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Case Report

# Bilateral nevus of Ota – A rare clinical presentation

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ABSTRACT

Nevus of Ota is a rare congenital hamartoma of the dermal melanocytes causing a blue-grey hyperpigmentation of the eye and adnexa. The condition is usually unilateral but rarely it can be bilateral. A 28-year-old lady from West Bengal, India, visited the Ophthalmology outpatient department with the complaints of blackish discoloration of left side of the face and periorbital region. On examination, a large (5x6 cm) lesion was noted on the skin of the left side of face, forehead, sclera, lower palpebral conjunctiva, ala of nose, hard palate along with sclera and iris of the right eye. Cutaneous examination revealed that the lesion is unilateral diffuse, homogenous, blue-gray colored macular pigmentation with ill-defined borders. The lesions do not undergo spontaneous regression, but the intensity of the blue-black discolored lesions may vary during menstruation, fatigue or change in weather. The patient needs to be followed up annually, checking at each visit for visual acuity, biomicroscopy, intraocular pressure, gonioscopy and fundus examination. A thorough knowledge of the disease process, possible complications and screening protocol can help the ophthalmologist to pick up sight threatening complications early, initiate prompt treatment which may avert possible visual loss.



## 1. Introduction

Hyperpigmentation is a common condition where the skin appears dark. Nevi are usually benign and scant attention is paid to them. However some nevi are significant, especially for the Ophthalmologist. This case is reported as it is a rare presentation of Nevus of Ota – also known as Oculo-muco-dermal melanocytosis or congenital melanosis bulbi. It is an uncommon congenital hamartoma of the dermal melanocytes causing a blue-grey hyperpigmentation of the eye and adnexa.[1] This condition was first reported by Ota and Tanino in 1939 as patchy, unilateral, bluish black discoloration of skin innervated by the ophthalmic and maxillary divisions of the trigeminal nerve.[2] Though this condition is included under oculocutaneous syndromes, it is not a true phacomatoses because of lack of involvement of the CNS.[3] Half of the cases present with a history of pigmented lesions since birth and the rest present in early adulthood or during puberty. This entity is usually unilateral with only 10% of cases being bilateral.[4] The ophthalmic complications of this disease include glaucoma, possibly due to direct invasion of melanocytes into the uveal tissue leading to blockage of the aqueous drainage, or malignant melanoma of the choroid and uvea.

## 2. Case Report

A 28-year-old lady from West Bengal, India, visited the Ophthalmology outpatient department with the complaints of blackish discoloration of skin around both the eyes. The discoloration was present around the left eye since birth, and now had gradually increased in size and started involving the other eye also. She also gave history of bluish discoloration of the left side of her face, forehead, and inside

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of mouth since birth. The discoloration was not associated with pain, itching, photosensitivity or any sudden increase in size. There were no complaints of diminution of vision or past history suggestive of glaucoma. Family history was non contributory; her parents were non consanguineous, patient denied taking any long term medication which are susceptible to develop hyperpigmentation such as topical Prostaglandin analogues, long term topical steroids, Oral contraceptive pills, hormone replacement therapy, tetracycline like minocycline, anticonvulsants like valproic acid, psychoactive agents like tricyclic antidepressants or phenothiazines, Oral antihypertensive agents like Angiotensin II antagonist, Calcium channel blockers or betablockers, anti cancer agents.

On examination, a large (5x6 cm) blue-grey colored lesion was noted on the skin of the left side of face, forehead, sclera, lower palpebral conjunctiva, ala of nose and hard palate. No other pigmentation or abnormalities were noted (Figure 1). Cutaneous examination revealed that the lesion was unilateral diffuse, homogenous, blue-grey colored macular pigmentation with ill-defined borders (Figure 2). Intraoral examination revealed similar lesions over the hard palate and buccal mucosa (Figure 3).

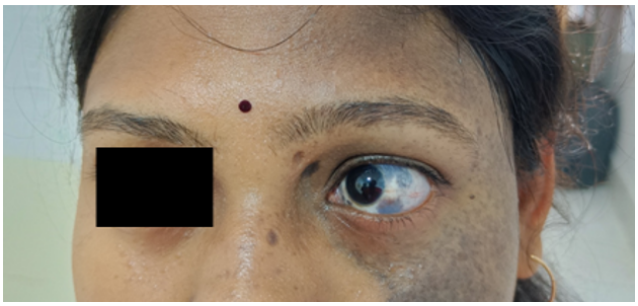

**Fig. 1:** Hyperpigmentation of skin over the left side of face and periocular skin

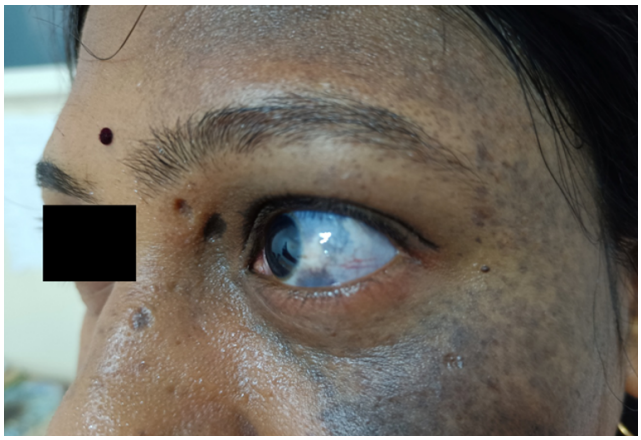

**Fig. 2:** Hyperpigmentation involving sclera and eyelids of left eye

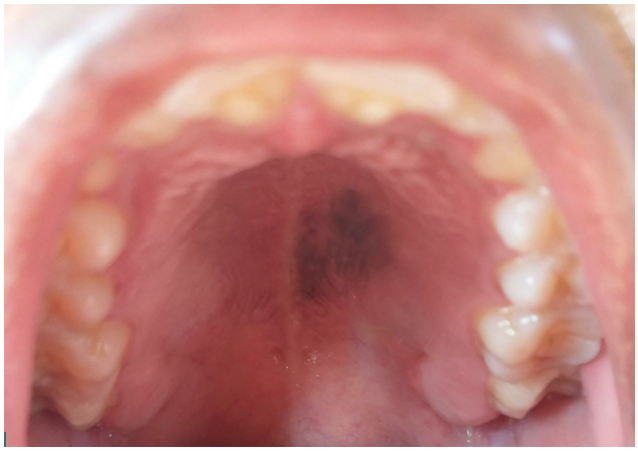

**Fig. 3:** Hyperpigmentation over the hard palate

On ocular examination, BCVA both eyes was 6/6 N6. There was dark brown hyperpigmentation in the temporal half of the iris and superior sclera in the right eye (Figure 4). The left eye also showed extensive hyperpigmentation of lids and upper, temporal and nasal part of sclera and iris (Figure 5). Both heterochromia iridis and iridum were noted. Intraocular pressure by Goldmann Applanation Tonometer was 14 mmHg and 16 mmHg in right and left eye respectively. The rest of the anterior segment was normal with no evidence of cataract or glaucoma. Gonioscopy was normal in both eyes and no hyperpigmentation of the trabecular meshwork was noted. Fundus examination of both eyes was unremarkable; no hyper pigmentation was noted in the choroidal vasculature, optic disc or macula. Baseline OCT was normal.

Based on the above findings, a clinical diagnosis of bilateral nevus of Ota was made. The patient was advised an annual eye examination, to watch for any sudden increase in size of lesion, diminution of vision or eye pain.

## 3. Discussion

Our patient had brownish black discoloration of the left side of face and periocular region since birth with involvement of iris and sclera of both eyes which is in favor of the diagnosis of bilateral nevus of Ota, which is a congenital hamartoma of dermal melanocytes in the distribution of the trigeminal nerve, most commonly limited to the maxillary and ophthalmic divisions. The condition is usually unilateral 90% but rarely bilateral 10%, thus depicting the rarity of this case.

Our patient had involvement of the eyelid thus differentiating it from ocular melanosis. The patient had no involvement of regions supplied by posterior supraclavicular and lateral cutaneous brachial nerves, thus differentiating it from nevus of Ito. Another condition mimicking this case is nevus of Hori, which is an acquired

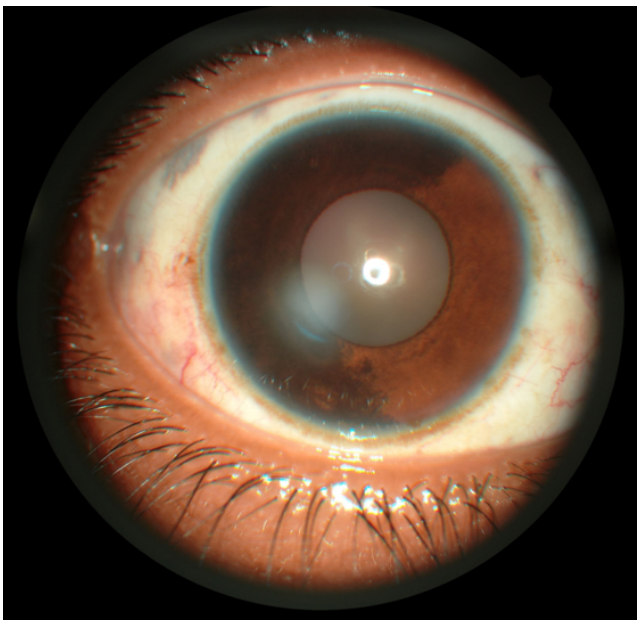

**Fig. 4:** Right eye showing hyperpigmentation of one half of iris (Heterochromia iridum)

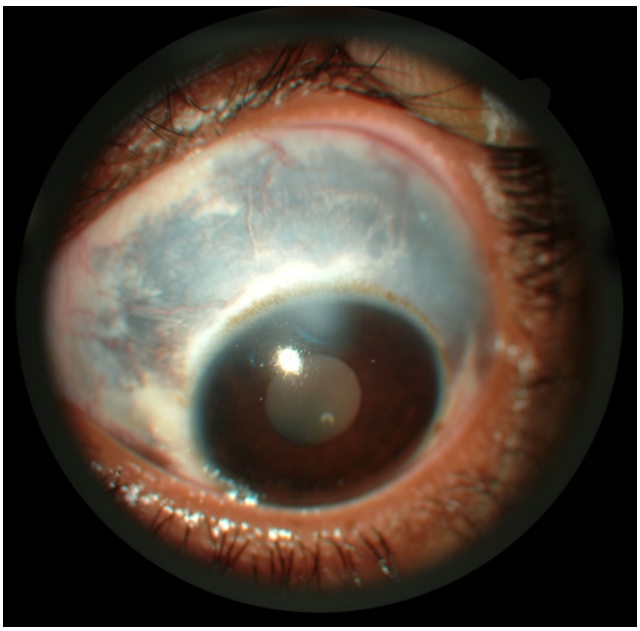

**Fig. 5:** Hyperpigmentation involving the superior part of sclera of left eye

condition with bilateral facial bluish grey macules without ocular or mucosal involvement.

Rare cases of sensory hearing loss have also been reported, in association with nevus of Ota but this was not present in our case.[5] The lesions do not undergo spontaneous regression, but the intensity of the blue-black discoloration of lesions may vary during menstruation, fatigue or change in weather.[6]

**Table 1:** Mishima classification of nevus of Ota[7]

| Subtypes | Intensity | Pigmentation | Areas involved |
|---|---|---|---|
| Type I | Mild | Light brown | Upper and lower eyelids and zygomatic area |
| Type II | Moderate | Deep slate Gray | Eyelids, zygomatic area, and base of nose |
| Type III | Intensive | Deep blue to brown | Affecting the first and second division of trigeminal neuralgia |

Biopsy often reveals the invasion of dendritic melanocytes in the dermis, leading to dermal melanosis. The genetic analysis of invaded dermal melanocytes mimics the genetic aberrations associated with either dysplastic nevi or melanoma in a significant number of cases. Possible molecular aberrations associated with risk of malignant transformation includes mutations of GNAQ and BAP1 genes.[8]

At present there is no evidence of glaucoma in our patient and there were no signs of malignant transformation. The patient needs to be followed up annually, checking at each visit for visual acuity, slit lamp biomicroscopy, intraocular pressure, gonioscopy and fundus examination. Biopsy may be required if there is sudden increase in the size of lesion as it is often an indicator of malignant transformation.

## 4. Conclusion

Bilateral nevus of Ota is a rare entity as the condition is usually unilateral. Both Ophthalmologists and Dermatologists need to be aware of these varied presentations as complications like glaucoma may result in irreversible blindness. A thorough knowledge of the disease process, possible complications and screening protocol can help the ophthalmologist to pick up sight threatening complications early, initiate prompt treatment which may avert possible visual loss. Clinical diagnosis is based on color, site and presentation. It is usually present since birth, but may occur during puberty and pregnancy. Skin biopsies are mandated when there is suspicion of malignant transformation.[9] Patients are advised to note any sudden change is color and size of lesion and also for visual disturbances. Annual review is recommended to evaluate

for glaucoma and malignant transformation.

## 5. Source of Funding

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## 6. Conflict of Interest

None.

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