Bilateral congenital nevus of Ota in association with Mongolian spot

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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. 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).
DISCUSSION

Nevus of Ota (Nevus fuscoceruleus ophthamomaxillaris) was first described by a Japanese dermatologist in 1939\(^1\). 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 dermis\(^2\).

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.

Nevus of Ota has been associated with idiopathic facial neuralgia, Sturge Weber syndrome, ipsilateral sensory neural hypoacusia, neurofibromatosis, primary retinitis pigmentosa and multiple blue nevi. 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.

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. 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. Although there are some reports of bilateral nevus of Ota in literature, 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