A Nevus of OTA with Intraoral Involvement: A Rare Case Report

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ABSTRACT

Nevus of Ota, which originally was described by Ota and Tanino in 1939. It is characterized as congenital or acquired hamartoma of dermal melanocytes, presents clinically as a blue or gray patch on the face within the distribution of the ophthalmic and maxillary branches of the fifth cranial (trigeminal) nerve. Involvement of the palatal mucosa occurs rarely in nevus of Ota, when it occurs, it usually blends with the oral mucosa and is typically irregular, ill defined and often present as a mottled patch. Nevus of Ota is rare in the Indian subcontinent. So far very less cases of nevus of ota with intraoral involvement have been documented in the English literature. We report a rare case of intraoral nevus of Ota in a 20 year-old female patient.

KEYWORDS: Nevus of Ota, Melanoma, Hamartoma, Glaucoma

INTRODUCTION

The nevus of Ota (nevus fusoceruleus ophthalmomaxillaris” or oculodermal melanocytosis) is a macular discoloration of the face, found most commonly in the Japanese people.¹ Nevus of ota develops when the melanocyte get entrapped in the upper third of the dermis. It involves the branches (ophthalmic and maxillary) of the trigeminal nerve and present unilaterally on the face. It is a condition that occurs rarely & affects only 0.014% – 0.034% of the population of Asia. The involution of oral mucosal membrane is astronomically recherche.² Nevus of Ota can be associated with sundry ocular abnormalities such as congenital glaucoma and melanoma. Malignant alterations can also occur in this nevus with the appearance of melanoma affecting the skin, orbit, iris, ciliary body, choroid and brain.³⁴⁵ Till date very less cases of nevus of ota with intraoral involution have been documented in the literature. Here, we report a rare case of nevus of Ota of the oral cavity in a 20 year-old female patient.

CASE REPORT

A 20-year-old female came to our department for regular check up. There was no significant medical history reported. Extraorally, patient had asymptomatic, well-demarcated bluish black macule on the right midface involving temporal, frontal, zygoma and the maxillary sinus area not crossing midline (figure 1, 2). Bluish black pigmentation of right sclera was also observed (figure 3). It was present since birth, and family history was non-contributory. There was no relevant drug history.

How to cite this article:
the hard palate, which was extended to the midline (figure 6). Other parts of the oral mucosa were normal. The patient was referred to dermatology for consultation, which did not report any abnormality other than the discoloration of the face.

Auditory examination also did not reveal any discoloration of the auricle and tympanic membrane. Ophthalmic examination does not reveal any abnormalities. The patient was not willing for any kind of treatment regarding the pigmentation. The patient was advised to report for regular follow up regarding nevus of Ota. The patient was subsequently referred for oral prophylaxis.

**DISCUSSION**

Nevus of Ota (nevus fuscoceruleus ophthalmaxillaris) is characterized by benign hamartomatous hyperpigmentation, clinically presented as a congenital or acquired blue or gray patch on the face, distributed on the ophthalmic and maxillary branches of the trigeminal nerve. Nevus of Ota is most commonly found in Asians affecting around 0.5% of the population. Very often, nevus of Ota occurs unilaterally in blacks and oriental women (4.8:1). Though most cases of nevus of Ota are unilateral (90%), in 5 to 10% of cases pigmentation can
occur bilaterally and may involve ocular, nasal and oral mucosal surfaces. Although malignant melanomas of the intraocular and central nervous system are more common in patients with nevus of Ota, the prevalence is still less than 4%. The nevus is caused due to incomplete migration of melanocytes from the neural crest to the epidermis during the embryonic phase. Consequently, the melanocytes enter the ophthalmic and maxillary branches of the trigeminal nerve creating spots on the nervous regions. Normally, the nevus of Ota appears at birth but can also occur in puberty or during pregnancy. 

Hirayama T. proposed a histological classification of the nevus of Ota into five types based on the locations of the dermal melanocytes. Superficial (type S), superficial dominant (type SD), diffuse (type D), deep dominant (type DD) and deep (type De). Nevi of Ota were originally classified based on clinical descriptions of pigmentation: type I (small lesions), type II (moderately sized lesions), type III (extensive lesions), and type IV (bilateral lesions).

Recently, a new classification of nevus of Ota based on the response to laser treatment has been determined. This new classification allows for the prediction of the clinical outcome of laser treatment.

There may be fluctuation in the color of the nevus of Ota according to personal and environmental conditions, such as fatigue, menstruation, insomnia, and cloudy, cold, or hot weather conditions. It is usually unilateral and is located in the areas innervated by the first and second branches of the 5th Cranial nerve. The lesion’s color depends upon the depth of involvement and the rate of the individual. The deeper lesions appear blue in color due to the Tyndall effect, whereas the more superficial lesions are slate gray in color.

The clinical differential diagnosis of skin lesions of nevus of Ota includes melanoma, Mongolian spot, blue nevus, and irregular melasma. Melasma presents clinically large diffuse blue-gray patches that tend to spontaneously resolve by age 3–6 years and typically tends to occur in lumbosacral area and rare in the face. Melasma has typical association with pregnancy and clinically appears as brown-to-gray brown patches with well-to-poor demarcation and irregular outline. It is bilateral and shows no palpate involvement. Another condition is a hemangioma, which will spare sclera as seen in the nevus of Ota. The oral melanotic macule can be misdiagnosed as nevus of Ota, but can be distinguished from nevus of Ota on the basis of size, as it is comparatively smaller, and with no involvement of the sclera.

Drug induced hyperpigmentation is usually acquired after ingestion of drugs like minocycline, amiodarone and gold. Nevus of Ito, described in 1954 by Minor Ito, as a dermal melanocytic condition of the shoulder. Nevus of Ota and Ito occur together, sometimes in the same patient, but Nevus of Ito is much less common.

The treatment options for nevus of Ota were limited before the introduction of laser in the clinical dermatology. The treatment options for nevus of Ota include cryotherapy, skin abrasion, microsurgery, cosmetic camouflage and laser. In recent years Q-switched Nd: YAG and Alexandrite lasers have become a gold standard for the treatment of nevus of Ota. Nevi of Ota have the potential to undergo melanomatous change. Malignant alterations can occur in the nevus of Ota with the melanoma affecting the skin, eye, ciliary body, choroid and brain. Malignant degeneration has occurred in 4.6% of reported cases and was more frequent in light skinned patients. The average age of the patient is about 60 years, though there is no known association between the age of the nevus and development of melanoma. Another well-known complication associated with oculodermal melanocytosis is glaucoma in the ipsilateral eye, which has been described in approximately 10% of patients. Glaucoma may be detected at birth or at a later age, which is often open angle, and is asymptomatic with loss of visual acuity, gradually. The pathology behind glaucoma in these patients is most likely deposition of melanocytes within the trabecular mesh-work at the iridocorneal angle.

There is no definitive diagnosis for this nevus. It may be clinically diagnosed, although, for confirmation, biopsy is indicated in patients of any age when the diagnosis is uncertain or in rapidly expanding or nodular lesions suggestive of malignancy. Periodic examination should be conducted regularly (twice a year) for early diagnosis of glaucoma or the more rare complication of malignancy.

CONCLUSION

Nevus of Ota with intraoral involvement is a rare entity. Oral health care providers should have a thorough knowledge regarding this entity, as it can lead to complications afterward, like glaucoma and melanoma if not diagnosed early and properly. Hence, the diagnosed cases of Nevus of Ota must be properly followed up and referral to a dermatologist and ophthalmologist early.

REFERENCES

Shivare P. et al.: Nevus of OTA - a rare entity


Source of Support: Nil
Conflict of Interest: Nil