

Bilateral congenital nevus of Ota in association with Mongolian spot

Hoda Rahimi, MD¹
 Maryam Yousefi, MD¹
 Mina Mirnezami, MD²
 Zahra Asadi-Kani, MD¹

1. Skin Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
2. Dermatology Department, Arak University of Medical Sciences, Arak, Iran

Correspondence Author:

Hoda Rahimi,
 Skin Research Center, Shahid Beheshti
 Medical University, Shohada-e Tajrish
 Hospital, Tehran, Iran
 Haft-e –Tir Hospital, Lorestan
 University of Medical Sciences,
 Doroud, Iran
 e-mail: hoda_rahimi@yahoo.com

Conflict of interest: None to declare

Received: January 26, 2011

Accepted: April 23, 2011

A 24-year-old woman presented with asymptomatic hyperpigmented bilateral patches on her temples, eyelids and forehead since birth. Furthermore, the patient had a congenital grey patch, compatible with Mongolian spot, on her buttock. She had no vascular or other cutaneous lesion. Histopathologic examination revealed bipolar dendritic melanocytes dispersed in a ribbon-like pattern between the collagen fibers and around the neurovascular bundles of the dermis. As far as we know, this is the first case of bilateral “congenital” nevus of Ota in association with a Mongolian spot reported in a patient.

Keywords: nevus of Ota, congenital nevus, dermal melanosis, mongolian spot, bilateral nevus.

Iran J Dermatol 2011; 14: 68-70

INTRODUCTION

Nevus of Ota, a dermal melanocytic nevus first described in Japan ¹, manifests as blue - black or gray-brown patchy/diffuse pigmentation that usually occurs unilaterally in areas innervated by the first and second branches of the trigeminal nerve ². Bilateral involvement is an exception, especially if it is congenital, as in our case. Hereunder, we report an interesting case of association between bilateral “congenital” nevus of Ota and Mongolian spot.

CASE REPORT

A 24-year-old woman presented with asymptomatic hyperpigmented bilateral patches on her face since birth. The lesions progressed with age. She was born to a consanguineous

marriage. Her medical and family history were unremarkable. Physical examination revealed diffuse blue-grey hyperpigmented patches with irregular borders on her temples, eyelids and cheeks (Figure 1). Hyperpigmentation of the conjunctiva and sclera was also noticeable (Figure 2). Ophthalmologic examination was normal. No oral or nasal mucosal pigmentation was observed. Furthermore, the patient had a congenital grey patch, compatible with a Mongolian spot, on her buttock (Figure 3), which had become less prominent since birth. She had no vascular or other cutaneous lesions. Her general examination was normal. Histopathologic examination of the lesions revealed bipolar dendritic melanocytes dispersed in a ribbon-like pattern between the collagen fibers and around the neurovascular bundles of the dermis (Figure 4).



Figure 1. Hyperpigmented bilateral congenital patches on the face and sclera



Figure 2. Blue-grey hyperpigmentation of the sclera and face (closer view)

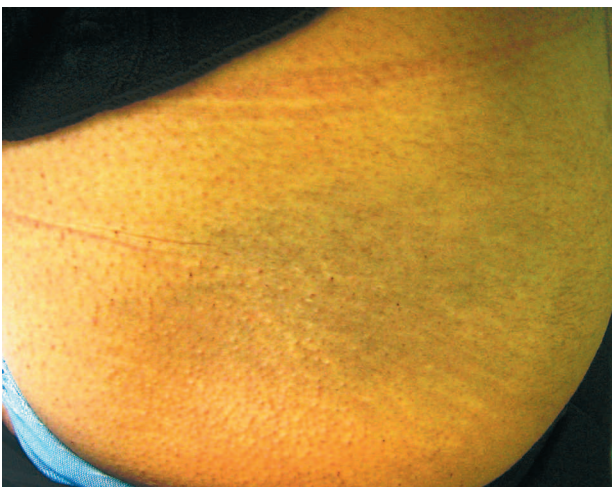


Figure 3. Mongolian spot on the left buttock of patient.

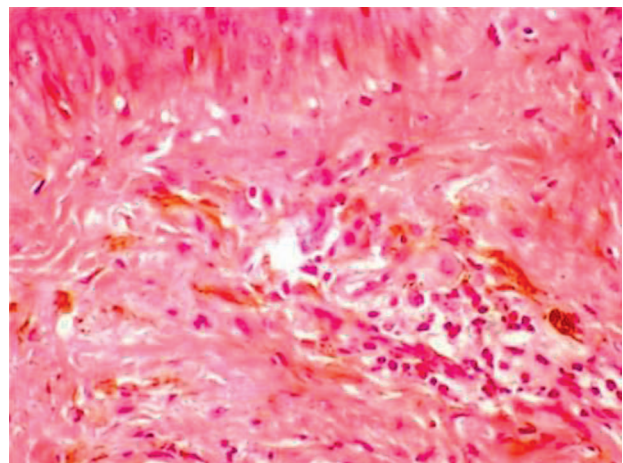


Figure 4. Histopathologic view of the facial lesion: dispersed melanocytes in a ribbon-like pattern between collagen fibers in the dermis (H&E *40)

DISCUSSION

Nevus of Ota (Nevus fuscoceruleus ophthalmomaxillaris) was first described by a Japanese dermatologist in 1939¹. Ota nevus can be congenital or acquired in adolescence. It occurs almost entirely in persons of Asian descent. The clinical manifestations are usually unilateral; only 5 percent of cases are bilateral. Clinically, blue-gray macular pigmentation with irregular borders involves skin

that is innervated by the first and second branches of the trigeminal nerve. Histopathology of the affected skin shows the presence of dendritic cells containing melanin in the dermis².

Extracutaneous manifestations include ocular involvement of sclera, episclera, conjunctiva, cornea, retina, and the uveal tract. Similar discoloration can be observed in the oral mucosa (buccal and palatal), as well as in nasal mucosa and the tympanic

membrane. Leptomeninges can also be affected. Open angle glaucoma and malignant melanoma involving the eyes are rare associations reported ³.

Nevus of Ota has been associated with idiopathic facial neuralgia, Sturge Weber syndrome ⁴, ipsilateral sensory neural hypoacusia, neurofibromatosis ⁵, primary retinitis pigmentosa and multiple blue nevi ⁶. Malignant transformation of the nevus of Ota to melanoma has been reported several times. Melanoma arising in the choroid, brain, orbit, iris, ciliary body, or optic nerve in association with a nevus of Ota has been described; therefore, careful observation is mandatory in these patients ⁶.

Various therapies have been successfully used. Cosmetic cover-up products can be used for camouflage. Cryosurgery and microsurgical treatments can leave disfiguring scars and are not recommended. Combined dermabrasion and the carbon dioxide snow method have produced good results ⁷. In recent years, use of laser therapy has been very effective and has given new hopes to patients with the nevus of Ota. The best results for the treatment of this condition are achieved with Q-switched Nd-YAG, ruby, and alexandrite lasers or a combination of them ⁸. Although there are some reports of bilateral nevus of Ota in literature ^{9,10}, almost all of them are acquired. The point of this case is that it was congenital. As far as we know, this is the first reported case of bilateral "CONGENITAL" nevus of Ota in association with Mongolian spot in a patient.

REFERENCES

1. Ota M. Nevus fuscocoeruleus opthalmomaxillaris. *Tokio Med J* 1939; 63: 1243-5.
2. Kishikawa T, Suzuki T, Sasakia Y, Aihara K, Hirayama T. Characterization of melanosomes and melanogenesis in cells cultured from Ota's nevus. *J Submicrosc Cytol Pathol* 1997;29 339-52.
3. Mishima Y, Mevorah B. Nevus Ota and naevus Ito in American Negros. *J Invest Dermatol* 1961;36: 133-54.
4. Irimia A, Preda M, Ciuca CA, Gavrilă CD. Sturge Weber syndrome in association with oculo-dermal melanosis. *Oftalmologia* 2007;51: 45-9.
5. Gupta A, Ram J, Jain IS. Nevus of Ota associated with neurofibromatosis. *Ann Ophthalmol* 1986;18: 154-5.
6. Rivers JK, Bhayana S, Martinka M. Dural melanoma associated with ocular melanosis and multiple blue nevi. *J Cutan Med Surg* 2001; 5: 381-5.
7. Hata Y, Matsuka K, Ito O, Matsuda H, Furuichi H, Ishizu N, Konstantinos A. Treatment of naevus Ota: combined skin abrasion and carbon dioxide snow method. *Plast Reconstr Surg* 1996; 97: 544-54.
8. Omprakash HM. Treatment of nevus of OTA by Q-switched, frequency doubled, ND:YAG laser. *Indian J Dermatol Venereol Leprol* 2002; 68: 94-5.
9. Kim SK, Kang HY. Centrally located acquired bilateral nevus of Ota-like macules (Hori's nevus): is this a novel type? *Eur J Dermatol* 2008;18:596.
10. Ruiz-Villaverde R, Sánchez-Cano D, Villaverde-Gutiérrez C. Bilateral naevus of Ota in a pregnant white women. *Clin Exp Dermatol* 2009; 34: 422-4.