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CASE REPORT

Nevus of Ota associated with intracranial melanoma: Case report and review of the literature

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Abstract: There is a known association between nevus of Ota and melanomas involving the brain parenchyma and/or the meninges. We present the unusual case of a 32-year-old African-American female with a nevus of Ota and a contralateral parenchymal, primary CNS melanoma. We discuss the unique features of this case and provide a brief review of the literature regarding nevi of Ota and associated CNS melanoma. Our patient is a 32 year-old, African-American female with a left-sided nevus of Ota who presented with a three month history of headaches and paresthesias involving her left face and arm. An MRI of the brain revealed a hemorrhagic mass in the right temporal lobe, which, after craniotomy, was determined to be a melanoma. Extensive imaging, ophthamologic examination and full-body skin examination revealed no other foci of melanoma. To our knowlege, this is the only case of a nevus of Ota associated with contralateral parenchymal melanoma in an African-American patient. The association of contralateral parenchymal primary CNS melanoma with nevus of Ota is extremely unusual. Futhermore, despite the association of nevus of Ota with CNS melanoma, the literature does not support routine screening of patients with nevus of Ota for CNS melanoma with imaging modalities.

Keywords: nevus of Ota; melanoma; skin

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The patient is a 32-year-old African-American female with no significant past medical history who had severe headaches and episodes of numbness on the left side of her face and left arm for several months prior to presentation. Of note, the patient had a large nevus of Ota on the left side of her face that had been present since birth (**Figure 1**). She stated that the nevus had been unchanged for at least a decade. On exam, there was slate-blue patch which covered most of the left half of her face. Most of the left sclera displayed similar blue pigmentation as did a portion of her soft palate. The patient had no pigmented lesions concerning for melanoma on full body skin exam. The patient's neurological exam was normal, except for subtle sensory deficits on the left face and arm. An MRI of the brain revealed a 5 cm hemorrhagic mass located in the right superior temporal gyrus. A subsequent staging work-up including CT scans of the chest, abdomen, and pelvis failed to reveal any other foci of melanoma, so it was concluded that the melanoma originated intracranially. The patient underwent craniotomy for extirpation of this lesion, and histopathological examination revealed it to be a melanoma. The patient did well initially; however, the melanoma recurred with leptomeningeal dissemination several months after surgery, and the patient expired.

Discussion

We present the case of an intracranial melanoma in a woman with unilateral nevus of Ota. Our case is unique in that the melanoma is contralateral to the patient's nevus of Ota and located in the brain parenchyma.

Nevus of Ota, often referred to as oculodermal melanocytosis, usually occurs as a flat or slightly raised blue-black or slate-gray unilateral discoloration in the distribution of the first and second divisions of the trigeminal nerve[1]. It is formed by melanoblasts that fail to migrate to the dermoepidermal junction and instead remain in the dermis[2].

Melanoblasts are the precursors of melanocytes, and

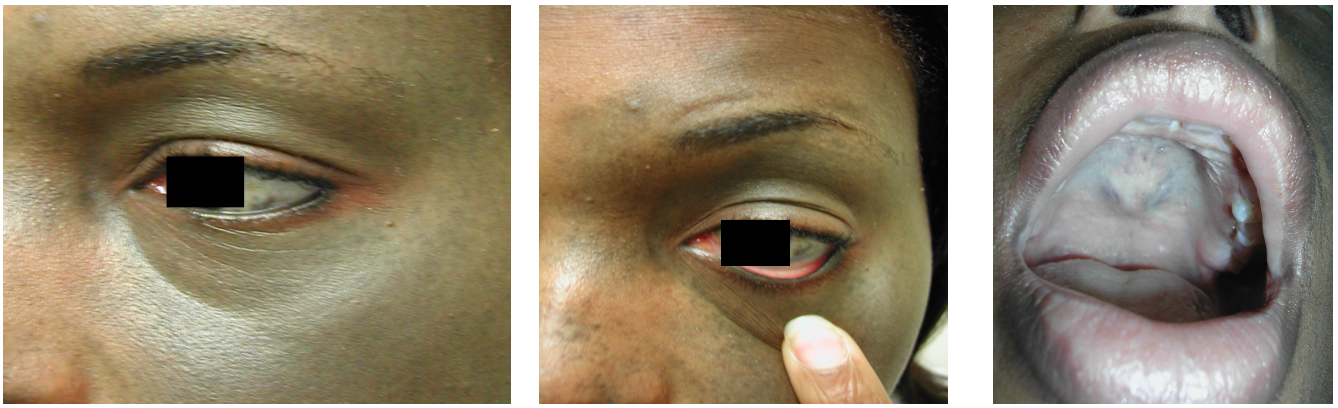

Figure 1. A large nevus of Ota on the left side of the patient's face

their arrested migration can cause pigmentary abnormalities in the nerve fibers, meninges, ocular structures, and inner ear[2].

Intracranial melanomas originate from the proliferation of melanocytic elements normally present in the leptomeninges (i.e. melanoblasts of neural crest origin) which can develop neoplastic patterns[3]. Leptomeningeal melanosis, a disproportionate number of melanotic cells in the meninges, has been considered a hamartomatous state characterized by excessive formation of melanoblasts that may predispose an individual to CNS melanomas[4]. Likewise, dural melanomas are thought to originate from abnormal dural melanin pigmentation[5].

There are 15 previously reported cases of intracranial melanomas associated with nevus of Ota[5–11]. These tumors have been located in the cerebral hemisphere, the optic chiasm, and the pineal gland[5–11]. The majority of CNS tumors associated with nevus of Ota have been leptomeningeal[6]. In nearly half of reviewed meningeal melanomas associated with Ota's nevus, a dural attachment and origin have been documented[5].

To our knowledge, our patient is the fifth reported case of intraparenchymal CNS melanoma associated with nevus of Ota[6,7]. In the prior four cases, the leptomeninx had associated pigmentation or melanoma. With the recurrence of intracranial melanoma in our patient, there was leptomeningeal dissemination. However, it is not known if she had leptomeningeal pigmentation associated with the melanoma at initial presentation. While previously reported cases have been associated with pigmentation of the leptomeninx, it has been demonstrated by magnetic resonance that intraparenchymal melanin deposition can occur independently of detectable leptomeningeal melanosis[12,13]. Therefore, it may be possible that our patient presented with intraparenchymal melanoma without associated leptomeningeal melanosis.

To our knowledge, our patient is the third reported case of contralateral CNS melanoma associated with nevus of Ota[6,8]. Each of the two cases previously reported, Sang *et al*. in 1977 and Balmaceda *et al*. in 1993, were thought to have been a unilateral nevus of Ota associated with bilateral diffuse leptomeningeal melanocytosis, one area of which underwent malignant degeneration[6,8].

Although the prognosis of intracranial melanoma is poor, the prognosis of solitary intracranial melanoma or dural melanoma is better than that of diffuse leptomeningeal melanoma, likely because the former lesions are more accessible to direct surgical removal[14,15]. However, the survival of patients with solitary intracranial melanomas depends largely on the extent of surgical treatment and the location of the tumor[3]. In a study of 81 patients who underwent surgery for solitary intracranial melanomas, the overall survival rate was more than 12 months in 20% of cases (16/81) and less than 1 month in 13.6% (11/81) of cases[3]. Disease free intervals for dural melanoma have been reported as 16–18 months postoperatively[15]. Patients with leptomeningeal melanoma have a median symptom-free course of 5 months and a median postoperative survival of 1 month[16].

Given the poor prognosis of intracranial melanoma, the question arises as to whether or not patients with nevus of Ota should undergo screening CT or MRI for intracranial masses. To the best of our knowledge, there is not literature in strong support of implementing such screening measures. Our review of the literature revealed that our case represents only the sixteenth case of intracranial melanoma associated with nevus of Ota. In light of the rarity of the association of the two, it might be more prudent to advise clinicians to have a high index of suspicion in a patient with a nevus of Ota presenting with neurologic findings rather than to suggest widespread head imaging of asymptomatic patients with a nevus of Ota.

Conflict of interests

The authors declare no potential conflict of interest with respect to the research, authorship, and/or publication of this article.

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