Nevus of Ota with Buccal Mucosal Pigmentation: A Rare Case

Shishir Ram Shetty¹, Subhas Babu G², Kumuda Arvind Rao³, Renita Castellino³

ABSTRACT
Nevus of Ota is a condition wherein the typical pattern of the bluish black pigmentation is noticed along with the cutaneous distribution of the trigeminal nerve. This condition is most prevalent in Japanese population but comparatively rare among Indians. We report a case of 23-year-old female presented with unilateral pigmented areas over the skin of forehead, malar area, ear and periocular area. Blackish-blue pigmented areas were also noticed on the sclera. Brownish-black diffuse pigmented areas were also noticed on the buccal mucosa of the same side. The presence of pigmentation on the skin over pinna and oral pigmentation made our case a rare incidence. Oral pigmentation associated with nevus of Ota especially on the buccal mucosa have rarely been reported in the past.

Keywords: Buccal mucosa, Oculodermal melanocytosis, Oral pigmentation, Nevus of ota.

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Introduction
Nevus of Ota or ‘Oculodermal melanocytosis’ is a congenital hyperpigmentation of the facial skin in the distribution of the first and second divisions of the trigeminal nerve which is often associated with increased pigmentation of the various ocular structures.¹ Pigmentation may affect sclera, conjunctiva, cornea, iris, choroid and less commonly, the optic nerve, retro bulbar fat, orbit periosteum and extra ocular muscles.² The dermal lesions are bluish, confluent, non hairy, flat, pigmented macules with poorly defined margins. The melanocytosis also affects the oral cavity, nasal mucosa, external auditory canal, tympanic membrane, orbital fissures, meninges and the brain.³

Case Report
A 23-year-old female patient reported to our department with decayed tooth in the lower jaw of six-month duration. The patient’s medical history was non-contributory. The patient had no deleterious habits. Extra-oral examination revealed the presence of hyperpigmented areas over the left periocular and malar areas extending to the midline. Superiorly, it extended to the hairline on the same side, inferiorly to an imaginary line joining corner of the mouth to left ear tragus (Figure 1A). Bluish-blackish discoloration of the sclera and pinna were noticed on the same side (Figures 1B and 1C). No other pigmentation or abnormalities were noticed during the general physical examination. Patient stated that these pigmented areas were present since birth and had gradually increased in size. The patient had never sought any treatment for it in the past. Brownish-black diffuse pigmentation was noticed on the buccal mucosa of the same side extending anteriorly from the angle of the mouth to the retro-molar area, and superio-inferiorly to the depth of the buccal sulcus (Figure 1D). Buccal mucosa on the right side showed no pigmentation. A thorough examination of the patient was done by a qualified dermatologist, otolaryngologist and ophthalmologist. Visual acuity and intraocular pressure were normal. Otoscopy and central nervous system examination did not reveal any abnormality. Endocral work-up and CT scan of the head revealed no abnormalities. After getting opinions from the concerned specialists, the diagnosis of Nevus of Ota was made. The other possible differential diagnoses were Sturge-Weber syn-

¹ Assistant Professor, Department of Oral Medicine and Radiology, A.B Shetty Memorial Institute of Dental Sciences, Mangalore, India.
² Professor and Head of the Department, A.B Shetty Memorial Institute of Dental Sciences, Mangalore, India.
³ Postgraduate Student, A.B Shetty Memorial Institute of Dental Sciences, Mangalore, India.
Correspondence to: Shishir Ram Shetty, Email: drshishirshettyomr@yahoo.com
drome, café-au-lait spots of neurofibromatosis, blue nevus, melasma and actinic lentigo. Sturge-Weber syndrome presents with unilateral Portwine stains and intraocular changes appearing similar to Nevus of Ota. Lesions of Sturge Weber syndrome have typical reddish-purple colour and regress as age progresses in contrast to Nevus of Ota which has a blackish colour and advances as age progresses. Besides, majority of the cases of Sturge Weber syndrome have a history of neurological disorder like epilepsy. A CT scan of the head was performed to rule out cerebral angiomatosis which is also a characteristic feature of Sturge-Weber syndrome. Neurofibromatosis was ruled out because of the lack of cutaneous nodules and the freckling in the axillary region. Blue nevus could be easily ruled out because of its typical bluish color. Melasma can be ruled out as it occurs during pregnancy and hormonal changes. Actinic lentigo presents with brownish papules or plaques. No ocular pigmentation is seen in any of the above discussed entities. The patient was then referred to dermatologist for laser therapy but the patient did not agree for the treatment. However, the patient has been on a regular follow-up for the past one and half years.

Figure 1. (A) Cutaneous pigmentation of periorbital and malar region on the left side of the face, (B) Pigmentation of lower and lateral part of left eye sclera, (C) Cutaneous pigmentation over the pinna of the left ear, (D) Left buccal mucosa showing diffuse areas of pigmentation.
Discussion
Hulke first described Nevus of Ota in 1860 but a detailed description was made first by Ota in 1939 wherein the typical pattern of the bluish black pigmentation along with the cutaneous distribution of the trigeminal nerve were described. This condition is most prevalent in Japanese population but comparatively rare among Indians. However, the exact prevalence rate in Indian population still remains unknown. This condition is more uncommon in males with female to male ratio of 5:1.

The exact etiology of this condition is unknown. Some researchers believe that melanocytes move from the neural crest to the skin during early embryonic life. Failure of complete migration into the epidermis before birth with ensuing dermal nesting produces characteristic blue epidermis before birth with ensuing dermal nesting bryonic life. Failure of complete migration into the epidermis before birth with ensuing dermal nesting

from the lesions have been reported, usually along with the ophthalmic division of the trigeminal nerve. Different disorders such as Sturge-Weber syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, multiple haemangiomas, spinocerebellar degeneration, ipsilateral deafness, and congenital cataract have been associated with Nevus of Ota.

Our patient was asymptomatic when she first reported to us and continued to be so since the past one and half years. Our case report intended to highlight a rare oral presentation of a relatively rare dermatological condition.

References