

Linear and whorled nevoid hypermelanosis: A rare familial case report

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ABSTRACT

Linear and whorled nevoid hypermelanosis (LWNH), also known as ‘zebra-like pigmentation’ is characterized by linear and swirling streaks of hyperpigmentation along the Blaschko’s lines without preceding inflammation and atrophy. It is mainly located on trunk and limbs. The hyperpigmentation may be present at birth or may develop by early childhood. Very rarely, familial cases have been described. Here, we are reporting one such rare case of familial LWNH in a 19-year-old female.

Key words: *Familial, Hypermelanosis, Hyperpigmentation.*

Linear and whorled nevoid hypermelanosis (LWNH), also known as ‘zebra-like pigmentation’, is characterized by linear and swirling streaks of hyperpigmentation along the Blaschko’s lines. The hyperpigmentation may be present since birth or develop by early childhood. Hyperpigmentation may progress for one to two years before stabilization. It generally occurs sporadically and located on trunk and limbs. The eyes, palms, soles, and mucous membranes are usually not affected. Very rarely, familial cases have been described. Here, we are reporting one such rare case of familial LWNH.

CASE REPORT

A 19-year old female presented to the outpatient department with her 2 sisters and mother with the complaint of dark-colored skin lesions over both lower limbs since early life (Fig 1). The lesions appeared over the lower leg at the age of 8 months and gradually progressed to involve both lower legs,

thighs, and back by the age of 2 years followed by involvement of the upper limbs also (Fig 2). No history of preceding inflammation, blisters or atrophy was noted. At birth, the girl was full-term, normal vaginally delivered, with normal growth and development. No history of consanguinity was present in the family. Over the past 5 years, she started noticing multiple hypopigmented lesions appearing over hyperpigmented streaks on the lower limbs after various topical treatments, the nature of which she could not elaborate. No prior history of itching was present.

She had 3 elder sisters and one younger brother. Out of them, the eldest sister was unaffected. Two were affected, having similar hyperpigmented streaks over the upper limb since birth and out of



Figure 1: Case showing classical linear hyperpigmented streaks over extensor surface of both lower limbs



Figure 2: Case showing the linear and whorled hyperpigmented streaks along the lines of Blaschko over the trunk



Figure 3: Sister of the case showing linear and whorled hyperpigmented streaks over the trunk



Figure 4: Mother of the case showing the remnant of whorled pigmentation over the abdomen

them, one died because of unknown reason (Fig 3). Her brother was unaffected. Her mother also had similar hyperpigmented streaks over the back and abdomen (Fig 4). They also gave a history of linear pigmented streaks in her maternal grandmother. There was a history of fading of lesions with time, in her mother and grandmother.

On examination, the vitals were stable. Her palms, soles and mucous membranes were not involved. No skeletal, cardiovascular, central or respiratory system involvement was observed. Multiple hyperpigmented macules and linear streaks arranged along the lines of Blaschko on extensor aspects of both lower and upper limbs were seen. In between the hyperpigmented streaks, multiple hypopigmented macules were also present.

All routine investigations were normal. No congenital abnormality was detected in X-ray, chest computed tomography

(CT) scan and 2D Echocardiography. On histopathological examination, increased epidermal melanosis in the basal layer and a mild perivascular lymphocytic infiltrate was seen. Some melanin pigment incontinence was also seen (Fig 5).

Based on clinicopathological correlation and history, a diagnosis of linear and whorled nevoid hypermelanosis was made in all the three successive family members. The family was reassured and counseled about the benign nature of the disorder. A periodic assessment was recommended to them. However, the exact reason for hypopigmentation was not clear.

DISCUSSION

LWNH is a rare pigmentary disorder characterized by linear and whorled hyperpigmented streaks along the Blaschko lines [1]. It occurs within a few weeks of life and progresses gradually for the next few years before stabilization. It is not preceded by inflammation or atrophy. There is sparing of the face, palms, soles, and mucous membranes.

The underlying cause of LMNH is poorly understood, it is thought to result from genetic mosaicism or chimerism [2,3]. LMNH generally occurs sporadically. Very rarely, familial cases have been described. Genetic studies suggest somatic mosaicism as a cause for LWNH with mosaic trisomy of 7, 14, 18, 20; X-chromosomal mosaicism have been reported [3,4]. Various skeletal anomalies (facial and body asymmetry, brachydactylic), cardiovascular diseases (ventral septal defect, tetralogy of Fallot) and central nervous system diseases (microcephaly, mental retardation, epilepsy) can be associated with LWNH [1,5].

Histopathologically, an increase in pigmentation of the basal layer and prominence or vacuolization of melanocytes is seen. Pigmentary incontinence is usually, but not always absent [1]. Similar findings were noticed in our cases.

The differential diagnosis [5] includes incontinentia pigmenti (IP), epidermal nevus and hypomelanosis of Ito. In incontinentia pigmenti, cutaneous manifestations pass through

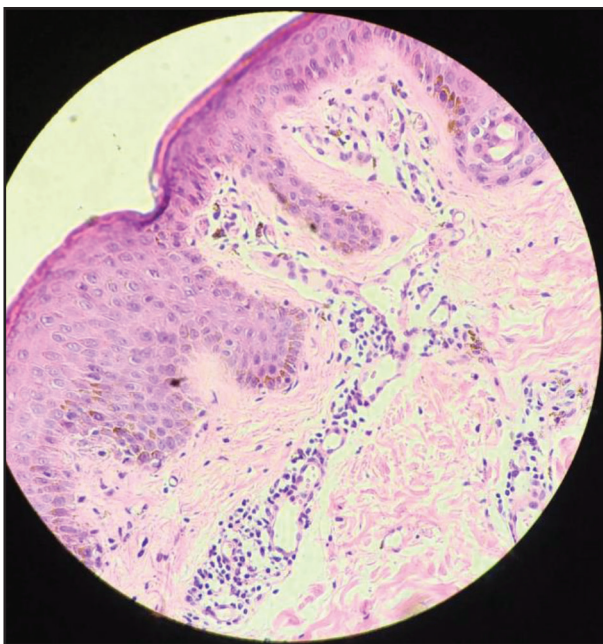


Figure 5: Histopathological picture of a biopsy taken from the leg showing prominent basal melanization without pigmentary incontinence (H and E stain, x40)

vesicular, verrucous, whorls or streaks like hyperpigmentation and hypopigmented scar stages. Skin appendages are usually affected by this. Histopathologically, basal cell degeneration, pigmentary incontinence, dermal melanophages, and tissue eosinophilia are seen. Lack of these features, rule out the possibility of IP in this case. Whereas, epidermal nevi, usually appear during infancy as pigmented streaks along the Blacklines followed by verrucous stage which is histopathologically characterized hyperkeratosis, acanthosis, papillomatosis, and elongated rete ridges.

CONCLUSION

The familial occurrence of LWNH in three successive generations is extremely rare conditions. To the best of our knowledge, this is the second case of LWNH occurring in three successive generations being reported in the literature from India.

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