had been worsening for 2 days prior to admission. She had no headache, hearing loss, or other manifestations of cranial nerve dysfunction. After admission, physical examination findings showed decreased sensation on the right side of the tongue and right-sided mild facial paralysis. There were no mucocutaneous or ocular pigmented lesions. Our study was conducted in accordance with the declaration of Helsinki, and was approved by the Ethics Committee of Tongji Hospital of Wuhan. Written informed consent was obtained from the patient.

### Imaging findings

Preoperative computed tomography (CT) displayed a slight hyperdense and well-circumscribed lesion in the right CPA region (Fig. 1a). MRI also revealed a solid oval mass (26 mm × 19 mm) attached to the dura of petrosal surface in the right CPA. The lesion was isointense on T1-weighted image (Fig. 1b) and of mixed signal intensity on T2-weighted image (Fig. 1c), and it showed intense and homogenous enhancement after gadolinium administration (Fig. 1d and 1e). No evidence of angiopathic abnormalities was found on magnetic resonance angiography scan (Fig. 1f).

### Therapeutic intervention

The right retrosigmoid suboccipital approach was performed and the lesion was microsurgically resected under continuous intraoperative neurophysiologic monitoring. Microscopy revealed a brown-black solid lesion in the right CPA region. The tumor seemed to derive from the dura mater, and the brainstem was slightly compressed. The lesion was a slightly soft, well-circumscribed brown-black pigmented tumor, and a gross-total resection was achieved.

### Pathological results

Tumor sections stained with hematoxylin and eosin (H&E) comprised of a large number of spindle-shaped cells, with a strip distribution and focal storiform pattern, and could be markedly observed in tumor tissues (Fig. 2a). Many spherical nucleus cells that contained granules of dark brown pigment were distributed among the tumor cells (Fig. 2b). However, there were rare mitotic images and areas of necrosis in the region of the tumor. The tumor cells showed strongly positive staining for some relatively special markers of meningeal melanocytoma including HMB-45, MelanA, S100, SOX10, and BRAF (Fig. 2c–2g). Furthermore, cellular proliferative activity assessed by Ki67 labeling index was low, approximately 5% (Fig. 2h). The final diagnosis of MM was established according to

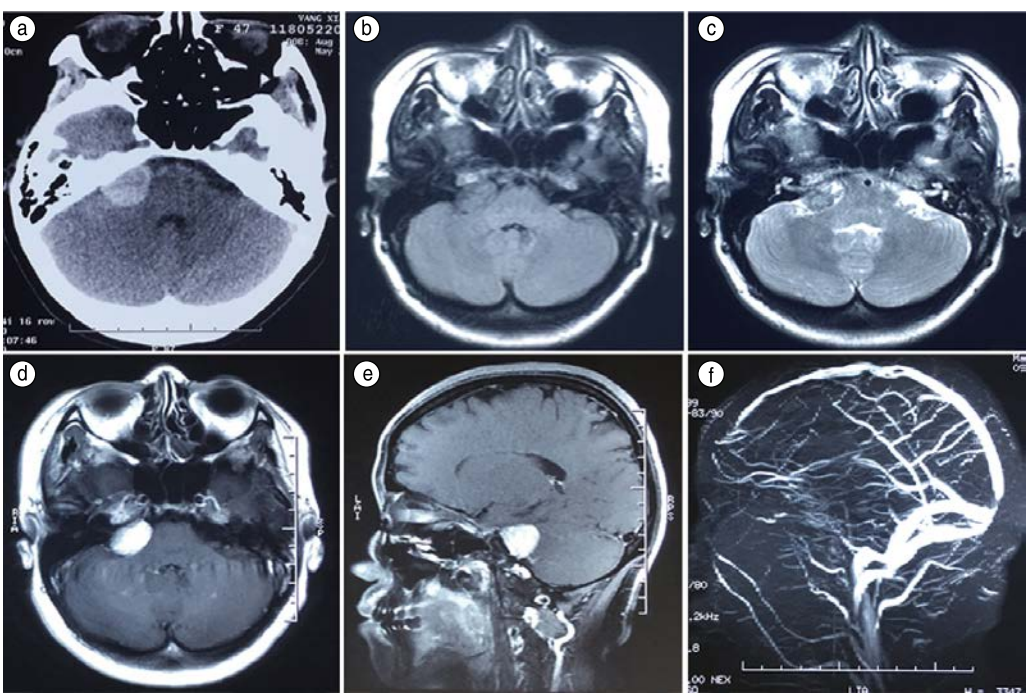


**Fig. 1** Imaging findings (a) CT showed a hyperdense and well-circumscribed lesion in the right cerebellopontine angle (CPA) region; (b and c) The lesion was isointense on T1-weighted sequences (b) and of mixed signal intensity on T2-weighted sequences (c); (d and e) T1-weighted images presented intense and homogenous enhancement in the right CPA; (f) Magnetic resonance angiography (MRA) scan showed no obvious abnormity

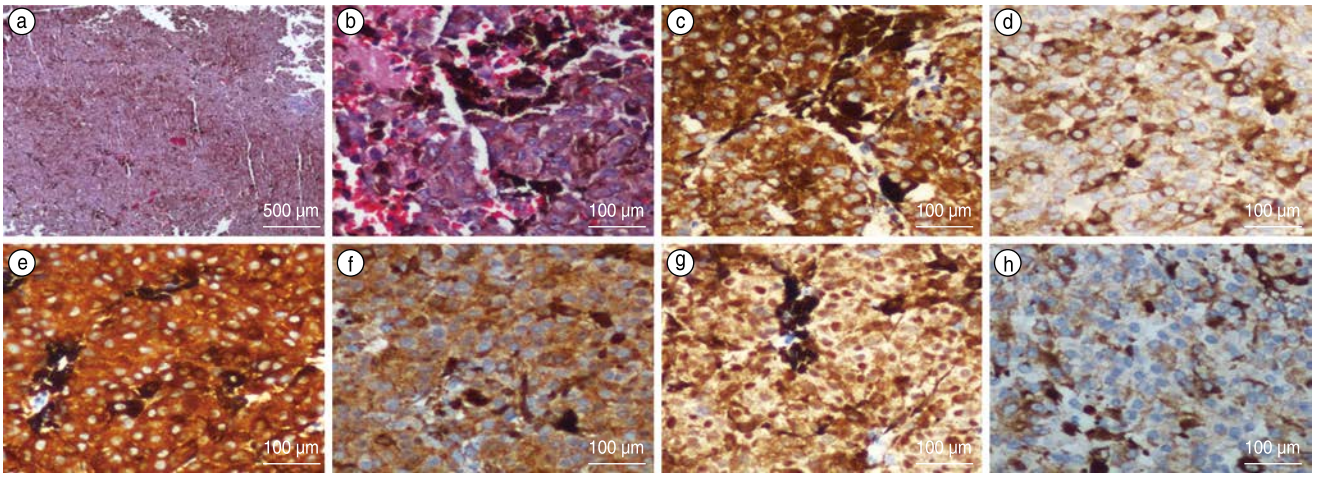


**Fig. 2** Histopathological findings. (a and b) Tumor cells arranged in fascicles interspersed with melanin-containing cells in H&E staining; (c–h) The tumor cells were positive for HMB-45 (c), Melan A (d), S100 (e), SOX10 (f), BRAF (g), and Ki67 (h). (× 200)

the histopathological and immunohistochemical results.

### Follow-up

Postoperatively, the symptoms eventually resolved, and there was no focal neurologic deficit or complications associated with surgery upon discharge. The patient underwent neither radiotherapy nor chemotherapy after surgery. During the two-year follow-up, no evidence of local recurrence was observed (Fig. 3a and 3b).

## Discussion

Primary pigmented tumors of the leptomeninges, which include pigmented meningioma, primary central nervous system (CNS) malignant melanoma, melancholic schwannoma, melanoblastosis, and MMs, are rare lesions in the brain [12–13]. MMs are exceptionally rare benign tumors of the central nervous system [8, 12]. Additionally, the occurrence of meningeal melanocytoma in the CPA region is extremely uncommon, with only ten cases previously described in relevant literature (Table 1). Although the tumor attaching to the meninges often grossly resembles meningioma on radiology [10, 14–15], MMs are ultrastructurally similar to melanoma in their histology and pathology [14–16]. Gratifyingly, they commonly shows a favorable clinical prognosis [17]. Importantly, because the biological behavior, management, and prognosis of MMs is different from meningioma, it is necessary to distinguish those tumors in order to make a suitable therapeutic strategy and prognosis for MM.

Lacking distinctive clinical symptoms, MMs are often difficult to distinguish from some usual lesions of the CPA before operation, such as vestibular schwannomas, meningiomas, and epidermoid tumors. Preoperative

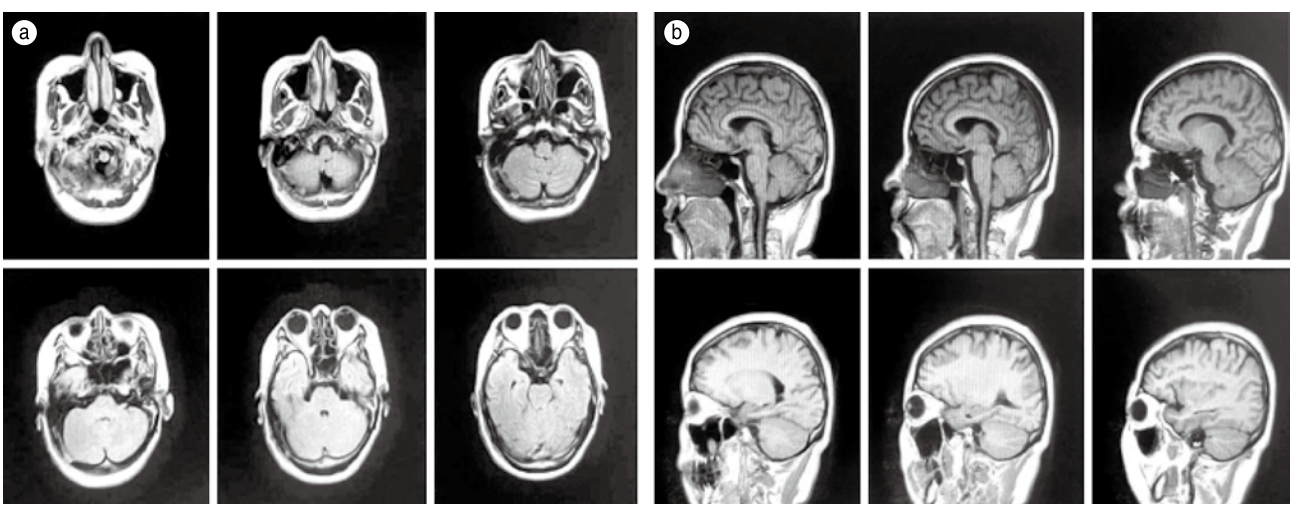


**Fig. 3** Imaging results of 2 years after operation (a and b). There is no recurrence of the tumor on magnetic resonance imaging

**Table 1** Cases of meningeal melanocytoma of CPA

| Case and reference | Age/sex | Tumour lacation | T1: T2 weighted imaging | Treatmecnt | Immunohistoche mical finding | Ki67(%) | Recurrence Time of Dead |
|---|---|---|---|---|---|---|---|
| Prabhu,1993 [32] | 67/Women | CPA | N | S | HMB45(+)S100(+) | – | NR/4month(D) |
| Gardiman,1996 [33] | 19/Men | CPA | N | S | Vimentin(+)S100(+)HMB45(–) | – | NR |
| Gardiman,1996 [33] | 43/Men | CPA | N | S | Vimentin(+)S100(+)HMB45(–) | – | Re/5month(D) |
| Clarke,1998 [21] | 30/Women | CPA | Hyper-intensity; N | S&R | Vimentin(+)S100(+) | – | Re/6month(D) |
| Hamasaki, 2002 [18] | 59/Men | CPA | Hyper-intensity; Hypo-intensity | S&R | HMB45(+)S100(+)HMB45(-) | – | NR/2 years |
| Horst, 2005 [26] | 38/Men | CPA | Hyperintense; iso-hypointense | S | MelanA(+) | – | Re/5 years |
| Gupta, 2007 [13] | 58/Women | CPA | Heterogeneously Hyperintense; hypointense | S | HMB45(+)vimentin(+)MelanA(+) EMA(–) | – | NR |
| Masaaki, 2011 [34] | 43/Women | CPA | Hyperintense; hypointense | S | HMB45(+)S100(+) | 4%–5% | Re/20 month |
| Gjergji, 2011 [10] | 47/Men | CPA | Hyperintense; iso-hypointense | S | – | 10% | Re/20 month |
| Rasha, 2018 [11] | 46/Men | CPA | Hyperintense; hypointense | S | S100(+) | – | N |
| This case | 47/Women | CPA | Iso-hypointense; Heterogeneously Hyperintense | S | HMB45(+)BRAF(+)MelanA(+) S100(+)SOX10(+) | 5% | NR |

CPA: cerebellopontine angle; N: no evidence; S: surgical; R: radiotherapy; Re: recurrence; B: biopsy; NR: not recurrence; D: dead

imaging studies are also nonspecific and inconclusive for MMs, leading to frequent preoperative misdiagnoses. On CT scan, MMs appear as a well-circumscribed, iso- to slightly hyperdense lesion with dural attachment [18–19]. Congruently, the tumor, in our case, was hyperdense and well-circumscribed on CT scan. The signal pattern of MMs on MRI scan varies widely, depending on melanin and presence of hemorrhage. Generally, the tumor was hyperintense or isointense on T1-weighted sequences and isointense or slightly hypointense on T2-weighted sequences with homogeneous enhancement on contrast-enhanced MRI [9, 19]. Recently, Shoko summarized the characteristic image features of 14 cases of intracranial MMs documented in English literature and found that eleven cases showed hyperintensity on T1-weighted images and were hypointense on T2-weighted images [20]. However, the MM in our case was isointense on T1-weighted sequences, with homogenous enhancement after gadolinium administration, and of mixed signal intensity on T2-weighted sequences, which is not consistent with the general characteristic MRI features of MMs. The degree and distribution of the melanin pigments and presence of hemorrhage in the tumor might have resulted in an uncommon presentation in the MRI in our case. We then summarized the MRI features of MMs in the CPA region in the English literature, which are generally in line with the characteristic MRI features, with high signal intensity on T1-weighted images and low signal intensity on T2-weighted images (Table 1). Collectively, it is difficult to provide preoperative value through MRI studies in the diagnosis of meningeal melanocytoma.

The definitive diagnosis of MM and the differential diagnosis are best achieved postoperatively through histopathological and immunohistochemical studies. The histopathological characteristics of MMs have been documented [10]. Briefly, the general morphology of MMs usually presents as an encapsulated black or dark brown solid lesion that is tightly attached to the underlying meninges [13], as seen in the present case. Microscopically, there are several melanin granules in the tumor cells. It can obviously be observed that tumor cells with prominently spherical nuclei are arranged in bundles and enriched with melanin pigments. Characteristic immunohistochemical reaction of this lesion includes a positive response to HMB-45, Melan-A, Vimentin, and S-100 [8, 12, 21]. HMB-45 and Melan-A are two specific markers detectable in most melanocytic lesions, and S-100 and vimentin are indicators of cells with mesenchymal origin. However, the antigen of EMA and Leu-7 are usually negative, differentiating MMs from melanotic meningioma and melanotic schwannoma [4, 22]. The differential diagnosis between MM and malignant melanoma can sometime be difficult as they share some common histological features. However, melanocytomas are characterized by nuclear pleomorphism, necrosis, and a lack of or decreased mitosis. Importantly, Ki-67 labeling index, which is an important marker to evaluate malignancy and predict the recurrence of tumors, is often expressed less than 5% in MMs and more than 10% in malignant melanomas [9, 23]. In this case, we found that the lesion was enriched with melanin, and the cells were arranged in sheaf with obvious nuclei. Immunohistochemical findings showed the antigens of HMB45, BRAF, MelanA, S100, and SOX10 to be positive, and the Ki-67 labeling index was approximately 5%, which corresponded with the pre-descriptive characteristics of MM on pathology and immunohistochemistry.

Although MMs are histologically benign tumors, malignant transformation and recurrence have been reported, despite complete surgical removal. In 2003, Uozumi *et al* reported the first case of malignant transformation of an intracranial MM. The patient

underwent gross total removal of the MM. However, four years later, the patient underwent radiotherapy and an additional operation because of a local recurrence. Histopathological examination revealed a malignant melanoma originating from a melanocytoma, and the patient died from cerebrospinal fluid dissemination. [24] Wang *et al* also presented a case of malignant transformation of an intracranial supratentorial MM which recurred as malignant melanoma 3 years after total resection [25]. Roser *et al* described a case of malignant transition of an intracranial melanocytoma into a melanoma 12 years after subtotal tumor resection of the initial tumor. The patient died 4 months after the second operation from a rapid diffuse meningeal spread of the tumor, despite a combination of whole brain radiotherapy and chemotherapy [23]. As for the recurrence of MMs, Koch *et al* also reported a patient with MM of the CPA who suffered a first local recurrence 5 years after surgical resection and a second local recurrence 6 years after diagnosis, with tumor cells seeding to the intracerebral and spinal meninges (Table 1) [26]. Naturally, the frequent phenomenon of malignant transformation or recurrence has prompted researchers to explore its pathogenesis. There are a few possible causes for the transformation or recurrence of MMs: first, most scholars believe that MMs exhibit relatively benign characteristics [27]; second, the region of the CPA is complicated, which precludes complete removal of the tumor [3]. However, with respect to the present case, no sign of local recurrence was observed at the postoperative two-year follow-up.

To the best of our knowledge, there is no specific guideline for the management of the MMs as they are rare tumors. The best therapeutic option for MMs is complete removal of the tumor whenever possible [28]. Rades reviewed 89 patients with MMs and found that the five-year survival rate and the five-year local control were 100% and 80%, respectively, in patients with complete resection, but only 46% and 18%, respectively, in those with incomplete resection [29], indicating the vital role of complete resection in the outcome of MMs. Complete tumor removal was also achieved in the present case, and no recurrence was observed at the postoperative two-year follow-up. However, failure of complete resection of the lesion can be attributed to several factors, such as poor tumor location, involvement of vital structures or skull base dura [8], excessive tumor bleeding [9, 21], and excessive tumor volume [30]. Noteworthy, Rades also found that the five-year survival rate was 100% in patients with combined incomplete resection and postoperative radiotherapy versus 46% in patients with incomplete resection only [29], suggesting that postoperative radiotherapy should be advised in cases of incomplete resection. Other cases of MMs, wherein tumors were partially removed by surgery and postoperative radiotherapy was beneficial, have been reported. Kurita *et al* described a case of MM from the Meckel's cave, wherein gamma knife radiotherapy was performed after the tumor was partially removed. The tumor showed marked shrinkage without complication after three years of irradiation [31]. Hamasaki *et al.* also presented a patient, with residual MM tumor in the CPA region, who underwent radiosurgery and showed no evidence of regrowth or metastasis 24 months later (Table 1) [18]. Clarke reported another case of MM in the CPA, wherein radiotherapy was performed after the tumor was partially removed. Although regrowth of the residual tumor was observed, the tumor was much less vascular after radiotherapy and a complete resection was achieved upon second operation (Table 1) [21]. Kuo *et al* reported that the survival time of MM between surgery alone and surgery plus radiotherapy was not significantly different. However, adjuvant radiotherapy showed a trend towards improved survival [28]. In our retrospective study for MM of the CPA, all 8 patients with CPA MM underwent surgical treatment, of which 2 patients underwent surgery plus adjuvant radiotherapy and 6 patients underwent surgery solely. The observed survival time was 4 months to 5 years. Table 1 shows the clinical characteristics and prognosis of the 11 cases of CPA MM. Therefore, to the best of our knowledge and based on the published data, complete removal of the tumor is the best therapeutic option for MM, and postoperative radiotherapy appears to improve both local control and survival. However, further clinical evidence-based research is needed to draw a definite conclusions regarding the benefits of radiotherapy in MM treatment. Although MM is rare in the central nervous system, it is often misdiagnosed as common tumors in the CPA region before surgical resection due to the lack of specificity in imaging and in the patient's symptoms. Although the tumor is usually considered to be benign, recurrence and metastasis have also been reported. Total resection is the best therapeutic option. Some adjuvant treatments, such as chemotherapy and radiotherapy, can be selectively considered. However, their curative effect needs further verification.

### Conflicts of interest

The authors indicated no potential conflicts of interest.

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