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Dermatology

AN UNUSUAL ASSOCIATION OF PARTIAL UNILATERAL LENTIGINOSIS AND CENTRAL HETEROCHROMIA

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ABSTRACT Partial unilateral lentiginosis (PUL) is a rare pigmentary disorder that is characterized by multiple clustered lentigens on a normal skin in a segmental pattern with a sharp demarcation at the midline. Heterochromia is a condition of the eye in which there is variation in iris colour of one or both eyes. There has been multiple associations of PUL and has also been associated with ocular involvement but has never been associated with heterochromia. Herein we report an undocumented association of PUL with central heterochromia.

KEYWORDS : Partial unilateral lentiginosis, Heterochromia iridium, Central Heterochromia, Pigmentary disorder, Somatic mosaicism

Introduction

Partial unilateral lentiginosis is a rare cutaneous pigmentation disorder characterized by multiple simple lentigines on the normal skin, with a unilateral segmental pattern, which is known as segmental lentiginosis, lentiginous mosaicism, unilateral lentiginosis, partial lentiginosis or agminated lentiginosis. Although there is usually no abnormality, the disorder can occasionally be associated with neurofibromatosis, or café-au lait spots.[1,2] Heterochromia is a condition of the eye in which there is variation in iris colour of one eye or both eyes.[3] PUL and heterochromia are melanin pigmentary disorders, but no documented correlation exists to our knowledge.

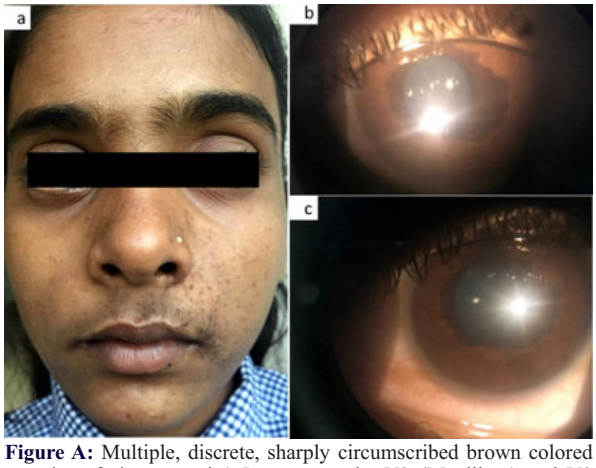

Figure A: Multiple, discrete, sharply circumscribed brown colored macules of size around 1-5 mm over the V2 (Maxillary) and V3 (Mandibular) divisions of the trigeminal nerve on the left side of face with a sharp midline demarcation.

Figure B & C: Slit Lamp Examination Revealed Central Heterochromia In Both Eyes.

Case Report

A 12 year old girl presented to the out patient department with multiple asymptomatic brown spotty pigmentation over left side of face. The lesions first appeared 5 years back on the lower part of left cheek and gradually multiple lesions appeared over the left part of nose and lips. No change was observed in the color, number and dimensions of the lesions during the past 2 years. No history of similar lesions on body and no family history was present. There was no significant medical history. General examination was found to be normal. Cutaneous examination revealed multiple, discrete, sharply circumscribed round to oval brown colored macules of size around 1-5 mm over the V2 (Maxillary) and V3 (Mandibular) divisions of the trigeminal nerve on the left side of face with a sharp midline demarcation (Figure 1). No other mucocutaneous abnormalities were noted. There was no evidence of any café au lait macules, axillary freckling, palmar freckling and neurofibromas. Slit lamp examination revealed central heterochromia in bilateral eyes. No other pathologic finding was detected during neurological, orthopedic and any other systemic examinations. Dermoscopic examination revealed an accentuated regular brown-colored symmetrical pigment network with a sharp demarcation at the periphery. Histopathology could not be performed as the patient refused. Results of routine laboratory examinations were within normal limits. Clinical and dermoscopic findings lead to a diagnosis of Partial unilateral lentiginosis with central heterochromia. The patient is being planned for Q- switch ND YAG laser for the lentigens.

Discussion

Partial unilateral lentiginosis (PUL) (also called segmental lentiginosis, lentiginous mosaicism, "zosteriform" lentiginous nevus or agminated lentiginosis) is a rare pigmentary disorder characterized by numerous clustered lentigines on the normal skin in a segmental pattern with a sharp demarcation at the midline. Although it can be seen anywhere on the body but face, neck, and extremities are the most common sites of involvement. Generally lesions are present at birth or first noticed during childhood but some adult cases have been described. The mean age is 5 years with a range from birth to 15 years.[1,2]

The pathogenesis is not completely understood, but a somatic mosaicism hypothesis has been proposed during embroyonic development, possibly due to a mutation in the neural crest melanoblasts. Several developmental pigmentary genes such as SNAI2, KITLG, EDNRB, EFNB1, EFNB2 have been related to pigmentary mosaicism and have a role in neural crest differentiation and melanoblast migration. Postzygotic somatic mutations could also be an explanation for the presence of both segmental Neurofibromatosis and PUL in the same patient.[2,4]

PUL should be differentiated from nevus spilus and nevus of Ota. Nevus spilus has hyperpigmented background speckled with multiple dark brown macules with a more stable course with time. Nevus of ota on the other hand presents with ocular pigmentation and mottled blue gray pigmentation unlike the brown colored pigmentation on the skin and eye in PUL.[5,6] Multiple other genetic diseases may present with

lentigines, such as Leopard syndrome, segmental neurofibromatosis, Peutz- Jegher's syndrome and Carney's complex.[7,8,9] These can be differentiated from PUL in that it has a typically unilateral and segmental distribution and lacks all other clinical signs and symptoms found in them.[1] Neurologic, endocrinological, or hematological disorders can be associated with this disorder, although most patients do not exhibit any abnormalities.[10] PUL has been associated with several cutaneous and systemic disorders (Table 1).[2]

Table 1: Reported Associations Of Partial Unilateral Lentiginosis

Colocalisation of café au lait macules usually in same distribution as lentigens
Ipsilateral Lisch Nodules
Segmental Neurofibromatosis
Colocalisation of blue nevus, Nevus of ota
Ipsilateral ocular involvement: Bulbar conjunctival or uveal pigmentation
Mental retardation, ipsilateral cerebrovascular abnormalities, focal epilepsy
Ipsilateral lateral popliteal nerve palsy
Ipsilateral sensory motor neuropathy
Ipsilateral rigid cavus foot
Probably Coincidental: Goitre, Anemia, Brochial asthma, celiac disease, Vitiligo,

Eye color is determined by melanin concentration and distribution within the iris tissues Heterochromia is a condition of the eye in which there is variation in iris colour of one eye or both eyes. There are a variety of causes, including hypopigmentary disorders such as Sturge Weber syndrome, Waardenburg syndrome, piebaldism & hypomelanosis of ITO.[3] There has been no association between heterochromia and PUL till date.

Ours is the first reported case of heterochromia associated with PUL. This report draws attention to the entity PUL and its possible associated ocular involvement. We cannot be certain whether the heterochromia in our patient represent a true association or a mere coincidence. Nonetheless, we report this case to increase the awareness among dermatologists regarding the ocular involvement in partial unilateral lentiginosis.

Conflict Of Interest Nil

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