LINEAR AND WHORLED NEVOID HYPERMELANOSIS WITH DELAYED MILESTONES

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PRESENTATION OF CASE

A 6-months-old female presented with progressively increasing diffuse dark-coloured lesions over the trunk and extremities with delayed milestones. According to the mother, the lesions started appearing after first week of life. Physical examination revealed streaks and whorls of hyperpigmented macules along the lines of Blaschko, located on the trunk and limbs [Figure 1]. The mother gave negative history of any preceding inflammation and positive history of two episodes of seizures in the first month of life, for which no cause was identified. Systemic examination showed no significant anomaly. The girl is the second child with a five-year-old unaffected sister. She has no similar case in her family. Haematological and biochemical investigations were within normal limits. Skin biopsy revealed increased melanin content in the basal layer with increased number of melanocytes. Basal cell degeneration and pigmentary incontinence in the dermis was not observed.

CLINICAL DIAGNOSIS

Linear and Whorled Nevoid Hypermelanosis (LWNH)

It was first described by Kaltar et al[1] in 1988, and is characterised by hyperpigmented macules along the lines of Blaschko without any preceding inflammation or atrophy. Lesions usually start occurring within the first few weeks of life, and are mainly distributed on the trunk and extremities, sparing palms, soles and mucous membranes. We present a case of LWNH with delayed milestones and a history of two episodes of neonatal seizures.

DISCUSSION OF MANAGEMENT

German dermatologist Alfred Blaschko, in 1901, described a pattern of lines that is followed by a group of skin diseases.[2] The lines form a V-shape or fountain-like pattern over the back, S-shape or whorled pattern over the anterior and lateral aspects of the trunk, linear pattern over the extremities and waves on the vertex scalp. The lines believed to trace the migration of embryonic cells[3] are invisible under normal conditions, but becomes visible in some diseases. Happle hypothesized that genetic mosaicism, characterised by the presence of two distinct populations of melanocytes with differing abilities for pigment production is the most likely cause of these diseases.[4] Several case reports with genetic analysis have documented this to be the cause, most of them reporting trisomies of several different chromosomes.[5,6,7]

These groups of diseases, termed as Blaschkoid dyspigmentation[8] include Linear and Whorled Nevoid Hypermelanosis (LWNH), incontinentia pigmenti, linear epidermal nevus, hypomelanosis of Ito and Goltz syndrome etc.

Kaltar et al in 1988 reviewed case reports with similar characteristics but with different names and grouped them into a single entity of Linear and Whorled Nevoid Hypermelanosis (LWNH).[1] LWNH is characteristically distinguished from the rest by the absence of basal cell degeneration. No pigmentary incontinence in the dermis and no preceding inflammation or atrophy. Majority of the cases have only cutaneous manifestations that usually stabilise after two years of age and in some cases even start to resolve. Chemical peels and 2% hydroquinone is used in some cases. Multisystem involvements in genetic defects are not uncommon and extracutaneous manifestations have been reported in many of the cases of LWNH, neurological abnormalities being the most common.[9]

Our patient, along with LWNH, presented with delayed milestones. Parents also reported her having two episodes of seizures in the first month of life, for which no cause was identified. Neonatal seizures without any identifiable cause have been reported before.[10] Further workup is needed to identify the cause of seizures and developmental delay in such patients and to establish their relation with LWNH. Multisystem involvements in LWNH, especially neurological points us to consider novel paediatric management rather than purely dermatological, where in most of the cases it is left to resolve on its own. This traditional approach delays the diagnosis and proper management of the extracutaneous manifestations, if present. And many a times these are not linked to LWNH. This leads to their bad prognosis and poor quality of life.

Figure 1

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