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Case Report

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

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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

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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.[6] Primary meningeal melanocytoma is considered a rare neoplasm, particularly

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when it occurs at the cerebellopontine angle. Nevus of Ota (aka oculodermal melanocytosis)[16] is a hyperpigmentation along the distribution of the ophthalmic and maxillary branches of trigeminal nerve;[2] 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.

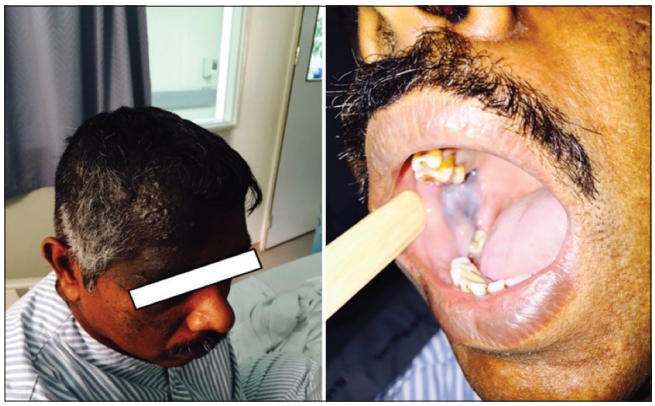

Figure 1: Showing right frontal (nevus of Ota) and buccal bluish

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

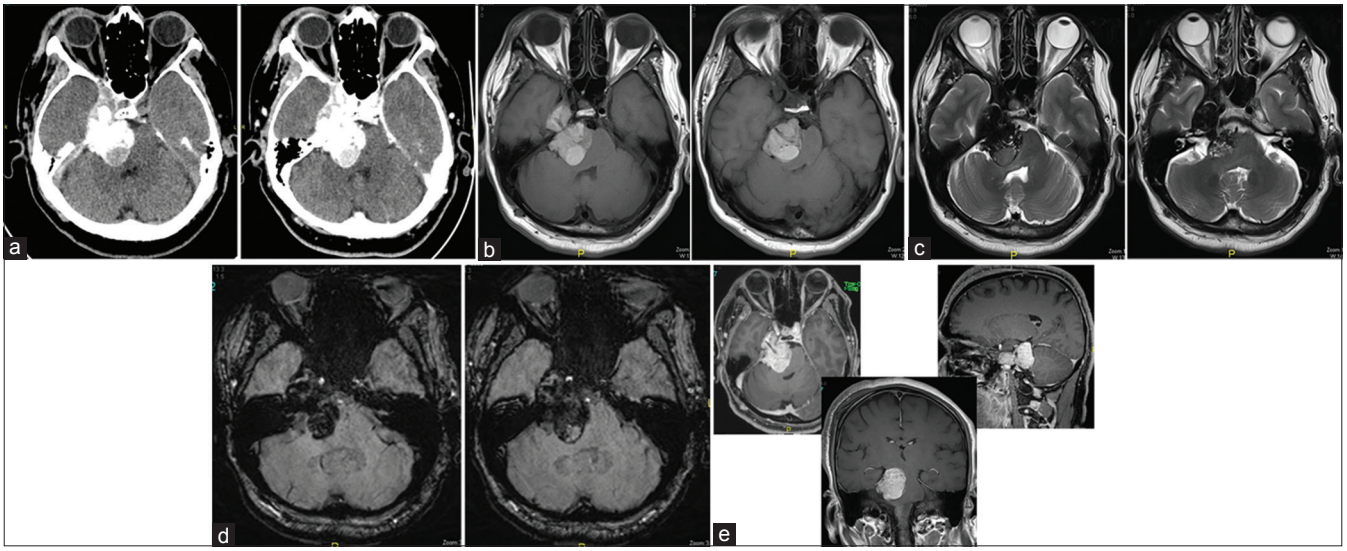

Figure 2: (a) Axial CT sections before and after IV contrast administration showing right-sided CP angle extra-axial mass lesion of mixed high and iso-attenuation value showing faint postcontrast enhancement extending over the petrous apex to the right parasellar region medial to the right temporal lobe. (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 hyperintenisty 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

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 retrosegmoid 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 fuscu-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.[3,5] 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.

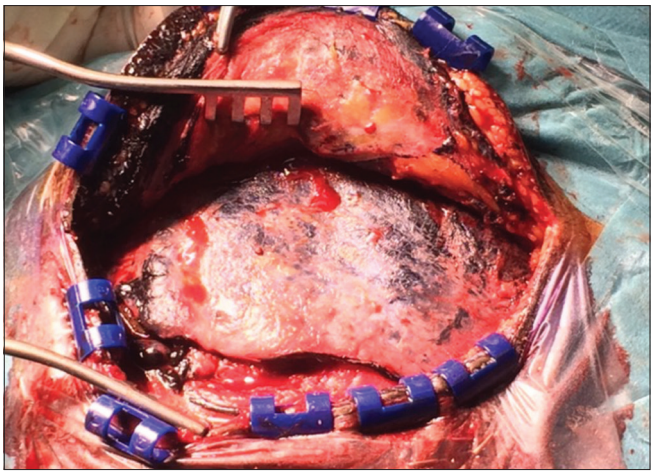

Figure 3: Bluish discoloration of the skin and temporalis muscle

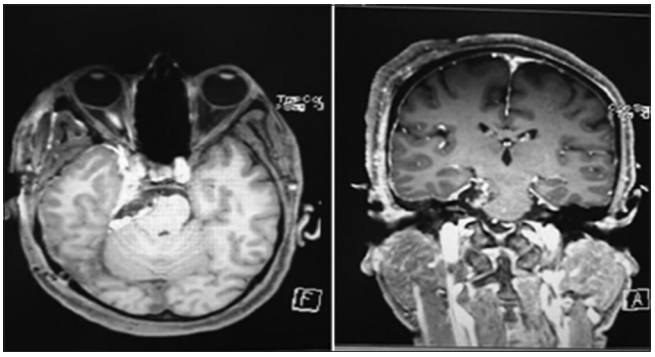

Figure 4: Postoperative MRI brain (axial and coronal) with contrast after lesion resection

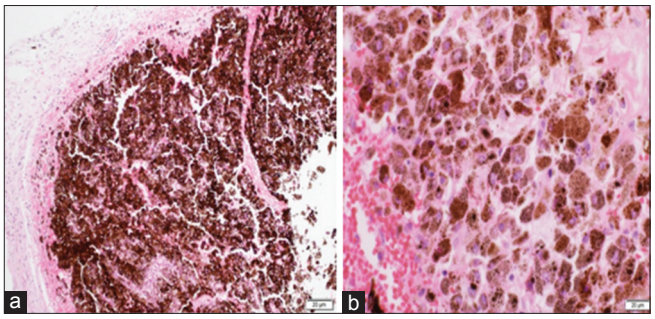

Figure 5: [a (Left) and b (right)] Meningeal nodules with heavy brownish pigmentation showing heavily pigmented melanocytes without atypia or necrosis

According to the World Health Organization, melanotic neoplasms of the central nervous system are classified into melanocytosis, melanomatosis, 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*.[17]	43/F	Meckel's cave	Abducent nerve palsy, ptosis	None	Parietal removal	+	Alive
Hino *et al*.[8]	75/F	Frontal	Loss of conscious, exophthalmos, hemiparesis	Yes	Parietal removal	–	Alive
Navas *et al*.[19]	25/M	Temporal	Ptosis	None	Pterional removal	-	Unknown
Our case	46/M	Cerebellopontine angle	Left arm weakness, vertigo	Yes	Posterior, frontotemporal removal	-	Alive
Piercecchi-Marti *et al*.[18]	25/M	Rolandic	Headache, seizure	None	Temporal removal	-	Alive
Rahimi-Movaghar and Karimi[9]	17/M	Parietal	Headache, blindness	None	Temporal removal	-	Alive

appearance containing a high amount of melanin in the cytoplasm.[3] Meningeal melanocytoma can be differentiated from malignant melanoma in which meningeal melanocytoma lacks excessive mitosis, necrosis, and hemorrhage, in addition to its own cytoarchitecture.[5] Meningeal melanocytoma is positive for S-100, vimentin, and HMB-45, whereas it is negative for epithelial membrane antigen.[5,7,9,15,18,19]

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.[18] Heterogenous density can be seen in intralesional hematoma.[5]

In the 95 case series of Rahimi-Movaghar and Karimi,[15] the recurrence rate of meningeal melanocyoma 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[15] and Rades *et al*.[14] case series. Chemotherapy has no remarkable rule in treating meningeal melanocytoma based on our review of the literature. Meningeal melanocytoma can sometimes behave malignantly and metastasize especially recurrent tumors.[5]

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].[1,5,11,13,15] 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.[10] Meningeal melanocytoma cells share the same embryonic origin of nevus of Ota being derived from neural crest cells,[1] 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.

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Conflicts of interest

There are no conflicts of interest.

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