CASE REPORT

'Nevi of Ota: the unusual birthmarks': a case review

Ravi Prakash Sasankoti Mohan,¹ Sankalp Verma,² Amit Kumar Singh,² Udit Singh²¹OMDR, Kothiwal Dental College, Moradabad, Uttar Pradesh, India²Department of Oral Medicine and Radiology, Kothiwal Dental College and Research Center, Moradabad, Uttar Pradesh, India**Correspondence to**Dr Ravi Prakash Sasankoti Mohan,
sasan_ravi@rediffmail.com**SUMMARY**

Nevi of Ota are usually characterised by unilateral, mottled, slate blue or dark brown macules on the forehead and face around the eye area. These are unusual skin discolorations in which melanocytes are found deeper than normal. Ota's nevus is usually congenital but may appear in early childhood or in puberty. We summarise a case report along with histological and management aspects of this cosmetically unappealing condition.

BACKGROUND

Nevus of Ota which was first reported by Hulke in 1860 before Ota's description in 1939 is characterised by unilateral, patchy, irregular, grey discolouration of skin supplied by ophthalmic and maxillary divisions of trigeminal nerve, especially the periorbital region, temple, malar prominence, forehead and nose. It results from an abnormal migration of melanoblastic cells of the primitive neural tube along the first and second divisions of the trigeminal nerve during embryogenesis. It is a rare entity seen in Indians and is more commonly reported in female patients, may be because they are more cosmetically concerned. Although, cosmetics is a usual complaint but different complications may also occur.

CASE PRESENTATION

A 25-year-old female patient visited the outpatient department for routine dental checkup. On extra oral examination, unilateral, bluish grey-coloured macule was present on the right side of face extending from periorbital region to the malar region and to the temple on the side. The case history revealed that this pigmented lesion was asymptomatic and was present since birth. There was no history of alteration in its size and no similar discolorations were present elsewhere on the body. She had not taken any treatment for this and the family history was also non-contributory. Intraoral examination revealed involvement of buccal mucosa, a bluish coloured plaque was observed on the entire left buccal mucosa.

INVESTIGATIONS

- Biopsy

DIFFERENTIAL DIAGNOSIS

- Freckles
- Lentigines
- Melasma
- Facial Café-au-lait patch
- Spilus nevus
- Postinflammatory hyperpigmentation
- Pigmentary incontinence owing to various causes
- Riehl's melanosis

TREATMENT

Pathologically, nevus of Ota is a melanocytic hamartoma that resides at the level of the middle dermis; therefore, to be effective, treatment modalities should extend to this depth. Various treatment modalities have been tried, which are as follows:

- Cryotherapy (may result in scarring)
- Surgical excision
- Laser (may result in scarring)
- QS ruby (595 nm)
- QS Nd:YAG (1064 nm)
- QS alexandrite (755 nm)
- Carbon dioxide
- Argon
- Chemical peeling
- Dermabrasion⁴
- Topical hydroquinone-based bleaching agents

Fractional photothermolysis, or fractional resurfacing, is a rapidly evolving modality, which, at its core, relies on the production of microscopic arrays of thermal damage of controlled size and density. Because the treated zones are of such small size and because the intervening areas are spared the thermal effects of the laser, healing is quite rapid, with patients only experiencing minimal adverse effects of a transient erythema. As a result, much of the inflammatory response and associated potential adverse effects are minimised compared with other conventional ablative resurfacing methods.

The effective strategies to prevent postoperative hyperpigmentation probably is to avoid direct exposure to sunlight, routinely to wear sunscreen cream and to apply topical hydroquinone early, preferably starting a few days after treatment.¹

OUTCOME AND FOLLOW-UP

The most serious complications include glaucoma and transformation to malignant melanoma; concomitant deafness.

DISCUSSION

Nevus of Ota is also known as nevus fuscoceruleus ophthalmomaxillaris or oculodermal melanocytosis. It is a dermal melanocytic hamartoma that presents as bluish hyperpigmentation along the first or second branches of the trigeminal nerve.² An increased number of dermal melanocytes located in the distribution of the ophthalmic and maxillary divisions of the trigeminal nerve has been reported.³ Nevus of Ota affects between 0.014% and 0.034% of the Asian population.¹ The pathogenesis of this condition is unknown, but several theories have been put forward which include:

1. Dropping-off of epidermal melanocytes
2. Migration of hair bulb melanocytes

To cite: Mohan RPS, Verma S, Singh AK, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008648

3. Reactivation of pre-existing latent dermal melanocytes, which are triggered by dermal inflammation, UV radiation or hormonal changes during pregnancy.²⁻⁴

A specific genetic defect underlying this abnormal cell migration has not been identified until now; nevertheless, a familial case of nevus of Ota has been reported.⁵

Exogenous factors may alter the intensity of pigmentation. Emotional excitement, fatigue, insomnia, unusually warm or cold weather may increase the colour tone. Hormonal fluctuations associated with menstrual cycle and menopause can also cause darkening of the lesion.

Based on the extent and distribution, Tanino has classified nevi of Ota as:

- ▶ Type I: mild
- ▶ Type II: moderate (as seen in our patient's case)
- ▶ Type III: intensive
- ▶ Type IV: bilateral

Extracutaneous involvement has been reported, especially ocular involvement in 60% cases. Pigmentation of mucosal membranes of head and neck is variable and tympanic membrane is the most commonly affected. Nasal mucosa, palate, buccal mucosa, pharynx, tongue, iris, uveal tract and fundus are also involved but in a very less number of cases.⁶

Histopathologically, in nevus of Ota, pigment-bearing cells are numerous and distributed diffusely throughout the upper and lower dermis. These are elongated, slender pigment-bearing cells dispersed in between the collagen fibres of the dermis and the long axis of pigment-bearing cells, that is, melanocytes are along the collagen fibres.⁷

Different disorders like Sturge-Weber syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, multiple haemangioma, spinocerebellar degeneration, ipsilateral deafness and cataract have been associated with nevi of Ota.³

Learning points

- ▶ The clinical distinguishing feature between nevi of Ota and acquired bilateral nevi of Ota like macules is that, in the latter there is extraoral involvement sparing the oral mucosa.
- ▶ Nevi of Ota is usually unilateral present on the face whereas nevi of Ito is seen on the shoulders and upper arms.
- ▶ Disorders like Sturge Weber syndrome, Klippel Trenaunay syndrome, neurofibromatosis, haemangioma, spinocerebellar degeneration have sometimes been reported with nevi of Ota.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Hidano A, Kajima H, Ikeda S, *et al.* Natural history of nevus of Ota. *Arch Dermatol* 1967;95:187-95.
- 2 Murakami F, Baba T, Mizoguchi M. Ultraviolet-induced generalized acquired dermal melanocytosis with numerous melanophages. *Br J Dermatol* 2000;142:184-6.
- 3 Stanford DG, Georgouras KE. Dermal melanocytosis: a clinical spectrum. *Aust J Dermatol* 1996;37:19-25.
- 4 Hori Y, Takayama O. Circumscribed dermal melanoses: classification and histologic features. *Dermatol Clin* 1988;6:315-26.
- 5 Chan HH, King WW, Chan ES, *et al.* In vivo trial comparing patients tolerance of Q-switched alexandrite (QS Alex) and Q-switched neodymium: yttrium-aluminum-garnet (QSNd:YAG) lasers in the treatment of nevus of Ota. *Lasers Surg Med* 1999;24:24-8.
- 6 Xu X, Chu AY, Pasha TL, *et al.* Immunoprofile of MITF, tyrosinase, melan-A, and Mage-1 in HMB45-negative melanomas. *Am J Surg Pathol* 2002;26:82-87.
- 7 Hidano A. Acquired, bilateral nevus of Ota-like macules [letter]. *J Am Acad Dermatol* 1985;12:368-9.

Copyright 2013 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow