

Case Report

Nevus of Ota with palatal involvement: a case report

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Abstract – Nevus of Ota is a macular discoloration of the face which is most commonly found in the Japanese and females are more commonly affected than males. It is first described by Ota and Tanino in 1939, involves the skin along the distribution of first and second division of trigeminal nerve. Involvement of the palatal mucosa occurs rarely in nevus of Ota, it usually blends with the oral mucosa and is typically irregular, undefined and often present as a mottled patch. We describe here the case of nevus of Ota with palatal involvement in a 13-year-old Asiatic boy. His past medical history revealed the presence of the lesion since birth. There were no other pigmented lesion locations. The exact etiology of nevus of Ota is still unknown but they have the potential to undergo melanomatous change.

Introduction

Nevus of Ota or oculodermal melanocytosis is a macular discoloration of the face which is most commonly found in the Japanese but non-oriental persons may also be affected [1]. Nevus of Ota, first described by Ota and Tanino in 1939, involves the skin along the distribution of first and second division of trigeminal nerve [2].

Nevus of Ota 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 [3].

It is said to be most prevalent in Japan where the incidence among the dermatology outpatients lies between 0.2% and 1%. Most of the cases reported are in females and oral cavity is infrequently involved [4,5].

There is an increased risk of uveal melanoma and glaucoma in these cases [6]. Malignant alterations can also occur in the nevus with the appearance of melanoma affecting the skin, orbit, iris, ciliary body, choroid and brain [3].

Oral involvement of the nevus of Ota is very rare, we report here the case of nevus of Ota with only palatal involvement.

Case report

A 13-year-old Asiatic boy was referred by his dentist to the maxillofacial department for evaluation of an asymptomatic palatal pigmentation. His past medical history revealed the

presence of the lesion since birth. There were no other pigmented lesion locations on the body. He did not take any regular medications. He didn't smoke or consume alcohol.

Extraorally, no pigmentation was observed.

Intraorally, an asymmetric, inhomogeneous with irregular edges diffuse hyperpigmentation affecting hard palate was noted. It was a blue-gray pigmented lesion 2 cm in diameter, flat and smooth in the midpalatal region (Fig. 1a).

The buccal mucosa, tongue and floor of mouth were normal.

Patient was asymptomatic with no effect on vision, sensory changes and hearing.

The patient was treated by total excisional with 5 mm margins under general anesthesia and placement of a protective palatal plate (Fig. 2).

The report stated that histological features were in keeping with a nevus of Ota.

The histologic analysis revealed a densely cellular dermal proliferation of fusiform cells, sometimes pigmented. The cells did not show cytonuclear atypia (Fig. 3).

The Ki-67 proliferative index was low. Mitoses were not detected. No other investigations were required.

The patient will be followed in the department at one week, three weeks, three months, then every six months after surgery. Complete healing of the palate was observed (Fig. 1b).

Discussion

The exact etiology of nevus of Ota is still unknown. However, nevus of Ota may represent melanocytes that have persisted and not migrated completely from the neural crest to

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Fig. 1. (a) Intraoral photography of Nevus of Ota affecting palatal mucosa. This showed an asymmetrical blue-gray lesion, approximately 2 cm in diameter affecting the hard palate (b) Healing after 2 months of surgery.



Fig. 2. Macroscopic view of lesion removed from the hard palate measuring 2 cm with 5 mm safety margins.

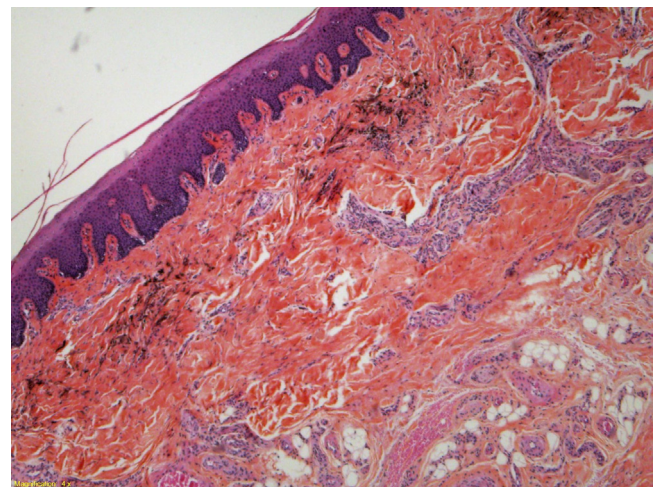


Fig. 3. Histological analysis of the lesion. Hematoxylin-eosin-saffron staining. The lesion corresponds to a dense cellular dermal proliferation, consisting of fusiform cells sometimes pigmented. The cells do not exhibit cytonuclear atypia. Mitosis patterns are not observed. (4x magnification).

the epidermis during the embryonic stage. Other theories which have been postulated is the active production by intradermal melanocytes [3].

The Asian population is more commonly involved affecting 0.014% to 0.034% of the population and females are more commonly affected than males at a ratio of 5:1 [6].

Normally, the nevus of Ota appears at birth but can also occur in puberty or during pregnancy. 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. The lesion's color depends upon the depth of involvement and the race 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.

Clinically, nevus of Ota presents as a brown, blue or gray patch on the face, which is congenital or acquired and is within the distribution of ophthalmic and maxillary branches of the trigeminal nerve. It can be unilateral or bilateral, but unilateral involvement is common (90–95%).

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% [3].

Nevus of Ota is asymptomatic though rare cases of sensory loss have been reported.

Extracutaneous sites like eyelids, and their adjacent skin areas, sclera, and conjunctiva, have also been involved. Spontaneous regression does not occur, although the intensity of the pigmentation may vary in relation to menstruation, fatigue or weather [2].

Table I. Classification of Nevus of Ota (Tanino's classification).

Types	Description
Type I	IA: Mild orbital type – Distribution over the upper and lower eyelids, periocular and temple region. IB: Mild zygomatic type – Infrapalpebral fold, nasolabial fold and zygomatic regions are affected. IC: Mild forehead type – Only forehead is affected. ID: Ala nasi alone is affected.
Type II	Moderate type – The lesions affect upper and lower eyelids, periocular, zygomatic, cheek and temple regions.
Type III	The condition is distributed over the scalp, forehead, eyebrows, and nose.
Type IV	Bilateral type.

Nevus of Ota often occurs in association with nevus of Ito which is a dermal melanocytic condition affecting the shoulder area. It can also be associated with other cutaneous disorders and ocular disease. Benign cutaneous and leptomeningeal conditions associated with nevus of Ota are phakomatosis pigmentovasculari, nevus flammeus, Sturge-Weber syndrome, Takayasu disease, Klippel-Trenaunay syndrome and neurofibromatosis.

Clinical differential diagnosis for skin lesions of nevus of Ota includes Mongolian spot, melasma, blue nevus, and drug induced hyperpigmentation [2].

Drug induced hyperpigmentation is usually acquired after ingestion of drugs like minocycline, amiodarone and gold whereas blue nevus may occur anywhere on skin. Sclera and oral mucosa are not involved in acquired bilateral nevus of Ota like macules (ABNOM) or Sun's nevus.

Mishima classified nevus of Ota into three types, depending on the extent and distribution of pigmentation [4]. 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 Di), deep dominant (type DD) and deep (type De) [3,7].

At this point, there is no ideal classification system but the classification laid down by Tanino in 1939 has remained the most useful clinical classification and in this classification Nevus of Ota has been classified into four major subtypes (Tab. I).

Oral lesions, which are rare yet significant are not represented in the above classification, prompting the authors of a case report about a palatal expression of Nevus of Ota to propose a modification of this classification and to include oral mucosal manifestations in the IE subclass [8–10].

There is no definitive diagnosis for nevus of Ota. They are diagnosed mainly by clinical examination and history. Skin biopsies are required only if clinical changes are suspected of malignant transformation within the involved skin, ocular tissues or mucosal tissues. Involvement of palatal mucosa occurs infrequently in nevus of Ota [4,6].

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.

Nevi of Ota have the potential to undergo melanomatous change. Malignant alterations can occur like 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.

Another well-known complication associated with ocular-dermal melanocytosis is glaucoma in the ipsilateral eye, which has been described in approximately 10% of patients [3].

A literature review was performed regarding case reports of palatal involvement of nevus of Ota and is summarized in the following table (Tab. II).

The review of literature showed that palatine involvement only, as the clinical case reported, was very rare. Extraorally, patients are most often affected at midface involving temporal, frontal, zygoma and the maxillary sinus area. Bluish pigmentation of sclera was also observed.

The diagnosis was clinical and confirmed by histological examination in most cases.

Treatment included regular follow-up to avert potential malignant transformation.

However, the decision to remove the lesion completely for our patient was made because of the difficulty of follow-up, and the risk of malignant transformation.

Conclusion

Nevus of Ota with intraoral involvement in males is a rare entity. Dentists 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.

Proper follow up and early referral to dermatologist and ophthalmologist is important in diagnosed cases of nevus of Ota.

Patients with nevus of Ota and especially with palatal pigmentation should always be examined by dentists. Dentists should have a thorough knowledge of nevus of Ota and its differential diagnosis to avoid misdiagnosis and, hence, the risk of malignant transformation. Patients should be taught awareness of the potential of glaucoma and malignancy.

Excision without previous biopsy of the lesion was the therapy of choice for this particular case, since there is still some controversy in the literature regarding the biopsy of pigmented lesions with malignant potential.

Table II. Table of cases reports of Nevus of Ota involving the palate reported in the literature.

Author/Year	Gender/Age	Description of lesions	Diagnosis	Management
Shivhare P <i>et al.</i> 2015	F/20	Bluish black discoloration was seen on right side of the hard palate which was extended to the midline.	NA	The patient was not willing for any kind of treatment regarding the pigmentation. Regular follow-up
Maguire J <i>et al.</i> 2019	F/48	Mild pigmentation of the left hard palate.	An incisional biopsy of the left buccal mucosa was completed. No other investigations were required.	Active monitoring owing to the potential for malignant change.
Solanki J <i>et al.</i> 2014	M/56	Blackish discoloration approximately 2 cm in diameter with diffuse margins was evident on the left side of soft palate.	Clinical diagnosis of Nevus of Ota was given with the consensus of dermatologist.	Regular follow up.
Virendra N. Sehgal <i>et al.</i> 2015	M/34	Several blue-pigmented macules with ill-defined borders merging into the surrounding mucosa of the hard palate.	No evidence indicative of malignancy was found on biopsy.	NA
Sharma G <i>et al.</i> 2011	M/22	Bluish discoloration approximately 2 cm in diameter with diffuse margins was evident on the left side of hard palate, which was somewhat crossing the midline.	Clinical diagnosis of nevus of Ota was given with the consensus of dermatologist.	Patient was advised to report for regular follow-ups of Nevus of Ota.
Mukhopadhyay A 2013	M/24	Few bluish pigmented patches were found in the right side of the hard palate.	No abnormality.	NA
Tomov G <i>et al.</i> 2015	M/52	Poor-defined lesion with bluish-black appearance on the right side of the posterior part of the hard palate.	NA	Follow-up to avert potential malignant transformation.
Guledgud MV <i>et al.</i> 2011	F/36	Well-defined lesion with uniform bluish-black appearance on the right side of the posterior part of the hard palate bordering the midline.	A diagnosis of Nevus of Ota was established as evidenced by the clinical findings.	The need for regular follow-up has been impressed upon the patient owing to the lesion's malignant transformation potential.

NA: Not Available.

Conflict of interest

All authors declare that there are no financial and personal relationships with other people or organisations that could inappropriately influence their work.

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Ethical approval

Exception from ethical approval because the study was a case report.

Informed consent

The authors declare that informed consent have been obtained.

Author's contribution

Conception and design of the study, or acquisition of data: A. Derache, M. Brix Drafting the article of revising it critically for intellectual content: A. Derache, M. Brix, E. Simon Final

approval of the version to be submitted: A. Derache, M. Brix, E. Simon.

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