

## Case Report

### **Oculodermal melanocytosis: A case series**

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**Abstract:** Nevus of Ota is a hamartoma of dermal melanocytes. The cause for it unknown, but certain theories have been postulated wherein it has been suggested that it results from failure of migration of melanocytes from the neural crest to dermoepidermal junction. It is also hypothesized that these aberrant dermal melanocytes may be influenced hormonally. Clinically, it depicts a characteristic pattern of the bluish black pigmentation along the cutaneous distribution of the trigeminal nerve. Literature review shows a very few substantial cases reported and incidence of such cases are very meagre in our Indian subcontinent. Prevalence of such cases is directed towards female population and its occurrence in oral cavity is quite rare. This is a case series of unilateral Nevus of Ota of a 28 year old male and 35 year old female patient with oral manifestations.

**Keywords:** Melanin, Nevus, Pigmentation

#### **INTRODUCTION**

Nevus of Ota (also known as "congenital melanosis bulbi", "nevus fuscoceruleus ophthalmomaxillaris", "oculodermal melanocytosis", and "Oculomucodermal melanocytosis") is hyperpigmentation of facial skin and mucous membranes in the course of distribution of the ophthalmic, maxillary and occasionally, the mandibular divisions of the trigeminal nerve. On the dermis this lesion appears as bluish black pigmented macules with ill defined margins [1]. The onset of nevi typically is either at birth (60%) or soon after birth, but few rare cases of acquired type have also been reported [2]. The involvement of oral mucosal membrane is extremely rare. This case series deals with a nevus of ota present in a twenty eight

year old male patient and thirty five year old female patient with intra-oral involvement.

#### **CASE REPORT-I**

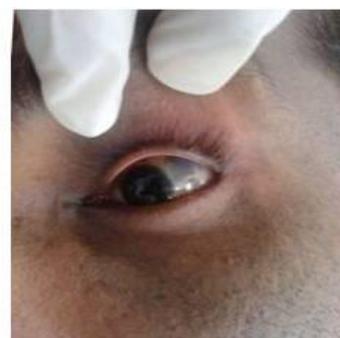
Twenty eight year old male patient came with a chief complaint of sensitivity with respect to his lower left back teeth since one month. There was no significant medical history reported. On examination there was an ill-defined bluish black pigmented region on the left side of his face extending superiorly from his hairline to inferiorly till the line joining the angle of the mouth (figure 1). Laterally it extended from the external auditory meatus to medially till the ala of nose involving the sclera of left eye (figure 2, 3)



**Figure 1**



**Figure 2**



**Figure 3**

Intraorally, brownish black pigmented region was evident on the hard palate bilaterally (figure 4). The buccal mucosa, tongue and floor of the mouth appeared normal. No other abnormalities were detected. On

specifically asking about the facial discoloration patient revealed that it was present since birth. A clinical diagnosis of nevus of Ota of left side of face with intra-oral palatal involvement was given.



**Figure 4**

#### **CASE REPORT-II**

Thirty five year old female patient came with a chief complaint of stains on all surfaces of teeth since two weeks. Personal history of the patient revealed a habit of pan chewing since ten years twice daily. There was no significant medical history reported. On extra oral examination there was an ill-defined bluish black

pigmented region on the right side of her face extending superiorly from her hairline to inferiorly about three centimetres away from the corner of the mouth (figure 5). Laterally it extended from the external auditory meatus to medially till the ala of nose involving the sclera of right eye (figure 6).



**Figure 5**



**Figure 6**

Intraorally, brownish black pigmented region was evident on the right side of the hard palate slightly crossing the midline( Figure 7). The buccal mucosa, tongue and floor of the mouth appeared normal. No other abnormalities were detected. On specifically

asking about the facial discoloration patient revealed that it was present since birth. A clinical diagnosis of nevus of Ota of right side of face with intra-oral palatal involvement was given.



Figure 7

## DISCUSSION

Nevus of ota is a dermal melanocytic disorder, whose exact etiology is still mysterious. However it has been suggested that it is failure of migration of melanocytes from neural crest cells to dermal epidermal junction during embryonic stages [3]. It is also been hypothesized that these dermal aberrations are influenced by certain hormones, through Hypothalamic-Pituitary-Ovarian axis mechanism.

Nevus of Ota mostly appears as a unilateral, macular, rarely papular or nodular, patchy brown, slate-blue or blue-black pigmentation, with deeper lesions appearing blue due to Tyndall effect of the dermal melanocytes [4]. Classification of the lesion is important to define the extent of the disease. Based on distribution, Tanino classified Nevus of Ota as mild, moderate, intensive and bilateral.

Based on the extent of the involvement, Tanino's classified Nevus of Ota as follows[5]:

Type I: IA - Mild orbital type: Upper and lower eyelids, periorbital, and temple region,

IB - Mild zygomatic type: Infrapalpebral fold, nasolabial fold, and zygomatic region

IC- Mild forehead type: Forehead alone,

ID-Alanasialone

Type II: Moderate type: Over the upper and lower eyelids, periocular, zygomatic, cheek, and temple regions.

Type III: The lesion involves the scalp, forehead, eyebrow, and nose.

Type IV: Bilateral type.

Both of the cases in this report appeared to be Type II variety of Tanino classification.

The major differential diagnoses for nevus of Ota are café- au- lait pigmentation of neurofibromatosis, hemangioma and Port-wine stain of

Sturge-Weber Syndrome. Café au lait pigmentation can present on the face but its uniform brownish flat appearance usually poses no difficulty in diagnosis [6].

From a dental practitioner's point of view, the condition that can also be easily confused with nevus of Ota because of lack of familiarity is Hemangioma. Hemangioma will typically not involve sclera as seen in Nevus of Ota. Another similar condition involving the same extent of area that can be confused with nevus of ota is Port wine stain (which maybe a part of Sturge-Weber syndrome), which can be differentiated from the color of appearance [7].

Therefore, the dentist should have awareness about nevus of Ota so that there are no unnecessary investigations like Ultrasonography or Angiography for the patient if he is suspected for Hemangioma [7].

These lesions are usually asymptomatic but rare instances of malignant melanoma arising from the lesions have been reported, usually along with the ophthalmic division of the trigeminal nerve [8].

The psychological impact of the lesions is high and early treatment would considerably reduce the stress later in life [2]. Dermabrasion, epidermal peeling, argon laser, Q-switched ruby, alexandrite and Nd: YAG laser have been used successfully for its treatment [7]. In our case the esthetic appearance was not a major concern for the patient hence no further treatment was undertaken.

## CONCLUSION

In this paper, two case reports of nevus of ota have been discussed. Nevus of ota with palatal involvement is a rare finding and should always be thoroughly examined by dentist and subsequently referred for dermatological and ophthalmological consultation, as malignant transformation and glaucoma has been documented. A thorough knowledge of nevus

of ota and its differential diagnosis is essential to avoid misdiagnosis.

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