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Cutaneous Mosaicism Along Blaschko Lines: A Case Series of Epidermal Verrucous Nevus and Linear Whorled Nevoid Hypermelanosis

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Abstract

Cutaneous mosaicisms are often seen along Blaschko lines. These lines represent ectodermal cell migration. Lesions derived from epidermal components typically follow Blaschko line patterns. Here, we present three cases of linear dermatoses along the Blaschko lines. Two cases of linear epidermal verrucous nevus and one case of linear and whorled nevoid hypermelanosis. Two cases are histologically confirmed. All three of our patients had normal development, and no systemic abnormalities were detected. Epidermal verrucous nevus and Linear whorled nevoid hypermelanosis are two conditions that arise from mosaicism, causing a clone of skin cells with an increased ability to produce pigment. Early clinical and histopathological diagnosis is essential. Most patients may seek treatment due to a poor cosmetic appearance.

Key words: Blaschko lines, Hypermelanosis, Mosaicism, Mutations; Somatic, Nevus; Epidermal, Skin diseases

Introduction

Several lines have been described in dermatology. Among these, Blaschko lines represent the developmental growth pattern of the skin and are not related to vascular, neural, or lymphatic structures.[1] These lines are usually not apparent under normal conditions but become prominent in disorders affecting skin pigmentation. They are arranged in V shaped lines patterns over upper back, S shaped patterns on the abdomen, inverted U-shaped patterns from the breast to upper arm and as perpendicular lines along the anterior and posterior aspects of the extremities. Here, we describe two such conditions presenting along a Blaschkoid distribution.

Case Report

Case 1: A 13-year-old boy came with complaints of asymptomatic multiple dark raised linear rough lesions noticed since birth. There was gradual increase in size and number of lesions in the past 1 year. Examination of eyes, oral cavity hair, teeth, and nails were normal, and systemic examination did not reveal any abnormalities. His growth and development were normal. Routine blood investigations, ECG, ECHO were normal. Family history was insignificant. On examination multiple hyperpigmented hyperkeratotic verrucous plaques were seen arranged in hourglass configuration over the face [Figure 1a], extending linearly from the xiphisternum to the right arm in an inverted u-shaped manner, lateralized pattern without crossing midline on the left side of the trunk [Figure 1b] and phylloid pattern over back [Figure 1c]. Clinical diagnosis of verrucous epidermal nevus was considered. Biopsy was taken which showed basket weave orthokertosis along with papillomatosis. Increased melanocytes with

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melanin incontinence with perivascular lymphocytic infiltrate were also seen in upper dermis [Figure 1d]. Based on clinical and histopathological changes diagnosis of linear epidermal verrucous nevus was made. Reassurance was given to his parents, and patient had been advised for laser therapy.

Case 2: A 56-year-old male came with complaints of asymptomatic dark brown linear discoloration over the chest and back for 15 years. Examination of eyes, hair, and teeth and other organ systems were normal. On examination, there were ill-defined brownish to black macules were seen extending from the xiphisternum to the upper extremities linearly on both sides [Figure 2a] and arranged in a V-shaped pattern over the back [Figure 2b]. Differential diagnosis of Linear and whorled nevoid hypermelanosis and Progressive cribriform and zosteriform hyperpigmentation were considered. Biopsy was taken from back lesion for histopathological examination. It showed orthokeratosis with increased pigmentation of basal keratinocytes and upper dermal oedema [Figure 2c]. Melanophages and inflammatory infiltrates were absent. Based on clinicopathological findings final diagnosis of linear and whorled nevoid hypermelanosis was made. Patient was counselled for laser therapyand was prescribed topical emollients.

Case 3: A 28-year-old male came with complaints of asymptomatic multiple dark raised rough lesions over face, neck, chest, back since birth. There was gradual increase in size and number of lesions.,. His development was normal. Family history was insignificant. Routine blood investigations were within normal limits. On examination, multiple well defined hyperpigmented hyperkeratotic verrucous plaques and papules arranged in phylloid pattern seen on neck extending on to right side of chest [Figure 3a] and in v – shaped manner seen on upper back [Figure 3b]. Biopsy could not be taken as the patient was not affordable Based on history and clinical examination diagnosis of epidermal verrucous nevus was given. Patient was reassured about benign condition of lesions and radiofrequency ablation with Nd-YAG laser was planned, however the patient was lost to follow – up.

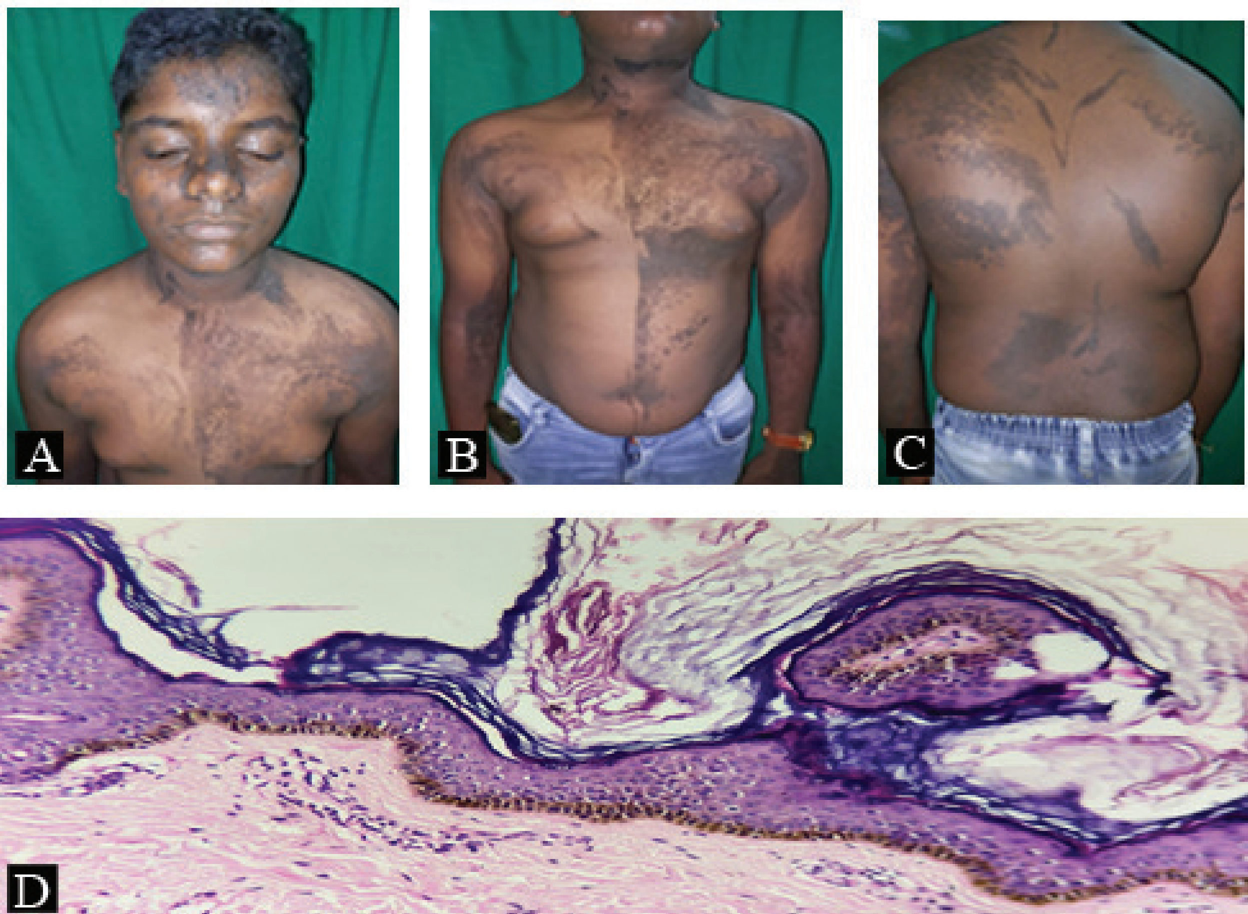

Figure 1 : (a) Hyperpigmented verrucous plaques and papules arranged along the Blaschko lines over face. (b,c) Hyperpigmented verrucous plaques arranged linearly in lateralised pattern and phylloid pattern on left side of chest, abdomen and back. D. Histopathological examination (40x) showing orthokeratosis, papillomatosis, increased pigmentation of basal layer and melanin incontinence.

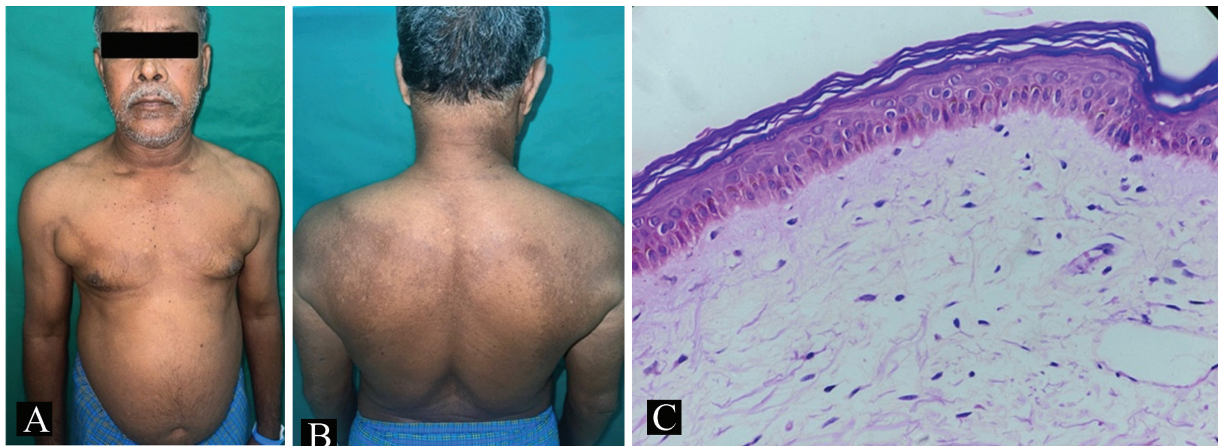

Figure 2: (a, b) Ill-defined brownish to black macules seen arranged linearly along the Blaschko lines over chest and back. (c) Histopathological examination (40x) showing orthokeratosis with increased pigmentation of basal layer.

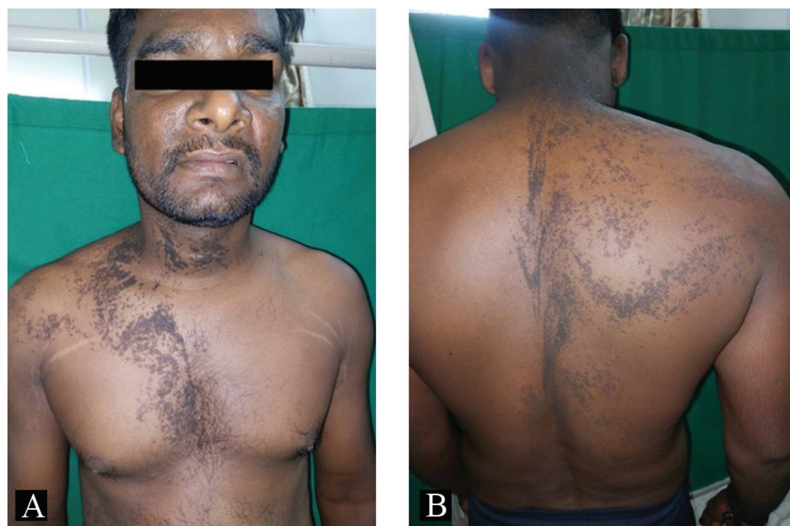

Figure 3: (a)Hyperpigmented verrucous papules arranged in phylloid pattern in face and neck. (b) multiple hyperpigmented papules arranged linearly along Blaschko lines on back.

Discussion

Different types of linear lesions which varies in morphology and histology can occur along Blaschko lines. These lines represent developmental pattern of skin. Many nevoid, congenital, and acquired disorders arise in linear pattern along Blasckho lines. Most of them arise due to cutaneous mosaicisms due to post zygotic mutations. They are two types of mosaicism such as functional or genomic mosaicisms.[2] Happle further described other patterns through which cutaneous mosaicism present like phylloid, block pattern, large patches without midline separation and lateralization pattern.[3] Linear lesions in dermatology not only follow Blaschko lines but can also be distributed along blood vessels, lymphatics, dermatomal, Koebner's phenomenon, infestations.[4]

Verrucous epidermal nevi are hamartomas mostly derived from keratinocytes but can also arise from hair follicles, sebaceous or sweat glands. Epidermal nevi are mainly caused by activating FGFR3 mutations in the human epidermis or secondary to a post zygotic mutations.[5] The lesions are present at birth. Some case reports of adult-onset epidermal nevus have been reported. Inflammatory component is known as Inflammatory linear epidermal verrucous nevus (ILVEN) characterized by itchy verrucous papules and papules. Epidermal nevi are mostly benign but in rare cases they may undergo malignant transformation.[6] Most of them can be diagnosed clinically. Biopsy and histopathological examination are required to differentiate it from other linear lesions like linear psoriasis, linear lichen planus, linear porokeratosis, Darier's disease. Patients with extensive lesions should be evaluated for epidermal nevus syndrome (ENS). It is characterized by involvement of ocular, cerebral, skeletal, renal, cardiovascular abnormalities.[7] No such abnormalities have been seen in our cases. Our patient had attained all developmental milestones and has normal IQ. Ophthalmological examination was done in

our patient which showed 6/6 vision. Many treatment options are available ranging from topical creams, keratolytics, laser therapies to surgical excision.

Linear and whorled nevoid hyper melanosis is a rare disorder of pigmentation which is characterized macular pigmentation in a linear or whorled streaky configuration. It presents at birth or within first 2 years of life.[8] Our patient had onset of lesions early in life which progressed and stabilized after some time. They may arise due to somatic mosaicism or chromosomal abnormality like trisomy 7,14,18 and 20.[9] Developmental delay and birth defects like skeletal anomalies, cardiac diseases, deafness can be associated with LWNH. However no systemic abnormalities were seen in our patient. Differential diagnosis for this condition includes progressive cribriform and zosteriform pigmentation, incontinentia pigmenti, hypomelanosis of ito, linear epidermal nevus. Linear whorled nevoid hypermelanosis and progressive cribriform and zosteriform pigmentation are part of same spectrum. The latter typically has a late onset and not associated with systemic abnormalities ,mostly due to somatic mosaicism.[10] Treatment options available for this condition are topical depigmenting creams, chemical peels, lasers.

Both conditions can be diagnosed clinically. Biopsy is required to differentiate them from other congenital and acquired conditions. Histopathologically, these two conditions can be differentiated with presence of acanthosis, papillomatosis, melanophages and pigmentary incontinence seen in linear epidermal verrucous nevi. While these changes are mostly absent in linear and whorled nevoid hypermelanosis. The latter is characterized by an increase in basal layer pigmentation without melanophages or pigmentary incontinence. Progressive cribriform zosteriform hyperpigmentation is considered as a localized spectrum of LWNH. Apart from histopathology dermoscopic examination can help in diagnosis of pigmentary lesions.

Treatment of these conditions is quite challenging. Various therapeutic options like topical keratolytics, hydroquinone, retinoids, cryotherapy, 532 Q-Switched Nd-YAG laser, 755nm QS Alexandrite laser, electrofulguration, and chemical peels may be used with variable clinical outcomes. In extensive cases multidisciplinary evaluation has to be done to rule out neurological, ocular, skeletal and cardiovascular complications.[11]

Conclusion

Linear epidermal verrucous nevus and linear whorled nevoid hypermelanosis represent a spectrum of cutaneous mosaicisms. Recent advances in genomics have led to identification of genetic mutations of these diseases. These pigmentary conditions are quite resistant to treatment. Most of people will seek treatment due to cosmetic concerns. Genetic counselling should be offered to parents of affected children.

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