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Linear and Whorled Nevoid Hypermelanosis in a Nigerian Child: A Case Report and Literature Review

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Abstract

Linear and whorled nevoid hypermelanosis (LWNH) is a rare skin condition characterized by linear streaks and swirls of macular hyperpigmentation along the lines of Blaschko. The disease may occasionally have associated extracutaneous manifestations. The exact pathogenesis is unknown and no satisfactory treatment modalities for LWNH are currently available. There are few case reports of the disease in literature worldwide and none has been previously reported in Nigeria. We are reporting one such rare case of linear and whorled nevoid hypermelanosis in a 9 month old Nigerian child. We report a 9 month old female who presented with abnormal dark skin pigmentation noticed at birth suggestive of LWNH with extracutaneous presentation including microcephaly, turricephaly, anisocoria, nystagmus and delayed developmental milestones. This shows the existence of this rare disease in Nigeria. Linear and whorled nevoid hypermelanosis though uncommon, is associated with significant morbidity especially among those with extracutaneous lesions. No effective treatment is available for this disorder.

Keywords

Linear, Whorled, Nevoid, Hypermelanosis, Nigerian

1. Introduction

Linear and whorled nevoid hypermelanosis (LWNH) is a rare skin condition characterized by linear streaks and swirls of macular hyperpigmentation along the lines of Blaschko. [1] Onset of hyperpigmentation usually occurs within the first few weeks of life, continues to progress for a year or two before stabilization. [2] The condition was first described in 1988 by Kalter and colleagues. [3] Different descriptive terms, including ‘‘zosteriform lentiginous nevus’’, ‘‘zebra-like hyperpigmentation in whorls and streaks’’, ‘‘reticulate hyperpigmentation distributed in a zosteriform fashion’’, and ‘‘reticulate hyperpigmentation of Iijima’’ had previously been used to describe patients with this same condition. [4-8]

The occurrence of LWNH is usually sporadic although familial cases have been reported.[2,9] Both sexes are equally affected.[10] The exact pathogenesis is unknown, although genetic mosaicism has been described.[11] Histopathological examination of skin biopsy of affected patients shows diffuse increased pigmentation in the basal layer and prominence of melanocytes without incontinence of pigment in the dermis. [12]

The linear streaks and swirls of macular hyperpigmentation occur in a reticulate pattern without any preceding inflammatory or palpable verruciform lesions. [1, 12] Sites of predilection include the trunk, face, neck, upper and lower limb. The palms, soles, scalp, and mucous membranes are typically spared. [12] Extracutaneous findings including neurological, skeletal, cardiovascular and ocular anomalies have been reported. [1, 9, 12] No satisfactory treatment modalities for LWNH are currently available. There are few case reports in Literature. To the best of our knowledge, known has been reported in Nigeria.

We are reporting one such rare case of linear and whorled nevoid hypermelanosis in a 9 month old Nigerian child.

2. Case Report

A 9 month old female referred to the Consultant Paediatric Clinic (CPC) of the University of Port Harcourt Teaching Hospital (UPTH) with complaints of abnormal dark coloured skin pigmentation, inability to properly open the left eye both noticed at birth, lateral deviation of the head of six months duration, inability to sit without support and crawl.

The child was delivered at term to non-consanguineous parents and had delayed developmental milestones. No family history of pigmentation disorder was elicited and she had 3 normal male siblings. On physical examination there were multiple, linear and whorled, hyperpigmented macules, arranged bilaterally and symmetrically (zebra-like), along the lines of Blaschko on the face, trunk, abdomen, and extremities including the palms and soles of the feet and predominantly right sided (Figure 1). The mucous membranes were however spared. The texture of skin was normal over the streaks as well as the unaffected skin. There were associated central nervous system abnormalities consisting of microcephaly, turricephaly, anisocoria, nystagmus and ptosis of the left eye. Also observed were musculoskeletal system abnormalities including torticollis, and facial asymmetry. Brain MRI showed multiple congenital cerebral anomalies comprising of pachygyria with lobar holoprosencephaly and complete agenesis of the falx and corpus callosum. Routine laboratory investigations including complete blood count, biochemical and urinary analyses were within normal limits. Skin biopsy and chromosomal studies were not done due to unavailability of facilities.

A diagnosis of LWNH was established based on clinical findings. The child had multidisciplinary review by the neurologist, ophthalmologist, dermatologist and physiotherapist; and she is currently being followed up at the Consultant Paediatric Clinic.

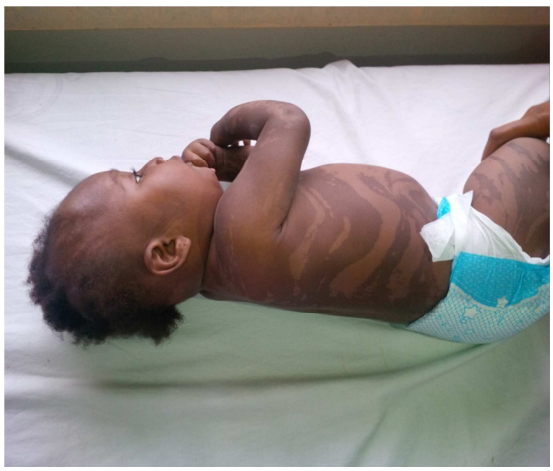

Figure 1. *Shows the index patient with linear and whorled hyperpigmented lesions following lines of Blaschko.*

3. Discussion

Linear and whorled nevoid hypermelanosis is a rare disorder characterized by reticulate hyperpigmented macules in a streaky configuration. [1, 3] It usually starts within the first few weeks of life with progression during the first 2-3 years of life before stabilizing. [2] In the index patient, skin hyperpigmentation was noticed at birth and is becoming more defined and prominent as the child grows. Although LWNH is a sporadic disorder; a familial case has been reported by Akiyama et al. [2] For the index patient, it is probably sporadic as other close family members are unaffected and there was no family history of pigmentation disorders. This is consistent with what has been previously reported in literature. [1, 5, 6]

In linear and whorled nevoid hypermelanosis, the trunk, extremities, neck, and face are the typical sites affected. [3, 13] The patient presented here showed the typical pattern and distribution of the skin lesions but with involvement of the palms and soles of the feet. The mucous membranes were spared.

Ertam et al [12] in 2008, published a case of an 11-year-old girl with hyperpigmentation along the lines of Blaschko over the entire body. The mental status of the patient was normal and no associated anomaly was detected in the physical examination or genetic analysis. Dermatologic examination revealed a whorled-like configuration of hyperpigmented macules on the neck, trunk, and buttocks, and a linear configuration of hyperpigmented macules, some of which were arranged in a parallel linear fashion on the extremities along the lines of Blaschko. Histopathologic examination confirmed the diagnosis. Dermatoscopic examination revealed linear or circular arrangement of streak-like pigmentations arranged in a parallel manner.

In 2011, Metta et al [9] reported LWNH in three successive generations. The main complaint was asymptomatic, dark coloured skin lesions over the body since early life. None of them had any history of warty lesions or blisters. There was no history of consanguinity or foetal deaths in the family. Apart from the three of them, no other family members have been affected. They all had multiple, whorled, hyperpigmented macules, arranged bilateral and symmetrical, along the lines of Blaschko on the abdomen, chest, and back. Systemic examination and mucosa were normal. Also, biochemical laboratory and radiological investigations were normal. Histopathological evaluation of the pigmented macules revealed increased pigmentation of the basal cell layer with melanocytes present up to the mid epidermis. There were focal areas of pigmentary incontinence noticed in the dermis. Genetic studies performed on the peripheral blood from the mother revealed trisomy on chromosome 20, but this study was found to be negative in the child.

The association of LWNH and extracutaneous features have been reported. [1, 9, 12, 14] Various neurological and ocular problems including severe mental retardation with macrocephaly, seizures, bilateral giant cerebral aneurysms,

nystagmus and anisocoria have been documented. [15, 16] We observed microcephaly, turricephaly, anisocoria, nystagmus, ptosis, flat occiput, delayed developmental milestones, facial assymetry and torticollis in our patient. Also, cranial MRI was abnormal in this patient with severe brain abnormalities detected. This is not surprising considering the clinical presentation of this patient.

The differential diagnosis of LWNH includes skin conditions that present with segmental hyperpigmented lesions such as incontinentia pigmenti, hypomelanosis of Ito, and epidermal nevus. In incontinentia pigmenti the skin lesions pass through four successive stages of vesicles, verrucous lesions, whorls or streak like hyperpigmentation and hypopigmented scars. [3] The index patient did not present with the above mentioned stages. The pattern of pigmentation in hypomelanosis of Ito resembles the reverse pattern of LWNH. [3] Epidermal nevi often present during infancy as hyperpigmented macules along lines of Blaschko. With time, they turn hyperkeratotic and papillomatous. The absence of verrucous lesions along with the absence of hyperkeratosis, acanthosis, and papillomatosis clinically ruled out the possibility of epidermal nevus. [3]

It has been shown that histopathological changes of linear and whorled nevoid hypermelanosis consist of diffuse hyperpigmentation in the basal layer and lack of pigmentary incontinence in the dermis. [12] However, due to the lack of facilities and parent's refusal, a skin biopsy was not done in the patient presented.

No effective treatment is available for this disorder. For lesions in cosmetically sensitive areas such as the face and the limbs, depigmentary creams have been tried with no improvement. [17] Such creams were however not available to our patient use at the moment.

Our patient is been followed up by the neurologist, ophthalmologist and physiotherapist due to the extracutaneous involvements to ameliorate the associated mobidity. The parents of our patient were also adequately counselled on child's condition.

4. Conclusion

Linear and whorled nevoid hypermelanosis though uncommon, contributes to significant morbidity especially among those with extracutaneous lesions. No effective treatment is available for this disorder.

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