PAEDIATRIC DERMATOLOGY

# A RARE CASE OF LINEAR AND WHORLED NEVOID HYPERMELANOSIS WITH GLOBAL DEVELOPMENTAL DELAY, SCOLIOSIS AND RETINAL DEGENERATION WITH DERMOSCOPIC FEATURES

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Background: Only forty cases of the rare sporadic Linear and Whorled Nevoid Hypermelanosis (LWNH) are described in medical literature worldwide, characterized by hyperpigmented, reticulated, streaky and whorled patches along Blaschko lines, without atrophy or preceding inflammation. It reflects an underlying mosaicism or chimerism and occasionally associated with systemic abnormalities.

Observation: A five year-old female presented with multiple uniformly hyperpigmented patches with midline demarcation over right posterior trunk, reticulate and blaschkoid over lower extremities and linearly arranged over right upper extremity. Lesions were asymptomatic and unchanged since birth. Dermoscopy showed brown structureless zones interrupted by dotted perifollicular hypopigmentation over right posterior trunk and anterior thigh, reticular pattern over right posterior thigh and linear brown streaks with alignment along Blaschko's lines over right upper extremity. Histopathology revealed basal layer hyperpigmentation, sparse superficial lymphocytic dermal infiltrates, melanocytic hypermelanosis and flat-topped papillomatosis. Hematological and biochemical tests were non-contributory. The patient has decompensated thoracic scoliosis with two cm divergence from plumb line. Developmental Pediatrics referral revealed global developmental delay and examination under anesthesia showed peripheral retinal degeneration on both eyes.

Key Message: The pathogenesis of LWNH is unknown though developmental somatic mosaicism leading to proliferation and migration of mixed populations of melanocytes with different potential for pigment production and genetics are possible causes with nearly all cases occurring sporadically. While data is lacking because of its rarity, affected individuals should be evaluated for disease associations (delays, skeletal and other systemic abnormalities). Different dermoscopic features described in three case reports before were all present in our patient (net-like, parallel streaks, dotted perifollicular hypopigmentation)

which is in keeping with its mosaic nature. No promise of chromosomal abnormalities can be found using sequencing. Together with limited information regarding effective treatment (peels, hydroquinone 2%, depigmenting agents,