

# “Nevus of Ota”: A Rare Oro-Facial Pigmentation- Short Review

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## Abstract

Nevus of Ota is a dermal melanocytic hamartoma of unknown cause and rare in Indian subcontinent, associated with ophthalmic, maxillary and mandibular divisions of trigeminal nerve. Oral involvement of nevus of ota is very rare and found mostly in females. This review highlights this obscure, but important oro-facial pigmentation.

**Keywords:** Nevus of ota; Oculodermal melanosis; Oro-facial pigmentation

## Introduction

Nevus of Ota is a macula-dermal discoloration occurring due to failure of melanocytes migration from the neural crest to derma-epidermal junction. More common in Asian and African population Hulk firstly described this condition as oculodermal melanosis in 1861 [1,2]. The Japanese dermatology professor Masao Ota, described “bluish-grey irregular hyper-pigmentation along the first and second divisions of the trigeminal nerve with frequent mucosal involvement as ‘Nevus fusco-caeruleus ophthalmomaxillaris and melanosis bulbi’ in 1939. In 1956, Fitzpatrick renamed the syndrome to oculodermal melanocytosis. Nevus of Ito, described by Minor Ito in 1954, is a dermal melanocytic condition affecting the shoulder area [3,4]. Worldwide prevalence of nevus of Ota is around 0.10-0.32%. Incidence rate in Asian population is around 0.04%-0.34%. It is more prevalent in female population with male to female ratio is 1.4:8 [5,6].

## Characteristics

Ophthalmic, Maxillary and Mandibular divisions of trigeminal Nerve are involved out of which maxillary is most common [7,8]. The color of the skin lesion is brown or blue, the diameter of the area is 1–10 cm or larger. The color or perception of the color of nevus of Ota may fluctuate according to personal and environmental conditions, such as fatigue, menstruation, insomnia, and cloudy, cold, or hot weather conditions [9,10]. The morphological appearance of nevus of Ota is classified as Homogenous (macular with equally distributed pigment density) Speckled (scattered spots of discoloration with similar or various pigment density) Mixed (speckled pattern on a background of homogenous macule) [11].

Patho-physiology is yet to be confirmed even if it has been postulated that Nevus of Ota and other dermal melanocytic disorders such as nevus of Ito, blue nevus and mongolion spots may represent melanocytes that have not migrated completely from neural crest to the epidermis during embryonic stage [12]. The two peak ages of onset in early infancy and in early adolescence suggest that hormones are a factor in the development of this condition [13].

## Tanino’s Classification

It was divided into 4 types on the basis of skin involvement area [14,15] (Figure 1).

Type I - Mild

Type Ia – Eye region type

Type Ib – Zygomatic region type.

Type Ic – Forehead type.

Type Id – Nostril type

Type II - Moderate.

Type III- Severe.

Type IV- Bilateral type.

Based on innervation area of the trigeminal nerve branches as below, named PUMCH classification [15] –

Type I: pigmentation macules involving one branch of the trigeminal nerve.

Type Ia: Ophthalmic nerve.

Type Ib: Maxillary nerve.

Type Ic: Mandibular nerve.

Type II: pigmentation macules involving two branches of the trigeminal nerve.

Type IIa: (ophthalmic and maxillary nerves; V1 + V2) of the trigeminal nerve;

Type IIb: (maxillary nerve and mandibular nerve; V2 + V3) of the second and third branch of trigeminal nerve.

Type III: (ophthalmic, maxillary, and mandibular nerves; V1 + V2 + V3) of the trigeminal nerve.

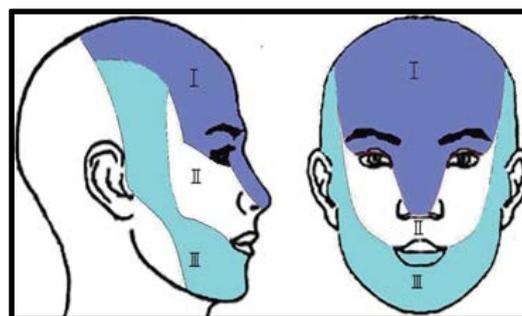


Figure 1: Showing ophthalmic, maxillary, mandibular divisions of trigeminal nerve affected by nevus pigmentation.

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Type IV (bilateral type): bilateral pigmentation macules respectively involve innervation areas of one or more branches of the trigeminal nerve.

Type IVa (symmetric).

Type IVb (asymmetric).

Type V: nevus of Ota with complications, such as Port-Wine Stains (PWS), telangiectasia, vitiligo.

### Clinical Features

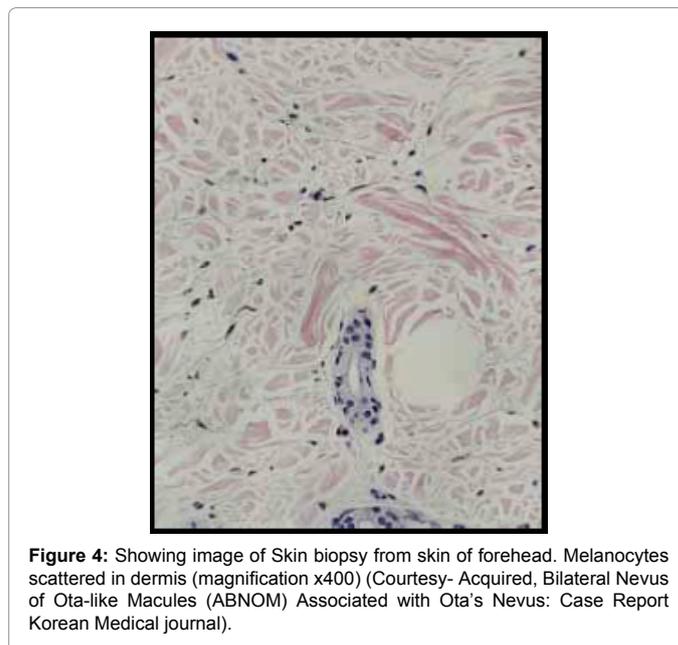
Mostly asymptomatic, can be associated with melanoma in rare cases (Figures 2 and 3). Ophthalmic division have ocular complications like glaucoma, melanoma associated with ciliary body, iris and optic nerve is also reported [16].

### Histopathologic Features

Skin biopsy reveals histopathologic view of dermal melanocytes, bipolar or oval in shape, scattered in the upper and middle portions of the dermis (Figure 4).

### Treatment

Topical therapy is of no value in the medical treatment of nevi of Ota. Previous treatment modalities, including cryotherapy, dermabrasion, and microsurgery, can be associated with scarring [17-20]. Development of the Q-Switched Nd: YAG laser (QSYL) and the Q-switched ruby laser (QSRL) has enabled complete, scar less

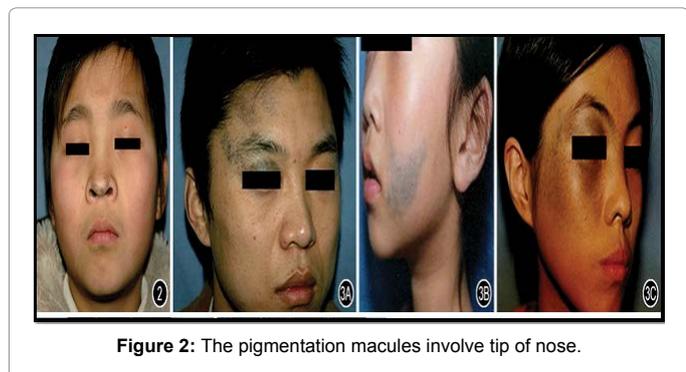


**Figure 4:** Showing image of Skin biopsy from skin of forehead. Melanocytes scattered in dermis (magnification x400) (Courtesy- Acquired, Bilateral Nevus of Ota-like Macules (ABNOM) Associated with Ota's Nevus: Case Report Korean Medical journal).

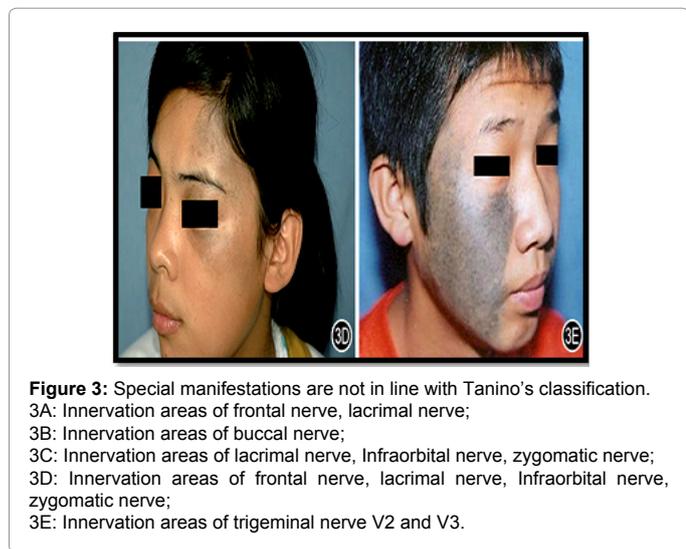
elimination of the pigmentation in patients with nevus of Ota. Without treatment, the skin lesions are permanent.

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**Figure 2:** The pigmentation macules involve tip of nose.



**Figure 3:** Special manifestations are not in line with Tanino's classification.  
 3A: Innervation areas of frontal nerve, lacrimal nerve;  
 3B: Innervation areas of buccal nerve;  
 3C: Innervation areas of lacrimal nerve, infraorbital nerve, zygomatic nerve;  
 3D: Innervation areas of frontal nerve, lacrimal nerve, infraorbital nerve, zygomatic nerve;  
 3E: Innervation areas of trigeminal nerve V2 and V3.

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