ABSTRACT

Linear and whorled nevoid hypermelanosis (LWNH) is a rare skin condition characterized by hyperpigmented macules and patches along Blaschko’s lines. 45 cases have been reported, with one quarter of patients having associated extracutaneous findings. LWNH is a rare skin condition. 45 cases have been described. 1–6

REFERENCES


INTRODUCTION

Linear and whorled nevoid hypermelanosis (LWNH) is a rare disorder of hyperpigmentation that typically presents within a few weeks of birth. It was first described as a distinct disease by Kalter in 1988.1 The pigmentation occurs along Blaschko’s lines with a streaky or swirl like pattern and it is not preceded by other skin lesions.2 It is also referred to as pigmented mosaicism. Approximately one quarter of patients may have associated extracutaneous findings, which can include the CNS, musculoskeletal, ocular systems and rarely heart.3 Histology is nonspecific showing increased epidermal pigmentation in affected skin. Our case study describes a patient who presented at birth with unilateral lesions.

CASE SUMMARY

An 11 year old male presented for evaluation of a “dark rash” present on his neck, trunk and upper extremities since infancy according to his mother. The rash was not itchy or painful and the lesions were not changing in size or migrating. His mother stated that the lesions were initially lighter when they started, but have been this color for about the last several years. Son and mother denied any significant past medical history, allergies, medications or family history of skin disease. Physical exam revealed linear and whorled hyperpigmented patches present on the right neck, trunk and upper extremities (figures 1, 2, 3). A Wood's lamp examination accentuated the pigmentation, confirming the intraepidermal location of pigment deposits. A diagnosis of LWNH was made based on the constellation of historical and physical exam findings. The patient was counseled on the benign nature of LWNH and refused treatment.

DISCUSSION

LWNH is a rare skin condition. 45 cases have been reported to date. Typically lesions present within a few weeks of birth. The extent of hyperpigmentation increases during the first 2-3 years prior to stabilizing.2 Di Lernia (2007) in a retrospective case series of 16 unrelated children with LWNH reported that 10/16 cases were unilateral like ours. The unilateral cases did not have any associated abnormalities, while 1/6 of the bilateral cases had associated abnormalities.3 Clinical findings include reticulate hyperpigmented macules and patches that coalesce to form streaks and whorls along Blaschko lines. An S-shaped or whorled pattern presents over the anterior and lateral aspects of the trunk, a V-shaped pattern is noted over the spine and a linear arrangement over the extremities and genitalia. These patterns were also observed in our patient. Palmoplantar skin, mucosa and hairs are typically spared.3 Most cases appear to be sporadic, due to mosaicism. Mosaic trisomy 7, 14, 18, 20 and X chromosomal mosaicism have been reported. However familial cases have been described.4 Differential diagnoses include incontinentia pigmenti, McCune-Albright syndrome, hypomelanosis of Ito and linear atrophoderma of Moulin. The third stage of incontinentia pigmenti shows hyperpigmented streaks in a Blaschko-like pattern. McCune-Albright syndrome has extensive cafe au lait macules that tend to be larger with jagged borders and may also follow lines of Blaschko. Hypomelanosis of Ito presents with hypopigmentation along lines of Blaschko in a similar pattern to LWNH and in extensive cases determining whether the lesions are hypopigmented or hyperpigmented may be difficult. Atrophoderma of Moulin presents during childhood or adolescence with unilateral Blaschkooid hyperpigmented, slightly depressed patches.5 Reports on other cases have described unsuccessful treatments with chemical peels and hydroquinone. Laser treatment may be a viable option, but literature is lacking.6

LWNH is a benign disease that must be distinguished from other entities. Despite its benign nature, up to one quarter of patients can have associated systemic findings. This warrants a thorough review of systems and a complete physical. The musculoskeletal, neurological and ocular systems are most commonly affected. Congenital heart disease, including ventricular septal defect and tetralogy of Fallot have been rarely reported.

Figures 1, 2, 3: Hyperpigmented macules and patches following Blaschko’s lines.