Case Report

Co-existence of Bilateral nevus of Ota with port-wine stain- A Rare Case Report

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Abstract

Nevus of Ota is a rare hamartoma of dermal melanocytes involving the ophthalmic and maxillary division of trigeminal nerve and presenting as blue-grey patches on the face. It may involve ocular, mucosal and orbital structures in addition to skin. It may be congenital or acquired. Whereas, Port-wine stain is a capillary vascular malformation present since birth. It appears in the form of red macules in childhood eventually increasing in size and deepening in color as age increases. We describe here a case of nevus of Ota with port wine stain presenting together in an eight year old girl.

Keywords: Bilateral Nevus of ota, hamartoma, port wine stain.

Introduction

Nevus of Ota, also termed as oculodermal melanocytosis is a rare congenital or acquired hamartoma of dermal melanocytes described first time by Ota and Tanino in 1939.[1]

It shows distribution along the ophthalmic and maxillary divisions of trigeminal nerve. It affects only 0.014% – 0.034% Asian people.[2] Female to male ratio is 5:1.1. [3] Clinically, condition presents as blue or grey macular patch on face either unilaterally or bilaterally. Nevus of Ota is usually unilateral, whereas only five to ten percent cases are bilateral.[3] Less than 20% show palatal involvement.[4]Port-wine stain (PWS)is a macular patch present since birth persisting throughout life. resulting due to capillary vascular malformation. Its prevalence in children accounts to three to five children per 1000 live births. Appears commonly on face as red macular patch darkening eventually with age. Other sites like neck, trunk and limb involvement are less common[5] Here, we present a rare case of bilateral nevus of Ota coexisting with port-wine stain in an eight year old girl.

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Case History

An eight year old girl presented to the Dermatology OPD with complaints of greyish pigmentation over the skin surrounding both eyes and in the oral cavity. She also had red lesions over chest, neck, back and upper limbs. Her mother gave the history of the presence of both these lesions since birth. The lesions over chest, back and limbs were initially small and red but gradually increased proportionately with her growth. There was no history of seizures or any other neurological deficits. The mother gave history of noticing grayish spots over the right eye, but there was no history of any visual disturbances. The girl attained her development milestones appropriately as per the age. She was not on any medications. Clinical examination revealed bluish-grey pigmented macules present symmetrically over bilateral peri-ocular regions, right upper eye lid and right conjunctiva, with involvement of hard palate (Figure 1). Blanchable well defined erythematous macules were present over chest, back and bilateral upper limbs (Figure 2).The macules had geographic margins. There was no warmth or thrills present over the macules and there were no bruits heard on auscultation. Her visual acuity was normal. Otorhinolaryngology examination was normal. Histological examination of grayish macule from right zygomatic area showed spindle shaped melanocytes with dendritic processes predominantly in the superficial dermis. Skin biopsy taken from the erythematous macule over left subscapular area revealed multiple dilated capillaries of various sizes with single layer endothelial lining in the papillary dermis (Figure 2) and deeply pigmented dendritic melanocytes in the dermis (Figure 2).

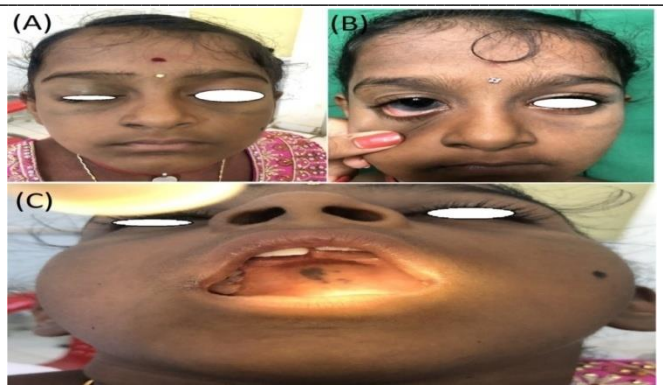

Fig 1: (A) Hyperpigmented macules present over right eyelid and right conjunctiva.
(B) Greyish macule present over right conjunctiva
(C) Hyperpigmented macule present over hard palate

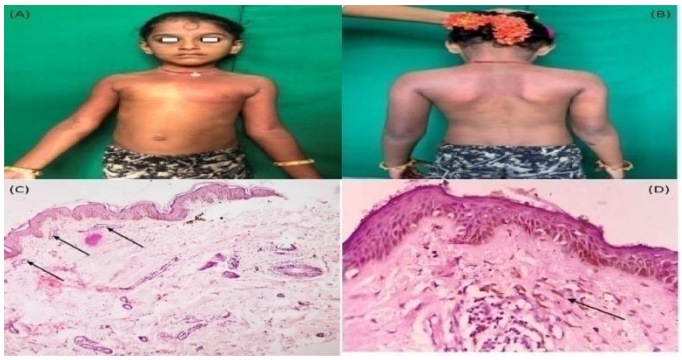

Fig 2:(A –B) Port wine stain : Erythematous macule present over bilateral upper limbs, chest and back
(C) Port wine stain: multiple dilated capillaries of different sizes with single layer of endothelial lining in papillary dermis (H and E stain, x10, Arrow)
(D) Nevus of Ota : Normal epidermis and deeply pigmented slender dendritic melanocytes scattered with collagen bundles in dermis (H and E stain, x40, Arrow)

Discussion

Nevus of Ota is caused due to incomplete migration of melanocytes from the neural crest to the skin in embryonic life. This results in dermal distribution of these melanocytic cells in the areas of innervations of ophthalmic and maxillary branches of trigeminal nerve.[6] This condition presenting as blue-gray macules,commonly involves areas like forehead, temple, periorbital area, cheek, and nose. Inside the oral cavity, hard palate is involved the most.[7]There may be variation in the color depending on physiological, environmental conditions and also on the depth of involvement of skin as deeper lesions appear blue in color due to the Tyndall effect,[6] whereas superficial lesions appear slate gray in color.Genetic factors and hormones may be the reasonfor predominance in females. In some cases it may be associated with open angle glaucoma or malignant melanoma.[6]Nevus of Ota is classified clinically into mild, moderate, severe and bilateral types.[4] Histologically, classification of nevus of Ota is based on the location of the dermal melanocytes. These include Superficial (type S), superficial dominant (type SD), diffuse (type Di), deep dominant (type DD) and deep (typeDe)[6]. In our case, the melanocytes were found to be predominantly distributed in the superficial dermis, thus falling under the Superficial Dominant (SD) type. The differential diagnosis of nevus of Ota include Hori's nevus[8], Mongolian Spot,[6,9] melasma and hyperpigmentation due to medications. Hori's nevus, also known as Acquired Bilateral Nevus of Ota-like Macule (ABNOM) presents in adults with bilateral involvement of face with blue- brown or slate-gray colored macules and without mucosal involvement [10] while Mongolian spot presents as large diffuse blue- to-gray patches typically occurring in lumbo-sacral area and resolves by 3– 6 years of age. It rarely occurs on the face. Drug induced hyper-pigmentation can be associated with minocycline, amiodarone and gold. Port wine stain appears as red macular patches initially but progresses over the years and turns purplish dark, with hypertrophy of the underlying soft tissue. This capillary malformation is usually diagnosed clinically. Almost 90% lesions are on the face however trunk, back and extremities are also involved. Most patients seek treatment due to cosmetic reasons. Syndromes commonly associated with Port wine stain are Sturge-Weber syndrome, Klippel-trenaunay syndrome. Nevus of Ota is diagnosed clinically but biopsy is confirmative. The introduction of pigment lasers have improved treatment outcomes in cases of nevus

of Ota. Other treatment options include laser, cryotherapy, dermabrasion with autologous cultured epithelium grafting, microsurgery and cosmetic camouflage. Q- switched Nd: YAG and Q- switched Alexandrite lasers have become gold standard in the treatment of nevus of Ota.[11] Treatment options for port wine stain include pulsed dye laser (PDL) and photodynamic therapy. Our patient was unwilling to undergo treatment with lasers owing to unaffordability.

Conclusion

Periodic examination should be conducted in such cases for early diagnosis of complications like glaucoma and melanoma.[2] They should also be screened for the emergence of complex vascular malformations, limb length discrepancies and Sturge-Weber syndrome.Our case has been reported owing to its rarity, as it involves bilateral Nevus of Ota with hard palate involvement coexisting with port wine stain involving the extremities.

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