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***CASE REPORT***

# A Rare Intraoral Manifestation of Nevus of Ota - A Case Report

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## ABSTRACT

Nevus of Ota is a dermal melanocytic nevus which is characterized by benign hamartomatous hyperpigmentation. Clinically it presents as a congenital or acquired blue or gray patch on the face and is distributed on the ophthalmic, maxillary, and mandibular branches of the trigeminal nerve. It is most commonly found in Japanese populations and is rare in the Indian subcontinent. It affects only 0.014 – 0.034% of the Asian population and is less common in the male population, with a male to female ratio of 1.4:8. The involvement of pigmentation over the pinna of the ear and the oral mucosa is extremely rare with very few cases reported in scientific literature. The case reported here is a case of nevus of Ota, with a rare intraoral presentation on the hard palate, crossing the midline.

**Keywords:** hyperpigmentation, melanocytic nevus, nevus of Ota

## INTRODUCTION

Nevus of Ota, also known as nevus fuscoceruleus-ophthal-momaxillaris or oculodermal-melanocytosis, is a macular discoloration of the face that is most commonly found in Japanese populations.[1] Hulke first described the nevus of Ota in 1860, however a more detailed description was made by Ota in 1939 wherein, the typical pattern of the bluish-black pigmentation along the cutaneous distribution of the trigeminal nerve was described.[2] It is a rare condition that only affects 0.014% – 0.034% of the Asian population.[1] Nevus of Ota presents unilaterally and consists of hyperpigmentation of the facial skin and mucous membranes in the distribution of the ophthalmic, maxillary, and occasionally the mandibular divisions of the trigeminal nerve.[3,1] The presentation of intraoral pigmentations are extremely rare, with very few cases documented in scientific literature.[4] This case report describes a case of nevus of Ota with a rare intraoral finding.

## CASE REPORT

A 45 year old male patient presented to the Department of Oral Medicine and Radiology, his chief complaint was a broken tooth in the right posterior region of his lower jaw for the past year. The patient reported no associated symptoms and his medical and drug history was noncontributory. The patient's dental history revealed restorations and multiple extractions of decayed teeth 2 years prior. On extraoral examination, a unilateral diffuse, irregular and macular blackish-grey pigmentation measuring approximately 4x4 cm was noted on the left temple area, malar area, periorbital area, root of the nose, ala of the nose, cheek region, pinna of left ear and the sclera [Figures 1 and 2]. The hyperpigmentation extended superiorly to the upper border of the left eyebrow, inferiorly 2 cm above the line joining the left angle of the mouth and tragus of the ear, anteriorly (medially) it extended to the root of the nose, and posteriorly (laterally) it was bounded by the left hair line.

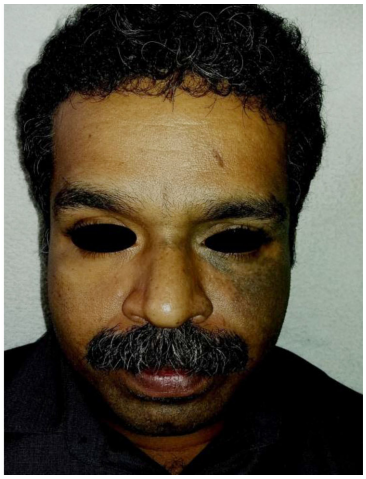

**Figure 1a**. Cinical presentation showing cutaneous pigmentation involving the left periorbital area, malar area, cheek area, and root and ala of the nose

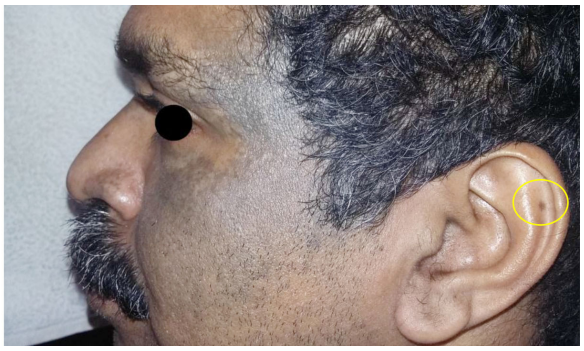

**Figure 1b.** Cinical presentation showing cutaneous pigmentation involving the left temple area, periorbital area, malar area, cheek area, pinna of the ear, and root and ala of the nose

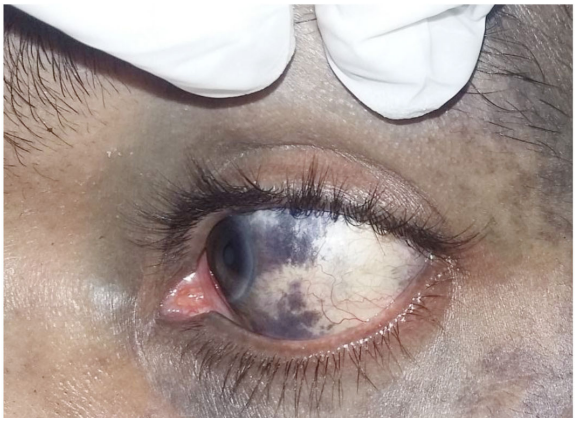

**Figure 2a.** Clinical presentation showing pigmentation of the left sclera on medial movement of the eye

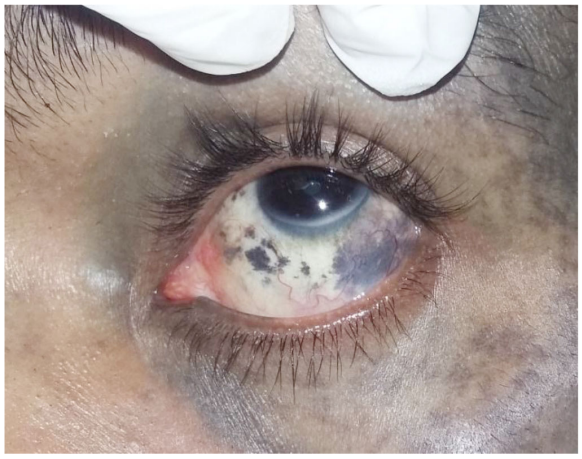

**Figure 2b.** Clinical presentation showing pigmentation of the left sclera on upward movement of the eye

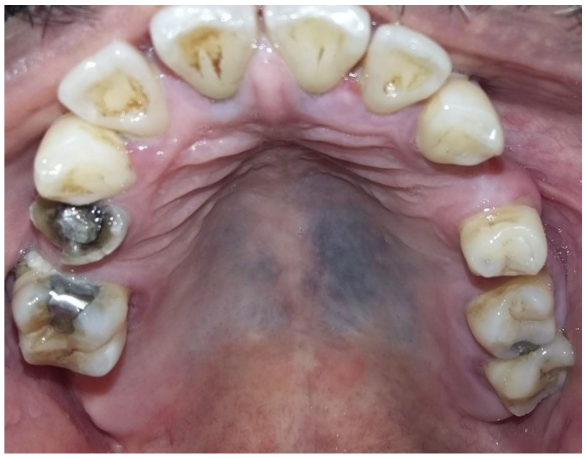

**Figure 3.** Clinical presentation showing intraoral pigmentation of the hard palate, crossing the midline

A general physical examination of the patient revealed no other areas of pigmentation on the body. On further questioning, the patient advised that the pigmentation had been present since birth. The patient reported no difficulty with his vision. On intraoral examination, similar pigmented areas on the hard palate, that crossed the midline, were noted [Figure 3]. The dorsal surface of the tongue showed fissuring. There were no other pigmented areas noted in the oral cavity. A dental examination revealed multiple filled, missing and decayed teeth, root stumps and chronic generalized gingivitis. Based on the disclosed history and clinical examination, a provisional diagnosis of nevus of Ota was given for the discoloration of the face. The differential diagnoses considered were blue nevus, actinic lentigo, Sturge-Weber syndrome, and café-au-lait spots of neurofibromatosis. A thorough examination of the patient was also completed by

**Table 1 -** Documented cases of intraoral nevus of Ota in chronologic order [1, 7, 11–14]

| Author | Location | Gender/ Age | Year |
|---|---|---|---|
| Dorsey And Montgomery | Buccal Mucosa | 16/M | 1954 |
| Mishima And Mevorah | Hard Palate (Along The Midline), Tongue | 35/F | 1961 |
| Mishima And Mevorah | Hard Palate (Unilateral) | 45/M | 1961 |
| Decosta And Carneiro | Buccal Mucosa | 23/M | 2011 |
| Reed And Sugarman | Hard Palate (Unilateral) | 43/F | 1974 |
| Yeschua | Buccal Mucosa | 27/F | 1975 |
| Page | Hard Palate | 59/F | 1985 |
| Rathi | Hard Palate | 30/F | 2002 |
| Karthik Kannan | Hard Palate | 32/F | 2003 |
| Parihar | Hard Palate (Unilateral) | 32/F | 2007 |
| Parihar | Hard Palate (Crossing Midline) | 33/M | 2007 |
| Mahima V | Hard Palate (Unilateral) | 36/F | 2011 |
| Gaurav Sharma | Hard Palate (Unilateral And Attempting To Cross Midline) | 22/M | 2011 |
| Shishir Ram Shetty | Buccal Mucosa | 23/F | 2011 |
| Peeyush Shivhare | Tongue, Gingiva And Hard Palate (Unilateral) | 20/F | 2012 |
| Jitendar Solanki | Soft Palate And Retromolar Trigone | 56/M | 2014 |
| Mengji Ashwini Kumar | Hardpalate (Along Midline) | 26/F | 2016 |
| Preethika G B | Hard Palate (Crossing Midline) | 28/M | 2016 |

a qualified dermatologist, ophthalmologist, and otolaryngologist who confirmed the diagnosis of nevus of Ota. The visual acuity and intraocular pressure were found to be normal and arcus senilis was present in both eyes, encircling the cornea. The otoscopy and central nervous system examination did not reveal any abnormalities. The endocrinal workup and CT scan of the cranium revealed no abnormalities. After obtaining expert opinions from the concerned specialties, a complete diagnosis of nevus of Ota was made. A recommendation was made to extract the root stumps, including the 45, begin a course of oral prophylaxis, and replace the missing teeth. Laser treatment for the pigmentations on the face was also suggested, however the patient refused. The patient is now on a regular follow up list.

## DISCUSSION

Nevus of Ota is a clinical condition that presents as a congenital or acquired bluish or grey patch on the face distributed along the branches of the trigeminal nerve.[3,5] Hulke first described nevus of Ota in 1860, however a more detailed description was made by Ota in 1939.[6,7] Considered a benign hamartomatous pigment, the nevus of Ota usually appears at birth but can also occur during puberty or pregnancy.[5,8] The condition is more prevalent in Japanese populations and is rare in the greater Asian population with a ratio of 0.014% – 0.034%. [1] It is less common in males when compared with females with the ratio of 1.4:8.[9] This case report documents a male patient who reports the nevus was present at birth. The exact aetiology of this condition is unknown, however some researchers have theorised that failure of complete migration of neural crest cells into the epidermis before birth, with ensuing dermal nesting and melanin production causes the characteristic greyish-blue patches.[7] Other factors such as hormonal alterations and exposure to ultraviolet radiation may play an important role in the pathogenesis of this condition.[7] Around 90% of cases of nevus of Ota are unilateral, however in 5 to 10% of cases pigmentation can be bilateral.[1] The above case had a unilateral presentation extraorally and bilateral presentation intraorally.

Areas that usually affected by pigmentation are the lower part of the forehead, eyes, temples, nose, zygomatic, and malar areas, and to some extent the ears and scalp. When it involves the ocular area it presents as hyperpigmentation of the sclera, cornea, retina, and iris, and may be associated with glaucoma. In rare cases the hard palate and tympanic membranes can also be involved.[10] In the case presented here, the discoloration involved the left temple region, malar area, cheek area, pinna of the ear, periorbital area, and intraoral involvement of the palate. The presence of pigmentation on the pinna of the ear and palatal mucosa, crossing the midline, makes this case particularly unique and rare. Shetty SR et al. also reported a case of nevus of Ota on the pinna of the ear and documented cases of intraoral nevus of Ota are outlined mentioned in Table 1.[7]

**Tanino's Classification**
Tanino's classification categorises pigmentation according to its distribution. The classifcation system has 4 major categories dependant on the area of involvement.
Type I – Mild
Type Ia – Eye region
Type Ib – Zygomatic region
Type Ic – Forehead region
Type Id – Nostril region
Type II - Moderate
The lesions affect the upper and lower eyelids, periocular, zygomatic, cheek, and temple regions.
Type III - Severe
The condition is distributed over the scalp, forehead, eyebrows, and nose.
Type IV- Bilateral type

The case reported in this article would be categorised as Type II according to Tanino's Classification, as the areas of involvement include the temple, malar, periocular, eyelid, and cheek areas. The presence of pigmentation on the pinna of the ear and the palatal mucosa, crossing the midline, makes this case both unique and rare as only a few cases have been documented.[7] If there is intraoral involvement it is most commonly present on the palate, followed by the buccal mucosa.[11] A skin biopsy is usually only required if any further changes occur in the pigmented areas, such as ulcerations or changes in colour.[15] The patient was offered a biopsy of the pigmented area to rule out any malignancy, however the patient refused as he had not experienced any problems associated with it. The final diagnosis of nevus of Ota was based on a thorough history and its striking clinical appearance, which eliminated a possible misdiagnosis. This diagnosis was also confirmed by a qualified ophthalmologist and dermatologist based on its clinical features.

There are various treatment modalities available for nevus of Ota involving the skin, including dermabrasion, epidermal peeling, and argon lasers. Nevus of Ota has also been successfully treated using Q-switched ruby, alexandrite, and Nd: YAG lasers.[1] This patient was offered laser treatment to reduce the pigmentation on his face, however he refused due to financial reasons.

Documented complications associated with nevus of Ota are glaucoma of the eye and malignant melanomas arising from the skin..[11] Most reported cases of malignant melanoma are usually from cutaneous lesions of nevus of Ota. The less commonly reported cases are those arising from the meninges and ocular areas. The Author's could find no reported cases of malignant melanomas in the oral cavity arising from a nevus of Ota.[11] One reported case, by Fistarol et al., concluded that the intraoral plaque type blue nevus may be confused with an intraoral malignant melanoma as they have very similar clinical appearances.[16] Even though there are no reported cases of melanoma arising from intraoral nevus of Ota, the risk is still very real and cannot be ignored because of malignancies that arise from the cutaneous nevus of Ota. [11] As such, long-term follow up is required for patients and the subject of this case report is being monitored at regular follow up appointments.

## CONCLUSION

A rare case of nevus of Ota, with palatal mucosal involvement that crosses the midline has been reported. Oral physicians should have a thorough knowledge of this rare dermatological entity for better patient management and prevention of any complications associated with it.

## REFERENCES


1. A Nevus of Ota with intraoral involvement: a rare case report. Int J Oral Health Med Res. 2015;2(2):76-9.
2. Alvarez-Cuesta CC, Raya-Aguado C, Vazquez-Lopez F, Garcia PB, Perez-Oliva N. Nevus of Ota associated with ipsilateral deafness. J Am Acad Dermatol. 2002;47(5):S257–9.
3. Sekar S, Kuruvila M, Pai HS. Nevus of Ota: A series of 15 cases. Indian J Dermatol Venereol Leprol. 2008;74:125-8.
4. Rathi SK. Bilateral nevus of ota with oral mucosal involvement. Indian J Dermatol Venereol Leprol. 2002;68(2):104.
5. Saracoglu S. Case report: Treatment of Nevus of Ota with 1064 nm Q-Switched Nd:YAG. J Laser Health Acad. 2013;2:42-5.
6. Ota M, Tanino H. Naevusfuscocaeruleusophthalmo-maxillaris and melanosisbulbi. Tokyo Iji Shinshi 1939;63(3133):1243–5.
7. Shetty SR, Babu S, Rao K, Castelino R. Nevus of Ota with buccal mucosal pigmentation: a rare case. Dental Res J. 2011; 8(1): 52-5.
8. Wang HW, Liu YH, Zhang GK, Jin HZ, Zuo YG, Jiang GT, Wang JB. Analysis of 602 Chinese cases of nevus of Ota and the treatment results treated by Q- switched alexandrite laser. Dermatol Surg. 2007;33(4):455-60.
9. Hidano A, Kajima H, Ikeda S, Mizutani H, Miyasato H, et al. Natural history of Nevus of Ota. Arch Dermatol. 1967;95:187–95.
10. Sharma S, Jha AK, Mallik SK. Role of Q-switched ND:YAG laser in nevus of Ota: A study of 25 cases. Indian J Dermatol. 2011;56:663-5.

11. Sharma G, Nagpal A. Nevus of Ota with rare palatal involvement: a case report with emphasis on differential diagnosis. Case Rep Dent. 2011;11:670-9.
12. Solanki J, Gupta S, Sharma N, Singh M, Bhateja S. Nevus of Ota - a rare pigmentation disorder with intraoral findings. J Clin Diagn Res. 2014;8(8):49-50. J Clin Diagn Res. 2014; 8(8): 49-50
13. Kumar MA, Venkatesh E, Srikanth D, Prashanth P.The case of a rare nevus with palatal, facial, and scleral pigmentation. Univ Res J Dent. 2013;3:131-3.
14. Oculodermal melanocytosis:A case series. Preethika GB, John A, Kashyap RR, Shetty D. Sch J Dent Sci. 2016;3(1):8-11.
15. Maheshwari R, Desai V, Sunil Kumar MV, Gaurav I. Unilateral nevus of Ota: A case report of oculodermal melanocytosis. J Dent Allied Sci. 2016;5:39-42.
16. Fistarol SK, Itin PH. Plaque-type blue nevus of the oral cavity. Dermatology. 2005;211:224-33.

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