Rare Case Report on Nevus of Ota

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• Abstract

Nevus of Ota is a hamartoma of dermal melanocytes. Clinically, Nevus of Ota is manifested as blue or gray patch on the face; such condition is congenital or acquired and is within the distribution of branches of the trigeminal nerve. The nevus can be unilateral or bilateral. In addition to skin, it may involve ocular and oral mucosal surfaces. The case of an 18-year old female with unilateral bluish black macule on the right side of the face since birth is presented. She also had a bluish patch on the right shoulder at birth, which disappeared when she turned 10 years. The pathogenesis of Nevus is unknown, and effective treatment has been realized with pigment-specific lasers.

• Keywords : Nevus of Ota, hamartoma, dermal melanocytes, bluish patch

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Introduction

Originally described by Ota and Tanino in 1939, Nevus of Ota is a hamartoma of dermal melanocytes¹. Clinically, Nevus of Ota is manifested as blue or gray patch on the face; such condition is congenital or acquired and is within the distribution of the ophthalmic and maxillary branches of the trigeminal nerve².

The nevus can be unilateral or bilateral. In addition to
skin, it may involve ocular and oral mucosal surfaces. A dermal melanocytic condition affecting the shoulder area, Nevus of Ito was initially described by Minor Ito in 1954. Nevus of Ito often occurs in association with Nevus of Ota in the same patient but is much less common\(^5\). Nevus of Ota is a facial dermal melanocytic lesion that is common among Asian patients. This usually affects the forehead, temple, eyelid, nose, ear, and scalp. Pigmentation may also be found in the oral mucosa, sclera, or tympanic membrane\(^6\).

The difference between Nevus of Ota and Nevus of Ito lies in its manifestation site. Nevus of Ota is on the forehead and face and around the eye area; Nevus of Ito is on the shoulder and upper arm area. Although it has the same appearance as Nevus of Ota, Nevus of Hori is not present at birth; it often affects both sides of the face\(^5\).

Various therapies have been successfully used for Nevus of Ota. Cosmetic cover-up products can be used for camouflage. Cryosurgery and microsurgical treatments are not recommended since they can leave disfiguring scars. On the other hand, chemical bleaching agents can damage the epidermal melanocytes and trigger permanent hypopigmentation or depigmentation\(^5-6\). The combination of dermabrasion and carbon dioxide snow method has produced good results\(^5-7\).

### Case Report

An 18-year old female visited the Dept. of ODMR with chief complaints of a 15-day old pain on the lower right back region of the jaw. The clinical examination revealed deep occlusal caries on the lower right first molar. Her general examination revealed a unilateral, asymptomatic bluish black patch present on the right side of the forehead, temples, malar aspects, ala nasi, and sclera. The patient revealed a history of pigmentation since birth. The patch is irregular in shape and is non-scrapable; it is not raised above the skin surface, extending instead from the forehead to the corner of the mouth on the right side. The patient also has a history of similar bluish patch measuring about 10mm on the right shoulder at the time of birth, but it gradually disappeared when she turned 10 years.

The eye examination revealed bluish black, asymptomatic, pigmented spots present in the sclera on the right eye only. The iris and pupil were normal on both sides. Intraoral melanocytosis was present on the marginal, attached, and interdental gingiva of the upper & lower teeth on the labial & buccal aspect and right buccal & labial mucosa as well as in the mid-palatine region. The patch present in the mid-palatine region is asymptomatic, bluish black, non-scrapable, and irregular in shape, measuring approximately 3 \(\times\) 2 cm from the periapical region of the upper anterior teeth up to the first molar region. The patient has no history of paresthesia or anesthesia associated with patch.

The following lesions were considered in the differential diagnosis:

- Blue nevus, Mongolian spot, Nevus of Ota

Mongolian spot appears as uniformly bluish without a brownish spotted component and manifests itself at birth or soon after birth and tends to disappear with age, whereas Nevus of Ota persists instead of disappearing. The blue Nevus is a sharply circumscribed, round or oval, soft nodule with bluish-black or slate-blue color whose diameter measures only a few millimeters as a rule; it is not detected in the final diagnosis.

Considering the characteristic clinical features of the lesion and age of the patient, she was provisionally diagnosed with...
early-onset type of Nevus of Ota based on her history of having pigmentation present since birth. According to the Tanino’s classification, it is a Moderate Type II Nevus of Ota.

The patient was referred to the Dept of Dermatology for further evaluation, where she was diagnosed with Nevus of Ota and advised to apply Melacare cream in the evening & Sun block/Sunscreen lotion for topical application for one month, after which the patient would be recalled. The lesion did not respond to conservative treatment. The patient was advised to undergo Q-switched ND-YAG Laser treatment since the lesions were asymptomatic; she refused laser treatment for facial pigmentation, however.

Discussion

Before Ota’s definitive description in 1939 by Tanino, Nevus of Ota was first described by Halbe in 1869 as nevus fuscoaceruleus ophthalmomaxillaris. Ota’s Nevus most often appears in the perinatal period or around puberty. The pigmentation of Ota’s Nevus consists of flat, blue-black, or slate-gray macules intermingled with brown, small, flat spots. The intensity of pigmentation may be influenced by fatigue, menstruation, insomnia, and weather.

Based on the distribution and extent of pigmentation, Ota’s Nevus was classified into four types by Tanino:

Type IA. Mild Orbital Type: light brown or slate spotted pigmentation limited to the upper and lower eyelids

Type IB. Mild Zygomatic Type: discrete brownish spots limited to the zygomatic region

Type II. Moderate Type: deep slate to brownish purple, relatively densely spotted pigmentation on the eyelids, zygomatic region, and base of the nose

Type III. Intensive Type: deep blue to bluish purple, densely spotted or almost diffused pigmentation on the eyelids, zygomatic region, base of the nose and ala nasi, forehead, external ear, postauricular region, and anterior part of the scalp

Type IV. Bilateral Type. For all types, the pigmentation was observed not to extend beyond the nasolabial fold and not to include the mandibular region.

Nevus of Ota is said to be more prevalent in Japan where the incidence among dermatology outpatients is 0.2~0.8 %. This condition is comparatively rare in India & uncommon among males. In fact, the male to female ratio is 1 : 5.

Nevus of Ota is a distinctive, mesodermal pigmentation of the facial skin; it is commonly combined with pigmentation of the eye. Although usually congenital, it is capable of benign progression.

This nevus is caused by the proliferation of dermal melanocytes. It is not malignant, but neither does it heal spontaneously. Laser therapy is effective. Nevus of Ota is further classified into the early-onset type wherein pigmentation is present at birth and the color darkens as the patient grows and the later-onset type, which occurs after puberty. Both types occur most frequently among Asian women and tend not to disappear spontaneously. Melanocytes are dispersed in the dermis. Pigmentation is present in the epidermal basal cell layer.

It is also closely related to the Mongolian spot & the blue Nevus. The difference between Mongolian spot & Nevus of Ota is that the former manifests itself at birth or soon after birth and tends to disappear with age-a uniformly bluish component without brownish spots. In contrast, the latter is present at birth (about 60%) or around puberty (about 40%);
it persists and has a brownish-blue color with powder-blast burn appearance. The blue Nevus is a sharply circumscribed, round or oval, soft nodule with bluish-black or slate-blue color whose diameter measures only a few millimeters as a rule.

The most serious but unusual complication is the development of malignant melanoma in the pigmented areas. Different disorders such as Struge-Weber syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, multiple hemangioma, ipsilateral deafness, & congenital cataract have been associated with Nevus of Ota.

In recent years, the use of laser therapy has been very effective, giving new hope to patients with Nevus of Ota. Various lasers are used in the treatment of pigmentary disorders, with the best results for the treatment of this condition apparently obtained with Q-switched Nd-YAG, ruby, and alexandrite lasers. The long wavelengths penetrate deeply into the dermis, triggering the selective photothermal and photomechanical destruction of dermal melanocytes. After irradiation, the shape of dermal melanocytes and their melanosomes is changed. Melanosomes are fragmented into smaller pieces, and their cell membranes are disrupted; the nucleus is fragmented or destroyed. The destruction of dermal melanocytes can be achieved without collateral injury to the surrounding tissue.

There is no significant epidermal pigmentation among patients with Nevus of Ota. Melanosomes in epidermal melanocytes are different from dermal melanocytes; they are smaller and greater in number. After laser therapy, epidermal melanocytes are reversibly changed; light microscopy shows the expansion of extracellular space, swollen mitochondria, dilatation of endoplasmic reticulum, and vacuolated melanosomes. One year after treatment, the normal cell structure is completely restored.

**Conclusion**

The disfiguration of the face as a cosmetic problem can be the result of various lesions affecting the face. Nevus of Ota is a relatively uncommon dark discoloration occurring around the face, eye, and shoulders. The lesions are usually benign & asymptomatic, but lifelong follow-up is required since few cases of malignant melanoma have been reported in literature. Although various modalities have been successfully used for Nevus of Ota, laser is very effective, and recurrence is rare.


