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Bilateral nevus of Ota in association with nevus flammeus: a case report

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Abstract

Nevus of Ota and nevus flammeus are distinct congenital pigmentary and vascular anomalies, respectively, with different etiologies and clinical implications. While both conditions can present unilaterally, their bilateral coexistence is rare. We report a 39-year-old Filipino female with a medical history of type 2 diabetes mellitus and dyslipidemia who presented to the dermatology clinic with concerns over a newly appearing benign nevus on the sole of her hand. Incidentally, dermatological examination revealed two significant congenital pigmentary lesions: bilateral nevus of Ota and a widespread port-wine stain. The nevus of Ota manifested as well-defined blue-gray macules and patches symmetrically distributed across her face, involving both nasal nostrils and conjunctiva. The nevus flammeus, a congenital vascular malformation, extended over both arms, the upper back, and the chest, presenting as a well-demarcated reddish-purple discoloration. These lesions had been present since birth, remained asymptomatic, and had never been previously evaluated. No systemic involvement was detected, and laboratory investigations were unremarkable. The diagnosis was made clinically. As the patient was not interested in treating the nevi, no treatment was planned. This case highlights the importance of obtaining a detailed family history in all similar cases to aid in understanding the genetic basis of these conditions and their potential coexistence with other congenital dermal anomalies.

Introduction

Nevus of Ota, also known as ocular dermal melanocytosis, is a congenital, benign gray-blue hyperpigmentation disorder of the facial area supplied by the first and second trigeminal nerves.[1] It mainly occurs due to the entrapment of melanocytes in the conjunctiva, sclera, and ipsilateral facial skin.[2] Genetically, 6% of nevus of Ota cases are caused by mutations in the *GNAQ* gene, coding for the alpha subunit of the Gq class of G-protein-coupled receptors.[3] This condition is more prevalent in females of Asian origin. In particular, Japan has an incidence rate of 1 to 2 per thousand individuals. Typically, manifestations begin unilaterally at birth, although bilateral nevus of Ota is rare, forming 10% of the confirmed cases.[4]

In contrast, nevus flammeus, commonly known as a port-wine stain (PWS), is a congenital vascular malformation characterized by a lifelong pink-to-reddish patch that can appear unilaterally or bilaterally on any part of the body. It is considered a common vascular disorder

affecting 0.3% to 0.5% of neonates.[5] Therefore, this report highlights a rare case of bilateral nevus of Ota in association with port wine stain.

Case Report

We report a 39-year-old Filipino female with a medical history of type 2 diabetes mellitus and dyslipidemia who presented to the dermatology clinic with concern over a recently appeared benign nevus on the sole of her hand, which had been gradually increasing in size. Upon examination, she was found to have two significant dermatological features besides her chief complaint, which had been present since birth and were unbothered by the patient. She had a bilateral nevus of Ota and a port-wine stain. The nevus of Ota was present as well-defined blue-gray pigmented macules and patches scattered across the patient's face bilaterally, involving bilateral nasal nostrils and bilateral conjunctiva (Figures 1 and 2). Additionally, the patient's port-wine stain was noted to cover bilateral arms and extend to the upper back and chest (Figures 3 and 4). This lesion has been present since birth and is characterized by its well-demarcated, reddish-purple discoloration. Systemic involvement was not observed in this case, and laboratory results were insignificant. The diagnosis was made based on clinical evaluation.

The patient expressed disinterest in treating the nevi; therefore, no treatment was scheduled. She was reassured and informed that no intervention had been performed.

Discussion

Nevus of Ota, also known as ocular dermal melanosis, is a benign form of melanosis that primarily affects the areas innervated by the trigeminal nerve, especially the ophthalmic (V1) and maxillary (V2) branches. It is characterized by distinct gray-blue hyperpigmentation caused by trapped melanocytes, usually presenting unilaterally.[1] This condition commonly involves the facial skin as well as the conjunctiva and sclera, with a higher prevalence in the Asian population, affecting 0.014% to 0.034% and occurring more frequently in females than males at a ratio of 5:1.[4] The clinical symptoms are typically found unilaterally, with only 5-10% of cases occurring bilaterally. The exact cause remains unknown, but various theories suggest impairment of melanocyte migration from neural crest cells to the epidermis and genetic factors, such as mutations in *BRAF*, *NRAS*, and *GNAQ*. Potential triggers include previous exposure to radiation or hormonal changes during puberty or pregnancy. Pathologically, it is characterized by numerous spindle-shaped or

dendritic melanocytes in the dermis, with lesions that are primarily macular (rarely papular or nodular) and can vary in color from brown to slate blue and gray-black, with deeper lesions appearing blue. Typically asymptomatic, rare cases may report sensory loss, and pigmentation can also affect extracutaneous sites like the eyes, oral cavity, and nasal mucosa. Diagnosis involves a thorough clinical assessment, including history and visual evaluations, along with ophthalmic examinations that assess visual acuity, intraocular pressure, and fundus examination. Dermatoscopy usually reveals bluish to slate-gray uniform pigmentation, and a skin biopsy may be warranted if there are signs of ulceration or changes in pigmentation. Understanding the features, potential causes, and diagnostic methods of nevus of Ota is essential for appropriate management and monitoring.[5]

Nevus of Ota is typically unilateral and sporadic. Bilateral and familial cases are rare.[6,7] Although bilateral acquired cases have been reported in the literature, congenital bilateral cases are relatively uncommon. Rahimi *et al.* reported the first case of bilateral "congenital" nevus of Ota in association with a Mongolian spot. It was described as asymptomatic blue-grey hyperpigmented patches over the temples, eyelids, and cheeks since birth.[8] Another case of bilateral nevus of Ota was identified in a brother and sister born to non-consanguineous parents. Both siblings had characteristic ocular and skin features consistent with nevus of Ota, while their parents showed no evidence of abnormal skin pigmentation.[9]

While establishing the differential diagnosis of the combination of bilateral nevus of Ota and nevus flammeus, it is important to rule out similar syndromes presenting with vascular and pigmentary anomalies. One of these syndromes is Klippel-Trénaunay syndrome (KTS), an uncommon congenital syndrome representing a triad of capillary malformations (port-wine stains), venous malformations (like varicose veins), and soft tissue or bony overgrowth, usually affecting one limb.[10]

In our patient, the absence of venous malformations and limb overgrowth renders KTS unlikely. This is a necessary differentiation since KTS is associated with complications like deep vein thrombosis, chronic venous insufficiency, and limb length discrepancy that demand multidisciplinary management.[11]

Ultimately, our patient's combination of bilateral nevus of Ota and nevus flammeus without vascular anomalies or limb hypertrophy is more in line with isolated vascular and pigment conditions rather than syndromic associations of KTS.

In this report, we present a rare presentation of bilateral congenital nevus of Ota in association with a port-wine stain, highlighting an unusual combination of two distinct congenital conditions in a single patient.

Conclusions

This case report highlights the rare occurrence of bilateral congenital nevus of Ota in association with port-wine stain. The increasing number of familial cases of nevus of Ota provides further support for its hereditary nature. We advise obtaining a comprehensive family history in all analogous cases to facilitate the understanding of the genetic foundation of this condition and its possible coexistence with other congenital dermal anomalies.

References

1. Patel BC, Egan CA, Lucius RW, et al. Cutaneous malignant melanoma and oculodermal melanocytosis (nevus of Ota): report of a case and review of the literature. J Am Acad Dermatol 1998;38:862-5.
2. Shetty SR, Subhas BG, Rao KA, Castellino R. Nevus of ota with buccal mucosal pigmentation: a rare case. Dent Res J (Isfahan) 2011;8:52-5.
3. Williams NM, Gurnani P, Labib A, et al. Melanoma in the setting of nevus of Ota: a review for dermatologists. Int J Dermatol 2021;60:523-32.
4. Pérez ME, Bley C, Cárdenas C. Nevus of Ota, a classic presentation. Medicina Clinica 2018;153:92.
5. Elmas ÖF, Kilitçi A. Dermoscopic findings of nevus of Ota. Balkan Med J 2020;37:116.
6. Trese MT, Pettit TH, Foos RY, Hofbauer J. Familial Nevus of Ota. Ann Ophthalmol 1981;13:855-7.
7. Kumari R, Thappa DM. Familial nevus of Ota. Ind J Dermatol 2006;51.
8. Rahimi H, Yousefi M, Mirnezami M, Asadi-Kani Z. Bilateral congenital nevus of Ota in association with Mongolian spot. Iran J Dermatol 2011;14:68-70.
9. Goyal S, Uwaydat SH, Phillips PH, Schaefer GB. Bilateral familial nevus of Ota. J AAPOS 2014;18:609-10.
10. Sharma D, Lamba S, Pandita A, Shastri S. Klippel–Trénaunay syndrome—a very rare and interesting syndrome. Clin Med Insights Circ Respir Pulm Med 2015;9:1-4.
11. Sung HM, Chung HY, Lee SJ, et al. Clinical experience of the Klippel-Trenaunay syndrome. Arch Plast Surg 2015;42:552-8.

Figure 1. Bilateral nevus of Ota with bilateral scleral involvement.

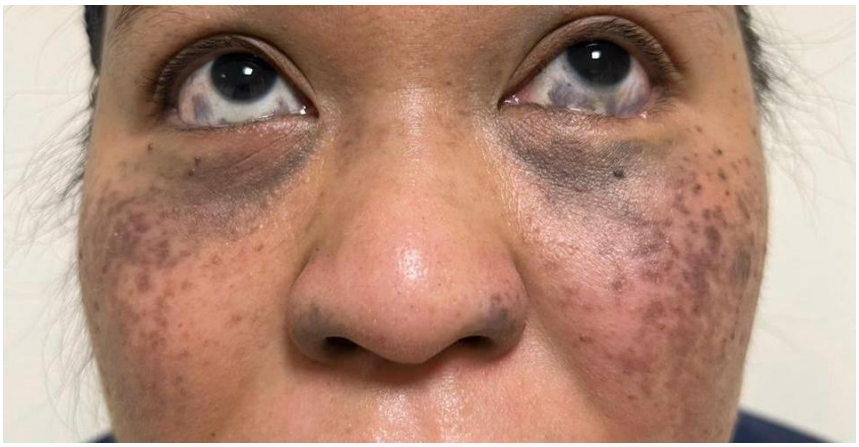

Figure 2. Bluish-grey patches on bilateral cheeks, side view.

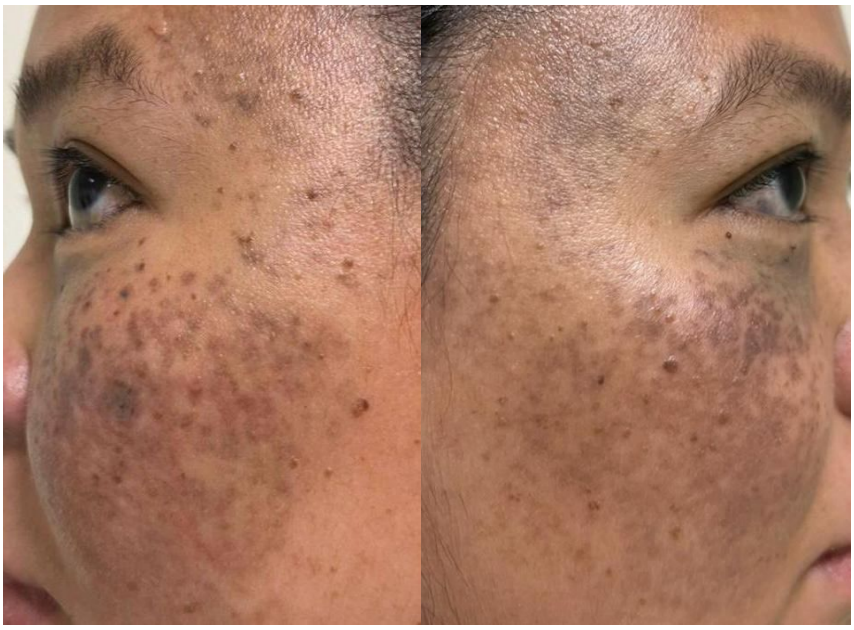

Figure 3. Blanchable reddish-purple patches on the anterior aspect of the chest and upper limbs.

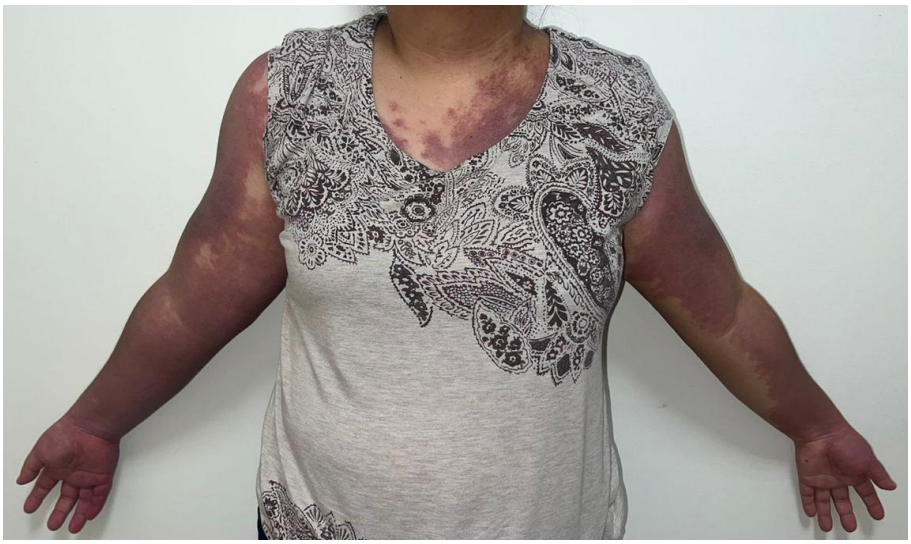

Figure 4. Blanchable reddish-purple patches on the posterior aspect of upper limbs.