

International Journal of Medical Science and Innovative Research (IJMSIR)

IJMSIR : A Medical Publication Hub Available Online at: www.ijmsir.com Volume – 3, Issue –4, August - 2018, Page No. : 217 - 220

Oculodermal Melanocytosis- A Brief Communication

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Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Oculodermal Melanocytosis is also known as Nevus of Ota. It is a hamartomatous melanocytic nevus of dermal melanocytes. We report a case series of two female patients with naevus of ota. This article briefly communicates the etiology, pathogenesis and various treatment modalities available in the management of these patients.

Introduction

Oculodermal melanocytosis or Nevus of Ota or nevus fuscoceruleus ophthalmomaxillaris was described by Ota in 1939.In Nevus of Ota there is blue hyperpigmentation of the face along the distribution of ophthalmic and maxillary branches of trigeminal nerve.^[11] This condition is most prevalent in Japanese population but comparatively rare among Indians. This condition is more common in females with female to male ratio of 5:1^[2] The typical lesion is unilateral though bilateral involvement may occur.^[3] It can be congenital or acquired.

Case Report

CASE 1-We report a case of 15 years old female with complaints of bluish discoloration of left sclera since early childhood. The discoloration gradually progressed to involve left upper and lower eyelids. The lesion was initially bluish in colour and gradually became darker over time. There was no history of redness, photophobia, watering, floaters, flashes of light, trauma,ocular medication, eve rubbing allergies. Family, or drug, personal, and socioeconomic history were not significant. General physical examination and systemic examination of patient was normal. On ocular examination multiple bluish grey patches were seen over the left upper and lower eyelid and sclera.(Figure 1,2) Visual acuity in right eye and left was 6/6.Pupillary reaction was normal in both eyes and ocular movements were also normal.On anterior segment examination of left eye episcleral pigmentation was present and right eye was normal. Intraocular pressure was 13mmof hg in right eye and 16mmof hg in left eye.Posterior segment examination was normal in both eyes.

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Figure1-Bluish discoloration of left upper and lower eyelid.



Figure2- Bluish discoloration of left sclera CASE2- Another female patient presented with complaints of bluish discoloration of right sclera and right side of face.There was no history of redness,photophobia, watering, floaters, flashes of light, trauma,ocular

medication. rubbing or allergies. Family, eye drug, personal, and socioeconomic history were not significant.General physical examination and systemic examination of patient was normal. On ocular examination multiple bluish grey patches were seen over the left upper and lower eyelid sclera left side of nose and face. (Figure 3) Visual acuity in right eye and left was 6/6. Pupillary reaction was normal in both eyes and ocular movements were also normal.Anterior segment examination of right eye shows episcleral pigmentation whereas left eye was normal.Intraocular pressure was 14mm of Hg in right eye and 15 mm of hg in left eye.Posterior segment examination was normal.



Figure3-Bluish discoloration of right side of face and sclera

Based on the distinct color, morphology, and location of the lesion, the condition was diagnosed as Nevus of Ota in both cases.Patients were counselled about the nature of disease, risk of long-term complications, and was advised regular follow-up.

Discussion

Nevus of Ota presents as a bluish hyperpigmentation along the ophthalmic and maxillary branches of the trigeminal nerve and occurs due to incomplete migration of melanocytes from the neural crest to the epidermis during the embryonic phase leading to dermal nesting and

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melanin production. Its occurrence in early infancy and in early adolescence suggests hormonal role in its development. The other etiological factors are genetics, hormones, infection, trauma, and ultraviolet light exposure.^[4] After its onset. Nevus of Ota may enlarge gradually and progressively and there may be increase in its pigmentation. It remains stable on reaching adulthood. The lesion's color depends upon the depth of involvement. The deeper lesions are blue due to Tyndall effect, in which, blue end of the light spectrum penetrates the deep dermis and is absorbed by dermal melanin^{[5].} In ocular melanocytosis, pigmentation occurs in conjunctiva, sclera, cornea, iris, choroid and less common in retrobulbar fat, orbit, periosteum, extraocular muscles and optic nerve. The ocular complications are glaucoma(10-15%) and uveal melanoma.^[6]

The other sites of involvement are oral cavity, nasal mucosa, external auditory canal, tympanic membrane, orbital fissures, meninges, and the brain^[7]. Differential diagnosis for nevus of Ota are mongolian spot, melasma, blue nevus, and drug-induced hyperpigmentation. Based on the extent of the involvement, Tanino classified nevus of Ota into four types:^[8]

Type I: IA	Mild orbital type	Distribution over the upper and lower eyelids.
Type I: IB	Mild zygomatic type	Involvement of zygomatic region.
Type I: IC	Mild forehead type	Involvement of the only forehead.
TypeI:ID		Involvement, of only ala nasi .
Туре II	Moderate type	Distribution over the upper and lower eyelids, periocular, zygomatic, cheek, and temple regions.
Type III	Severe type	The lesion involves the scalp, forehead, eyebrow, and nose.
Type IV	Bilateral type	Bilateral involvement

There is no definitive diagnosis for nevus of Ota. Diagnosis is mainly by history and clinical examination. In cases where malignant changes are suspected, skin biopsy is required .^{[9].}The various treatment modalities for nevus of Ota include cryotherapy, skin abrasion, microsurgery, cosmetic camouflage and laser. Recently Q-switched Nd: YAG and Alexandrite lasers have become a gold standard for the treatment of nevus of Ota^{.[10]}

Conclusion

Nevus of ota is a rare condition. Various topical and laser treatments have been used successfully in treating the cosmetic blemish associated with this condition.

It can affect vision if ocular complications like glaucoma and uveal melanoma occurs, therefore regular follow up is required.

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