Case Report

Primary meningeal melanocytoma of the cerebellopontine angle associated with ipsilateral nevus of Ota: A case report

Rasha Elbadry, Ahmed Abd Elazim, Kazim Mohamed, Mamdouh Issa¹, Ali Ayyad²

Abstract

**Background:** Cerebellopontine angle represents a complex anatomical area of the brain. A cerebellopontine angle lesion could be a vestibular schwannoma, meningioma, epidermoid cyst, or less likely, arachnoid cyst, metastasis, lower cranial nerves schwannoma, lipoma, hemangioma, paraganglioma, or vertebra-basilar dolichoectasia. Primary meningeal melanocytoma is a rare neoplasm, especially when it occurs at the cerebellopontine angle. Nevus of Ota (aka oculodermal melanocytosis) is a hyperpigmentation along the distribution of the ophthalmic and maxillary branches of trigeminal nerve; it occurs due to entrapment of melanocytes at the upper third of the dermis. It may not present at birth and may show up at puberty.

**Case Description:** We describe a case of primary meningeal melanocytoma of the cerebellopontine angle associated with nevus of Ota in a 46-year-old male patient presented with 7-day history of left arm weakness and vertigo. Computed tomography and MRI showed right-sided cerebellopontine angle mass, which was resected. Histopathology confirmed the meningeal melanocytic lesion and revealed its nature.

**Conclusion:** Primary meningeal melanocytoma of the brain is a rare but benign tumor; the association between meningeal melanocytoma and nevus of Ota is also rare and possibly explained by their common embryonic origin from neural crest cells. There are six cases reported so far in literature including our case for meningeal melanocytoma associated with nevus of Ota.

**Key Words:** Melanin, melanocytoma, meninges and cerebellopontine angle, nevus

INTRODUCTION

Cerebellopontine angle is considered a delicate anatomical region of the brain. A wide variety of cerebellopontine angle lesions exist. These could be a vestibular schwannoma, meningioma, epidermoid cyst, or less likely, arachnoid cyst, metastasis, lower cranial nerves schwannoma, lipoma, hemangioma, paraganglioma, or vertebra-basilar dolichoectasia. Primary meningeal melanocytoma is considered a rare neoplasm, particularly

How to cite this article: Elbadry R, Abd Elazim A, Mohamed K, Issa M, Ayyad A. Primary meningeal melanocytoma of the cerebellopontine angle associated with ipsilateral nevus of Ota: A case report. Surg Neurol Int 2018;9:245. http://surgicalneurologyint.com/Primary-meningeal-melanocytoma-of-the-cerebellopontine-angle-associated-with-ipsilateral-nevus-of-Ota-A-case-report/
when it occurs at the cerebellopontine angle. Nevus of Ota (aka oculodermal melanocytosis)\textsuperscript{16} is a hyperpigmentation along the distribution of the ophthalmic and maxillary branches of trigeminal nerve;\textsuperscript{22} it occurs due to entrapment of melanocytes at the upper third of the dermis. It may not present at birth and may show up at puberty. We present a rare association between Nevus of Ota and cerebellopontine angle primary meningeal melanocytoma.

**CASE REPORT**

A 46-year-old Sri Lankan male patient without comorbidities presented with 1-week history of left arm weakness and vertigo of gradual onset and progressive course. He denied headache, vomiting, or convulsions. He had a history of nevus of Ota at the right side of the face. There is a positive family history of nevus of Ota.

The patient was fully conscious and oriented, vitally stable, and has a bluish discoloration of the right frontal, periorbital region and buccal mucosa [Figure 1], right upper motor neuron lesion facial palsy, decreased sensation over the right ophthalmic and maxillary territories of trigeminal nerve, and left upper motor neuron lesion weakness more in upper limb than in lower limb.

Computed tomography showed right-sided CP angle extra-axial mass lesion of mixed high and iso-attenuation value and showed faint postcontrast enhancement extending over the petrous apex to the right parasellar region medial to the right temporal lobe [Figure 2a]. With MRI, axial T1-weighted images showed lobulated intracranial extra-axial hyperintense lesion at the right CP angle and creeping along the petrous apex to the right parasellar region compressing the brain stem and indenting the medial aspect of the right temporal lobe. Axial T2-weighted images showed the complex texture of the lesion with predominant low signal intensity and cystic areas of iso to high signal intensity. Axial susceptibility-weighted images showed ferromagnetic effects of the contents of the lesion with patchy areas of low signal intensity within a cystic lesion. Axial, sagittal, and coronal contrast-enhanced T1-weighted images of the brain showed increase in the hyperintensity of the lesion

![Figure 1: Showing right frontal (nevus of Ota) and buccal bluish discoloration](image)

![Figure 2: (a) Axial CT sections before and after IV contrast administration showing right-sided CP angle extra-axial mass lesion of mixed high and iso-attenuation value showing faint postcontrast enhancement extending over the petrous apex to the right parasellar region medial to the right temporal lobe. (b) Axial T1-weighted images showing lobulated intracranial extra-axial hyperintense lesion at the right CP angle and creeping along the petrous apex to the right parasellar region compressing the brain stem and indenting the medial aspect of the right temporal lobe. (c) Axial T2-weighted images showing the complex texture of the lesion with predominant low signal intensity and cystic areas of iso to high signal intensity. (d) Axial susceptibility-weighted images (SWI) showing ferromagnetic effects of the contents of the lesion with patchy areas of low signal intensity within a cystic lesion. (e) Axial, sagittal, and coronal contrast-enhanced T1-weighted images of the brain showing increase in the hyperintensity of the lesion denoting its enhancement and showing the extension of the lesion from the right CP angle region to the right parasellar region with its mass effect on the brain stem and the right temporal lobe](image)
denoting its enhancement and showed the extension of the lesion from the right CP angle region to the right parasellar region with its mass effect on the brain stem and the right temporal lobe [Figure 2b-e].

The patient underwent two-staged operations to resect the lesion utilizing two different approaches to secure proper resection. The first stage was through right retroseigmatic approach via 3-cm craniotomy. Facial, trigeminal, glossopharyngeal, and vagus nerves were visualized. There was a bluish lesion surrounding the trigeminal nerve. The part of the lesion that was accessible removed completely and part of it was resembling a hematoma. The second stage was through a right frontotemporal craniotomy; there was bluish discoloration of the skin, temporalis muscle, and dura [Figure 3]. Total resection of the tumor was achieved [Figure 4]. The tumor was invading the cavernous sinus and we used Dolenc approach (extradural transcavernous approach) to reach it; however, it was not invading the Gasserian ganglion.

Meningeal nodules with heavy brownish pigmentation showed heavily pigmented melanocytes and S-100 positivity without atypia or necrosis concluding a meningeal melanocytoma [Figure 5a and b].

**DISCUSSION**

Nevus of Ota is a bluish nevus usually found unilaterally in the face involving branches 1 and 2 of the trigeminal nerve, appearing in the sclera, mucous membranes, and adjacent periorbital skin because of the presence of dermal melanocytes.[17] There are 26 cases of melanocytic lesions of the face described by Ota in 1939, also known as nevus fusco-caeruleus ophthalmomaxillaris.[12] It usually occurs among Asians compared with other races.[8] Women-to-men ratio is 4:1 with majority presenting at birth, but it can also present at puberty.[4]

Disorders that can be seen with nevus of Ota are Sturge–Weber syndrome, Klippel–Trenaunay syndrome, arteriovenous malformation, neurofibromatosis, Takayasu disease, and melanomas.[8]

Meningeal melanocytomas are benign-pigmented tumors arising from melanocytes; their incidence to occur in the brain is rare (0.06–0.1% of brain tumors) compared with melanomas.[3] Rahimi-Movaghar and Karimi[15] described 95 cases of meningeal melanocytoma; 45 cases were intracranial, mostly infratentorial, and 50 cases were located in the spine and spinal roots. The disease tends to occur more in women with a median age of 45 years in intracranial cases but lower age in spinal cases.[15]

Clinical presentation of meningeal melanocytomas depends on the anatomical location and neurological structures compressed. Meningeal melanocytoma can present with intracerebral hemorrhage; Hino et al.[5] reported a 75-year-old woman presented with intracerebral hemorrhage related to a meningeal melanocytoma with a history of ipsilateral nevus of Ota.

According to the World Health Organization, melanotic neoplasms of the central nervous system are classified into melanocytosis, melanomatositis, melanocytoma, and malignant melanoma.[6]

Histopathologically, meningeal melanocytoma cells appear in sheets or bundles with occasional whorl
Table 1: Reported meningeal melanocytomas

| Reference                  | Age/Sex | Site            | Symptoms                                      | Hemorrhage | Operation          | Radiotherapy | Outcome |
|----------------------------|---------|-----------------|-----------------------------------------------|------------|--------------------|--------------|---------|
| Botticelli et al.          | 43/F    | Meckel’s cave   | Abducent nerve palsy, ptosis                   | None       | Parietal removal   | +            | Alive   |
| Hino et al.                 | 75/F    | Frontal         | Loss of consciousness, exophthalmos, hemiparesis | Yes        | Parietal removal   | -            | Alive   |
| Navas et al.               | 25/M    | Temporal        | Ptosis                                        | None       | Pterional removal  | -            | Unknown |
| Our case                   | 46/M    | Cerebellopontine angle | Left arm weakness, vertigo                  | Yes        | Posterior, frontotemporal removal | -          | Alive   |
| Piercejchi-Marti et al.    | 25/M    | Rolandic        | Headache, seizure                             | None       | Temporal removal   | -            | Alive   |
| Rahimi-Movaghar and Karimi | 17/M    | Parietal        | Headache, blindness                           | None       | Temporal removal   | -            | Alive   |

appearance containing a high amount of melanin in the cytoplasm. Meningeal melanocytoma can be differentiated from malignant melanoma in which meningeal melanocytoma lacks excessive mitosis, necrosis, and hemorrhage, in addition to its own cytoarchitecture. Meningeal melanocytoma is positive for S-100, vimentin, and HMB-45, whereas it is negative for epithelial membrane antigen.

Radiologically, meningeal melanocytoma appears homogenously hyperintense on T1-weighted images with contrast enhancement by gadolinium and hypointense on T2-weighted images owing to its melanin content. Heterogenous density can be seen in intralesional hematoma.

In the 95 case series of Rahimi-Movaghar and Karimi, the recurrence rate of meningeal melanocytoma was 26.3%, whereas the tumor-related death was 10.5%. The best treatment option for meningeal melanocytoma is not well supported based on our literature review. Complete resection versus incomplete resection plus or minus radiotherapy showed a nonsignificant difference in overall survival rates based on treatment modalities of Rahimi-Movaghar and Karimi and Rades et al. case series. Chemotherapy has no remarkable role in treating meningeal melanocytoma based on our review of the literature. Meningeal melanocytoma can sometimes behave malignantly and metastasize especially recurrent tumors.

The association between meningeal melanocytoma and nevus of Ota is rare, based on our review of the literature; there are six cases of meningeal melanocytoma associated with nevus of Ota including our case [Table 1].

Our case is the first case of cerebellopontine angle meningeal melanocytoma to be associated with nevus of Ota. Nevus of Ota arises from melanoblasts that originate from neural crest cells; melanoblasts migrate to the skin, leptomeninges, ocular structures, and inner ear. Meningeal melanocytoma cells share the same embryonic origin of nevus of Ota being derived from neural crest cells, hence the possible association.

CONCLUSION

Primary meningeal melanocytoma of the brain is a rare but benign tumor; the association between meningeal melanocytoma and nevus of Ota is also rare and possibly explained by their common embryonic origin from neural crest cells. There are six cases reported so far in literature including our case for meningeal melanocytoma associated with nevus of Ota.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Botticelli AR, Villani M, Angiari P, Peserico L. Meningeal melanocytoma of Meckel’s cave associated with ipsilateral Ota’s nevus case report. Cancer 1983;51:2304-10.
2. Tran HH, Kono T. Nevus of Ota: Clinical aspects and management. Skinmed 2003;2:89-98.
3. Chow M, Clarke DB, Maloney WJ, Sangalang V. Meningeal melanocytoma of the planum sphenoidale: Case report and review of the literature. J Neurosurg 2001;94:841-5.
4. Hidano A, Kajima H, Ikeda M, Miyasato H, Niimura M. Natural history of nevus of Ota. Arch Dermatol 1983;51:2304-10.
5. Hino K, Nagane M, Fujioka Y, Shiokawa Y. Meningeal melanocytoma of cerebellopontine angle: A case report with 12 years follow up. Basic Appl Pathol 2011;4:99-102.
6. Jaiswal S, Jaiswal AK, Bijay M, Behari S, Pandey R. Primary meningeal melanocytoma of cerebellopontine angle: A case report with 12 years follow up. Basic Appl Pathol 2011;4:99-102.
7. Kawaguchi T, Kawano T, Kazekawa K, Nakashima S, Honma T, Kaneko Y, et al. Meningeal melanocytoma in the left frontal region. Brain Tumor Pathol 1998;15:58-62.
8. Kopf A, Weidman AI: Nevus of Ota. Arch Dermatol 1962;85:75-88.
9. Kurita H, Segawa H, Shin M, Ueki K, Ichi S, Sasaki T, et al: Radiosurgery of meningeal melanocytoma. J Neurooncol 2000;46:57-61.
10. Maize J, Ackerman B: Pigmented Lesions of the Skin. Philadelphia: Bernard, Lea and Febiger; 1987. p. 73-162.
11. Navas M, Pascual JM, Fragas J, Pedrosa M, Shakur S, Carrasco R, et al: Intracranial intermediate-grade meningeal melanocytoma with increased cellular proliferative index: An illustrative case associated with a nevus of Ota. J Neurooncol 2009;95:105-15.
12. Ota M: Nevus fusco-caeruleus ophthalmomaxillaris. Jpn J Dermatol 1939;46:369-72.
13. Piercecchi-Marti MD, Mohamed H, Liprandi A, Gambarelli D, Grisoli F, Pellissier JF: Intracranial meningeal melanocytoma associated with ipsilateral nevus of Ota. Case report. J Neurosurg 2002;96:619-23.
14. Rades D, Heidenreich F, Tatagiba M, Brandis A, Karstens JH: Therapeutic options for meningeal melanocytoma: Case report. J Neurosurg 2001;95:225-31.
15. Rahimi-Movaghar V, Karimi M: Meningeal melanocytoma of the brain and oculodermal melanocytosis (nevus of Ota): Case report and literature review. Surg Neurol 2003;59:200-10.
16. Rapini RP, Bologna JL, Jorizzo JL: Dermatology: 2-Volume Set. St. Louis: Mosby. ISBN 1-4160-2999-0; 2007.
17. Stuart C: Naevus of Ota. Br J Dermatol 1955;67:317-9.
18. Uematsu Y, Yukawa S, Yokote H, Itakura T, Hayashi S, Komai N: Meningeal melanocytoma: Magnetic resonance imaging characteristics and pathological features: Case report. J Neurosurg 1992;76:705-10.
19. Winston KR, Sotrel A, Schnitz SJ: Meningeal melanocytoma: case report and review of the clinical and histological features. J Neurosurg 1987;66:50-7.