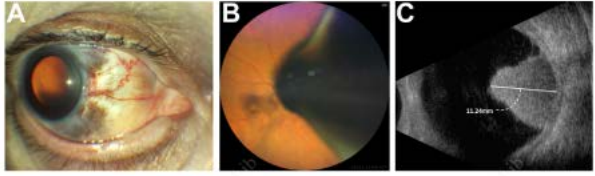

Ocular Melanocytosis with Uveal Melanoma

A 70-year-old man presented with decreased vision in the left eye. The examination revealed diffuse blue changes to the left sclera, patches of hyperpigmentation, and large conjunctival vessels in the nasal quadrant (**A**). Additionally, a pigmented intraocular mass abutting the lens (**B**) was seen nasally. Ultrasound biomicroscopy and B-scan ultrasonography (**C**) confirmed a ciliary body mass. The blue scleral pigmentation was present since birth; he had no other skin findings. A diagnosis of congenital ocular melanocytosis with a ciliary body melanoma was made. The patient underwent enucleation, and histopathologic evaluation confirmed the melanoma and gene expression profiling demonstrated Class II tumor characteristics (Magnified version of Figure **A-C** is available online at www.aaojournal.org).

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Definition: Oculodermal melanosis (Nevus of Ota, melanosis oculi), also known as oculodermal melanocytosis, oculomucodermal melanocytosis, or congenital melanosis bulbi, is a benign mesodermal melanosis involving the distributions of the ophthalmic and maxillary trigeminal nerve with associated hyperpigmentation of the eye and its adnexa. Entrapment of melanocytes in the upper third of the dermis leads to gray-blue macular hyperpigmentation of the conjunctiva and sclera and ipsilateral facial skin, usually occurring unilaterally. Scleral involvement occurs in greater than two-thirds of cases and is associated with an increased risk of developing glaucoma. Rarely, there can also be intraoral involvement with similar pigmented lesions seen on the palate.

Etiology/Prevalence

Nevus of Ota typically presents at birth but can also appear in puberty or during pregnancy. Nevus of Ota presents more commonly in females than males, with a 5:1 ratio. It also occurs predominantly in people of Asian and African descent. Although white individuals are least likely to develop Nevus of Ota, they are the most likely to develop malignant melanoma associated with this condition.

Risk factors

There are no known risk factors for Nevus of Ota. It is most commonly seen in the Japanese population but can rarely present in the Indian and White populations. Patients with Nevus of Ota are at risk for glaucoma and melanoma. Those with Nevus of Ota extending into the eye have an increased risk of developing glaucoma (10% of patients) as invasion of melanocytes can block drainage of aqueous, leading to elevated intraocular pressures. Malignant cutaneous or ocular melanoma may develop in these patients. 1/400 patients can develop uveal (typically choroidal) melanoma in the affected eye, and risk factors for malignant transformation include: related cutaneous or palatal melanocytosis (especially in the temple); scleral involvement of the superior, nasal, or temporal quadrants; choroidal melanocytosis; and diffuse iris melanocytosis .

Ocular melanosis is a condition differentiated from Nevus of Ota by the absence of eyelid involvement.

Nevus of Ito is a condition differentiated by involvement of hyperpigmentation of the neck, shoulders, axilla, and upper extremity.

Nevus of Hori is a condition almost identical to Nevus of Ota, but this condition is bilateral on presentation, as opposed to the unilateral Nevus of Ota.

Prognosis

Nevus of Ota is typically benign with an excellent ophthalmic and dermatologic prognosis with or without treatment. Yearly screening for glaucoma and malignant melanoma by an ophthalmologist and dermatologist is recommended. However Shields et al found that patients with uveal melanoma associated with oculodermal melanocytosis have a twice as likely risk for metastasis compared to those without melanocytosis.