LINEAR AND WHORLED NEVOID HYPERMELANOSIS

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ABSTRACT: Linear and whorled nevoid hypermelanosis is a rare sporadic skin condition characterized by swirling streaks of hyperpigmention following the lines of Blaschko. The hyperpigmentation often occurs alone, but additional symptoms have been described in individual cases. An 18 year old girl presented with insidious onset, asymptomatic, generalized, hyperpigmented streaky lines since late childhood without any other systemic abnormality. Histopathology showed increase in basal cells containing melanin without any dermal change, consistent with linear and whorled nevoid hypermelanosis.
KEY WORDS: Linear, whorled, nevoid hypermelanosis, lines of blaschko.

INTRODUCTION: Linear and whorled nevoid hypermelanosis (LWNH) was first described by Kalter et al[1] in 1988 as a sporadic pigmentary anomaly characterized by swirls and streaks of macular(flat) hyperpigmentation following the lines of Blaschko.Usually occurring as isolated disorder of pigmentation, associated chromosomal abnormalities with central nervous, cardiovascular, musculoskeletal and ocular abnormalities have been reported.

CASE REPORT: An 18 year old female complained of hyperpigmented (dark coloured) streaky skin lesions on face, trunk and extremities, occurring insidiously, since late childhood. Lesions were first noticed on the face and subsequently elsewhere. Lesions were asymptomatic but cosmetically disfiguring. Retrospective history from mother, failed to elicit presence of fluid filled or elevated skin lesions preceding the pigmentation. She was born of full term, normal vaginal delivery, to non-consanguineous parents, with normal growth and development.Family history was not suggestive.

General and systemic examination including detailed musculoskeletal, neurological, ocular and cardiovascular assessment was normal. Cutaneous examination revealed macular hyperpigmented lesions in streaks and whorls following the lines of Blaschko. Lesions were distributed as bilaterally symmetrical linear streaks on upper and lower extremities while on the chest and trunk, lesions were patchy whorls which did not cross the midline. Facial lesions were seen over the forehead and pre-auricular areas.No skin surface changes like roughness or scaling was noted over the lesion or elsewhere. Palms, soles, mucosa, hair, nails, teeth were normal.

Differential diagnoses considered were epidermal naevus, incontinentia pigmenti and linear and whorled naevoid hypermelanosis.

Hemogram and metabolic profile presented no abnormalities. X ray skull and ECG were normal. CT scan brain, echocardiography and chromosomal studies could not be done due to financial constraints. Skin biopsy revealed-mild papillomatosis and diffuse increase in basal cells containing melanin without any dermal change, which was consistent with linear and whorled naevoid hypermelanosis. The benign nature of the disease was explained to the patient. She was prescribed glycolic acid 6% crème for the facial lesions along with sunscreen.

DISCUSSION: Kalter et al. described LWNH as usually starting within the first few weeks of life with no preceding inflammation or palpable lesion and progressing during the 2nd-3rd years of life before stabilizing[1]. Patients typically present with reticulate hyperpigmented macules that coalesce to form streaks and whorled hyperpigmentation following the lines of Blaschko[2] and do not cross the midline. The trunk, extremities, neck, face and genitalia are the typical sites affected[2, 3, 4]with sparing of palms, soles, eyes and mucous membranes. Similar observations were made in our patient.

LWNH has also been described under different names as zosteriform hyperpigmentation, zosteriform lentiginous nevus and 'zebra like pigmentation'[5] referring to its streaky distribution along lines of Blaschko resembling stripes on a zebra. First described and drawn in 1901 by Alfred Blaschko[6], the lines of Blaschko represent the developmental growth pattern of the skin and its appendages and donot correspond to any known nervous, vascular or lymphatic structures[7]. No single theory as yet, clearly elucidates the localization of these lines but their characteristic pattern is mostly thought to be due to presence of two genetically different clones of cells in an individual, derived from a single zygote representing a human`mosaicism'[8, 9]. As such, LWNH is said to be due to developmental somatic mosaicism causing proliferation and migration of two mixed populations of melanocytes with different potential for pigment production.[1, 10]

Nearly all cases have occurred sporadically, however apparent genetic transmission has been described in two families [5, 11].Children with LWNH often have otherwise normal physical and neurologic development, however individual cases occurring along with chromosomal anomalies like mosaic trisomy of 7, 14, 18, 20 and X chromosomal mosaicism, congenital heart diseases (ventricular septal defect and tetralogy of Fallot)[12], CNS diseases (microcephaly, arhinencephaly and epilepsy)[13], psychomotor delay, deafness and brachydactyly have been mentioned in the literature. This patient did not have any extracutaneous presentation reported with LWNH though high end radiological and chromosomal studies could not be done.

The differential diagnosis includes incontinentia pigmenti and epidermal nevus. Incontinentia pigmenti[14],has four successive cutaneous stages: vesicles, verrucous lesions, whorl- or streak-like hyperpigmentation, and hypopigmented scars, with scarring alopecia and peg-shaped teeth. No history of preceeding lesions with histological absence of basal cell degeneration, incontinence of melanin pigment, tissue eosinophilia, and melanophages in the dermis ruled out the possibility of incontinentia pigmenti in the present case.

Epidermal nevi[15] are often noticeable during infancy as hyperpigmented streaks along Blaschko's line, which become papillomatous and hyperkeratotic with time. Extensive skin involvement is often associated with skeletal, ocular, and nervous system anomalies. In the absence of verrucous(raised) skin lesions and histopathologic evidence of hyperkeratosis, acanthosis, elongation of rete ridges, papillomatosis, the possibility of epidermal nevus was not entertained.

No effective treatment is available for LWNH though chemical peels [3], depigmenting agents [16]and lasers have been tried without much success.Cosmetic camouflage does offer some promise.

Office of Rare Diseases Research (ORDR), National Institutes of Health(NIH), U.S. Department of Health and Human Services, records only 45 cases of LWNH to be reported till date[17]. This case has been presented keeping in mind its rarity. And also, to stress upon the fact that, LWNH may be associated with congenital anomalies and thus require detailed examination and investigations.

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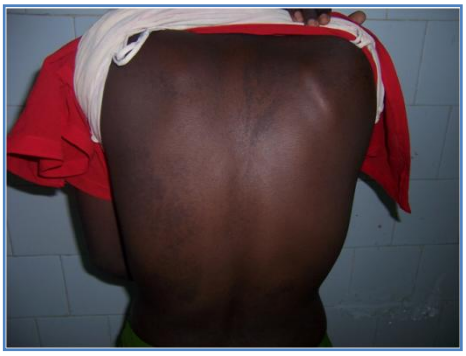

FIG. 1: Bilateral macular hyperpigmentation with sharp mid-line demarcation on the trunk

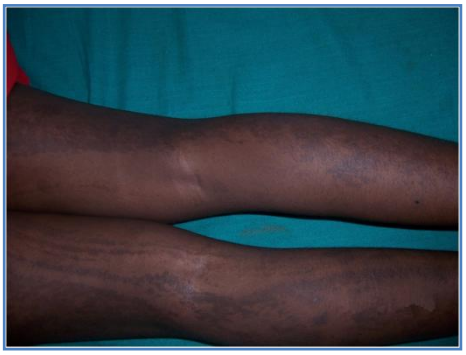

FIG. 2: streaks of macular hyperpigmentation following the lines of Blaschko on the legs

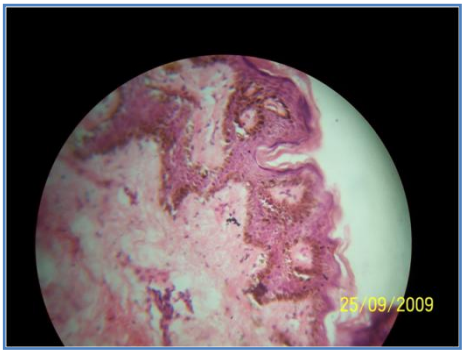

FIG. 3: Photomicrograph showing epidermal melanosis without pigment incontinence in the dermis

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