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Clinico-pathological correlation of linear dermatoses along the lines of Blaschko: An observational study

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ABSTRACT

Background: Several epidemiological studies have described linear dermatoses; however, only few studies have correlated linear dermatoses along Blaschko's lines with the histopathological findings. The aims of this study were to investigate the clinical patterns of various linear dermatological lesions along Blaschko's lines and investigate the correlations between these dermatological lesions and their histopathological features.

Materials and Methods: Patients who attended our dermatology out-patient department with linear dermatoses along the Blaschko's lines were included in the study. Detailed history was obtained, clinical examination was performed, and a final provisional clinical diagnosis was noted. Subsequently, biopsy of the lesions was performed for histopathological examination. Of 62 patients who presented with linear lesions along the Blaschko's lines, 45 patients underwent biopsy and histopathological examination.

Results: Of 45 patients who underwent biopsy, clinico-pathological correlation was observed in 100% of those diagnosed with lichen striatus and linear morphea, 89% of those diagnosed with hypomelanosis of Ito, 80% of those diagnosed with linear epidermal nevus, 75% of those diagnosed with lichen planus, and 67% of those diagnosed with nevus depigmentosus.

Conclusions The importance of correlation of linear lesions with their histopathological features in dermatology cannot be over emphasized. However, 100% correlation may be wishful thinking, and a correlation can help choose the appropriate line of management. Our results highlight this discrepancy and add to the knowledge on linear dermatosis.

1. Introduction

The concept of Blaschko's lines were first introduced by Alfred Blaschko in 1901.[1] He originally described these lines as "a system of lines on the human skin that linear nevi and dermatoses follow".[1] Linear cutaneous lesions are those that occur along imaginary skin lines like Blaschko's lines, lymphatics, and blood vessels or those with dermatomal distributions. These may be primary lesions or secondary lesions due to autoinoculation or Koebner's phenomenon.[2] Unlike other dermatoses, linear lesions along Blaschko's lines from a V-shape over the spine, S-shape on the lateral and anterior aspects of the trunk, and an inverted U-shape from the breast area onto the upper arm. They follow perpendicularly longitudinal direction on the extremities and whorls on the abdomen.[3] Two mechanisms proposed to explain Blaschko's lines include mosaicism and chimerism. A number of congenital and acquired conditions follow these lines. Almost all epidermal nevi follow Blaschko's lines. Lesions in X-linked disorders, such as incontinentia pigmenti (IP) and Goltz syndrome as well as chromosomal disorders, such

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as hypomelanosis of Ito also follow these lines. Such as pattern of distribution aids in their diagnosis in newborns, infants, and adults. Other nevoid epidermal disorders, such as linear lichen planus, nevoid psoriasis, Darier's disease, and Hailey-Hailey disease, linear porokeratosis, and blaschkitis are some of the acquired diseases that occur along the Blaschko's lines. The occurrence of these acquired lesions has special significance in terms of the epidemiology, clinical presentation, treatment, and prognosis. Several studies have investigated individual diseases along the Blaschko's lines. However, no studies have included Blaschko's lines as the central focus. This study is an attempt in that direction.

The importance of linear lesions in dermatology cannot be over emphasized. Their distribution also helps in elucidating the pathogenesis and prognosis. These lesions guide dermatologists in selecting the appropriate line of management. Even though the pattern of distribution of linear dermatoses acts as one of the diagnostic tools, their correlation with associated clinical syndromes and histopathology is needed frequently for better diagnostic accuracy. Several publications have described various individual linear dermatological lesions and their histopathological correlation in small numbers of patients. There are very few large-scale studies that have correlated various linear dermatological lesions with their histopathological features and associated syndromes. Therefore, the aim of this cross-sectional study was to investigate the correlation between dermatological lesions and their histopathological features to aid in diagnostic accuracy and algorithms, management, and improved therapeutic outcomes.

2. Materials and Methods

In this cross-sectional study, all patients with clinical evidence of lesions along the Blaschko's lines who attended our out-patient department between December 2016 and June 2018 were screened. The only inclusion criterion was patients with linear dermatosis along the Blaschko's lines who provided informed consent for participation in this study and undergo biopsy. This study was approved by the Institutional Ethics Committee of our institute (Approval No.). All procedures were performed in accordance with the Helsinki Declaration of 1975, as revised in 2000.

Patient details were collected in a predesigned proforma and included age, sex, occupation, and address. Detailed history was obtained with particular emphasis on the onset, duration, course of each lesion, mode of spread, progression, and any associated systemic findings. Family history regarding consanguinity in parents and similar history in other family members was obtained. After thorough general and systemic examinations to identify any systemic findings, other investigations were performed, as necessary in each patient.

For each lesion, the site, shape, size, surface, color, and distribution pattern were noted. Additionally, the mucosal surfaces, scalp and hair, nails, palms and soles, and genitals were evaluated. After diagnosing linear dermatoses along Blaschko lines, photographs of all lesions were obtained and punch biopsy was performed. The histopathological features of the biopsy specimen were correlated with the clinical findings.

2.1. Statistical analysis

Categorical data are presented as frequencies and proportions. The data were analyzed using SPSS v22 (IBM Inc., Armonk, NY, USA).

3. Results

Age distribution

The characteristics of the patients and the lesions are summarized in Table 1. A majority of patients in this study were younger than 20 years of age; furthermore, 55%–60% were younger than 10 years of age. The second peak was noted in pre-marital patients, which highlights the patients' anxiety to understand such disorders prior to marriage. Only potentially serious and extensive disorders like IP and epidermal nevus syndromes presented during infancy. There were no predominant patterns noted in terms of the sex, socioeconomic status, and educational status of these patients. In this study, 6.5% of patients had a history of consanguinity. These disorders are usually sporadic, which could explain the low rate of consanguinity.

3.1. Evolution of lesions

Details of the lesions are summarized in Table 2. Approximately two-thirds of the patients in our study reported proximal-to-distal evolution of the lesions. This is in accordance with the embryological migration of the cutaneous cells in the ectoderm, neuroectoderm, and mesoderm from the dorsal midline along the lateral walls to reach the corresponding ventral midline. It also highlights the natural contours of Blaschko's lines over the skin. The lesions were similarly distributed on the right and left sides, and only 6.4% of patients had bilateral involvement.

3.2. Symptoms and previous treatment

A majority (75%) of patients in our study were asymptomatic at presentation (Table 2). They usually presented because of anxiety and cosmetic concerns. However, itching was the primary complaint in symptomatic patients. Additionally, a majority of patients (62%) had not received any oral or topical treatments previously. Organ involvement was not seen in our study

3.3. Type and side of involvement

The narrow band pattern (69%) was commonest distribution followed by the broad band and checkerboard patterns. The commonest side of involvement was the left side (48%) followed by the right side (45%) and both sides (4%). Lower limbs were the commonest sites (33.9% on thighs; 29% on legs) involved followed by the head and neck (27.4%), back (27.4%), arms (27.4%), and forearms (22.6%) with infrequent involvement of the hands (8.1%), feet (9.7%), and chest (2.6%). The palms and soles were completely spared. Genital lesions, which have been well-described in the literature, were not seen in our study.

3.4. Clinical diagnosis

The clinical diagnoses in the patients who underwent biopsy are summarized in Table 3; similarly, the clinical diagnoses in those who did not undergo biopsy are summarized in Table 4 . Lichen striatus was the commonest dermatosis followed by hypomelanosis of Ito, lichen planus, linear epidermal nevus, nevus depigmentosus, and linear morphea. These six linear lesions constituted approximately 80% of cases. Multifactorial inflammatory dermatoses were commoner than genodermatoses.

3.5. Clinico-pathological correlation

The correlation rate between the clinical diagnoses and histopathological features in patients who underwent biopsy are summarized in Table 5. Of 62 patients, 45 (72.6%) patients underwent punch biopsy. Overall, clinic-pathological correlation was observed in 84.4% of them. Of eight patients who were clinically diagnosed with linear lichen planus, correlation with histopathological features was observed in six patients (75%). The remaining two patients had histopathological features of lichen sclerosis et atrophicus and lichen striatus, respectively. The high rate of clinico-pathological discrepancy in our study might be due to atypical clinical features at presentation compared with the classical clinical cases in the a previous study.All patients diagnosed with lichen striatus had histopathological features of lichen striatus. Previously, Gianotti et al. had reported a correlation of 50% in lichen striatus.[4]The high rate of our clinic-histopathological correlation might be due to common clinical presentation of these linear lesions and predominant involvement of children, which usually does not warrant investigations to confirm the diagnosis for a dermatologist. Of five patients who were diagnosed with linear epidermal nevus, four patients (80%) had consistent histopathological features, while one patient (20%) had histopathological features of nevus sebaceous. Of three patients who were diagnosed with nevus depigmentosus, two patients (66%) had consistent histopathological features. Of nine patients diagnosed with hypomelanosis of Ito, eight patients (88.8%) had histopathological features of hypomelanosis of Ito, while one patient (11.2%) had clinico-pathological discrepancy. The high accuracy of clinical diagnosis of hypomelanosis of Ito might be due to the characteristic history and clinical features. Of four patients who were diagnosed with linear morphea, all four patients (100%) had histopathological features of morphea. In patients with various other clinical diagnoses, clinico-pathological correlation was noted in patients with IP, linear porokeratosis, inflammatory linear verrucous epidermal nevus (ILVEN), and linear vitiligo. However, clinico-pathological discrepancy was noted in two patients who were diagnosed with linear pityriasis rosea (histopathological features of lichen planus) and linear sebaceous horn (histopathological features of epidermoid cyst).

In our study, 17 patients refused to undergo biopsy due to various reasons like anxiety toward invasive procedures, asymptomatic lesions, non-progression of lesions for several years, and limited therapeutic options even if the diagnosis is confirmed using histopathology.

4. Discussion

Our present study was the first of its kind in north Karnataka and attempted to highlight the profiles of patients with linear dermatosis along the Blaschko's lines.

Blaschko's lines, also known as cutaneous lines of embryogenesis,[3] represent patterns of several nevoid and acquired skin diseases on the human skin and mucosae.[2] These lines are characteristic of the mosaicism of the epidermis and probably represent the routes of ectodermal cell migration from the neural crest. These lines do not correspond to other patterns, such as Langer's lines of cleavage,[5] Voigt's lines,[6] embryonic clefts,[7] pigmentary demarcation lines,[8] or the lines of lymphatic drainage, blood supply, or neural elements.[9]

Five main types of Blaschko's lines have been described. Type 1a includes narrow bands while type 1b includes broad bands of these lines. Type 2 includes a checkerboard pattern, a flag-like pattern with a strict midline separation. Type 3 includes a phylloid pattern in which there are multiple leaf-like or oblong macules reminiscent of the floral ornaments of nouveau style of art. Clinical example is novel neuro- cutaneous syndrome in the form of phylloid hypomelanosis. Type 4 includes a patchy pattern without midline separation. Type 5 includes a clear midline demarcation and is classically seen in congenital hemidysplasia with ichthyosiform erythroderma and limb defects (CHILD) syndrome.

The embryological basis for the distribution pattern of these lines remains an enigma. Jackson suggested that these lines are determined not only by epidermal tissues and structures but also dermal tissues as well. Stretching of the skin[10] during embryogenesis, and an inborn error of morphogenesis due to a single mutant gene[11]have

Table 1: Characteristics of patients and lesions along Blaschko's lines.

		Count	%
Age, years	<10	20	32.3%
	11–20	19	30.6%
	21–30	8	12.9%
	31–40	6	9.7%
	41–50	3	4.8%
	51–60	4	6.5%
	>60	2	3.2%
Sex	Female	37	59.7%
	Male	25	40.3%
Consanguineous marriage	Present	4	6.5%
Family History	Present	0	0%
Duration of lesions	Since birth	22	35.5%
	< 6 months	25	40.3%
	7–12 months	5	8.1%
	1 to 5 years	6	9.7%
	> 5 years	4	6.5%

Table 2: Details of the lesions in patients with linear dermatoses along Blaschko's lines.

		Count	%
Symptoms	Itching	16	25.8%
	None	38	61.3%
Treatment	Topical Application	20	32.3%
	Topical and Oral treatment	4	6.4%
	Broad band	18	29.0%
Pattern	Checkerboard	1	1.6%
	Narrow band	43	69.4%
	Left side	30	48.4%
Side	Right side	28	45.2%
	Both sides	4	6.4%
	Proximal to distal	38	61%
	Static	13	21%
Progression of lesions	Distal to proximal	9	14.5%
	Extending	1	1.6%
	Back to front	1	1.6%
	Head and neck	17	27.4%
	Chest	14	22.6%
	Abdomen and pelvis	10	16.1%
	Back	17	27.4%
	Thigh	21	33.9%
	Leg	18	29.0%
Site	Arm	17	27.4%
	Forearm	14	22.6%
	Hands	5	8.1%
	Palms and soles	0	0%
	Genitalia	0	0%
	Feet	6	9.7%

Table 3: Clinical diagnoses in patients with linear dermatoses along Blaschko's lines who underwent biopsy.

Nature of the generalized condition	Linear condition	Number of patients (n=62)	%
X-linked dominant single-gene disorder	Incontinentia pigmenti	1	1.6%
Autosomal dominant single-gene disorder	Linear porokeratosis	2	3.2%
	Linear epidermal nevus	7	11.2%
	Lichen striatus	15	24.1%
	Linear lichen planus	9	14.5%
Multifactorial inflammatory disorder	Linear morphea	4	6.4%
	Segmental vitiligo	2	3.2%
	Linear pityriasis rosacea	1	1.6%
	Linear psoriasis	1	1.6%
	Linear sebaceous horn	1	1.6%
Presumed autosomal dominant lethal disorder rescued by mosaicism	ILVEN	1	1.6%
	Linear comedonicus	2	3.2%
	Hypomelanosis of Ito	12	19.3%
Chromosomal anomalies	Nevus depigmentosus	3	4.8%
	Linear and whorled nevoid hypermelanosis	1	1.6%

Table 4: Clinical diagnoses in in patients with linear dermatoses along Blaschko's lines who did not undergo biopsy.

Diagnosis	Number of patients (n=17)
Lichen striatus	5
Hypomelanosis of Ito	3
Linear epidermal nevus	2
Linear comedonicus	2
Linear lichen planus	1
Linear porokeratosis	1
Linear psoriasis	1
Segmental vitiligo	1
Linear & whorled nevoid hypermelanosis	1

Table 5: Correlation rate between the clinical diagnosis and histopathological features in patients with linear dermatoses along Blaschko's lines.

	Count	%
Lichen planus	6/8	75%
Lichen striatus	10/10	100%
Linear epidermal nevus	4/5	80%
Nevus depigmentosus	2/3	66.6%
Hypomelanosis of Ito	8/9	88.8%
Linear Morphea	4/4	100%
Overall	39/45	86.6%
Others	4/6	66.6%

also been proposed as the underlying mechanisms. Their characteristic distribution explains the presence of two different clones of cells during early embryogenesis. Therefore, possibly, these lines represent a form of human 'mosaicism' where two or more genetically distinct cell populations are present in an individual derived from a single zygote. These different clones may be due to lyonization (random inactivation of one of the two X chromosomes in all cells in females), post-zygotic somatic mutations during early embryogenesis, half-chromatid mutation before fertilization,[1,10] or chromosomal non-disjunction or chimerism. These lines probably represent boundaries between populations of normal and mutant cells and may represent the dorso-ventral outgrowth of mutant clones. An explanation for the fountain-like pattern of Blaschko's lines on the back is that transversal proliferation of precursor cells starts from the primitive streak but is interfered with the longitudinal growth and increasing flexion of the embryo. However, no single theory clearly explains the origin and localization of

Blaschko's linesbecause cutaneous mosaicism need not always follow Blaschko's lines.[9]The earlier the mutation, the more widely dispersed and more intimately mixed are the mosaic clones and, consequently, longer are the lines of migration. Its timing in relation to the process of lyonization, lateralization, and organogenesis will also influence the pattern. The pattern of cutaneous mosaicism also varies according to the cell-type that is affected.[12] In females, synchronous lyonization occurs in all cells approximately during the 1000-cell stage; therefore, the two clones are intertwined from the beginning. Consequently, Blaschko's lines in X-linked disorders are typically narrow and numerous. An important exception is the CHILD syndrome, which characteristically includes large unilateral blocks of abnormal skin, possibly because the mosaicism may be due to later somatic mutations on the X chromosome and not due to lyonization. In males, X-linked dominant disorders are usually fatal, sometimes even in utero. This lethal phenotype can only be rescued by mosaicism. Not all X-linked skin disorders follow Blaschko's lines. For example, women who are heterozygous for Fabry's disease, Wiskott–Aldrich syndrome, and chronic granulomatous disease demonstrate either no lesions or scattered lesions in a non-linear pattern. A possible explanation is that these abnormalities are systemic, and a local deficiency in parts of the skin is corrected from elsewhere, usually the bone marrow.[12] Some genes on the X chromosome do not undergo random inactivation.[13] For example, the gene on the short arm of the X chromosome that encodes steroid sulfatase escapes inactivation. This is the why the epidermis of female carriers of X-linked recessive ichthyosis lacks a mosaic pattern of scaling.[10]

IP is a rare genodermatosis with X-linked dominant inheritance. Most of the patients in this genodermatosis (about 96%–97%) are females. The linear skin lesions reflect mosaicism secondary to X inactivation. IP type 1 (sporadic form) was differentiated from type 2 (inherited form). Patients with type 1 IP had chromosomal (X/autosomal) translocations, often at Xp11, in association with pigmentary abnormalities.[12] IP type 2 represents the X-linked dominant disorder caused by mutations in the NEMO gene.[14] Additional manifestations of IP include linear absence of hair and sweat glands, nail dystrophy and nail tumors, asymmetric breast development, supernumerary nipples, missing and conical teeth, microphthalmia, retinal vascular anomalies, cataracts, mental retardation, seizures, spastic hemiplegia, diplegia, tetraplegia, optic atrophy, skull anomalies, scoliosis and pulmonary hypertension. In a retrospective study of 40 patients of IP by Razda et al., it was found that during the neonatal period, erythema, vesicles and hyperkeratotic lesions were rarely absent in the patients with IP. Ocular and neurological abnormalities were frequent (20% and 30%, respectively) but rarely severe (8% and 7.5%, respectively).[15]

Several inflammatory skin conditions may occasionally be distributed along Blaschko's lines. They usually appear years after birth, thus suggesting environmental contributions. Several conditions demonstrate Koebner's phenomenon as well. The linear form of these disorders may therefore reflect mosaicism for a susceptibility mutation.[12] This loss of heterozygosity may occur from a mutation, deletion, or DNA recombination and lead to the formation of a keratinocyte clone that is more susceptible to development of the skin disease. This concept was first introduced in 1991 to describe linear psoriasis and, since then, has been applied to segmental forms of atopic dermatitis, lichen planus, erythema multiforme, pemphigus vulgaris, vitiligo, and granuloma annulare.Segmental disease may be superimposed on non-segmental lesions, with the segmental lesions usually being more difficult to treat.[16]

Lichen striatus is an asymptomatic, uncommon, self-limited, linear dermatosis of unknown etiology that generally affects children between the ages of 4 months and 15 years. In other studies, reported male:female ratio has varied from 1:1.6 to 1:2.49. Environmental agents, particularly viruses, have been implicated, given the predominance of the disorder in young children and its seasonal variation. However, a viral association has not been proven via serologic testing or cultures. Lichen striatus may represent a manifestation of an atopic diathesis. Patrizi et al. reported an association of lichen striatus with atopic dermatitis in 70/115 children.[17] In another series, 80% of patients had atopy.[18]

Scattered linear lesions often occur in patients with lichen planus (LP) and are a result of scratching and the Koebner phenomenon.[10] Less commonly, unilateral streaks or bands of LP are seen that are longer and wider than the trauma- induced lesions seen along the Blaschko's lines. Some authors consider linear LP as an intermediate entity between LP and lichen striatus. Linear LP accounts for less than 0.2% of all patients with LP,[16] except in Japan, where up to 10% of reported cases are linear.[19] The linear variant of LP can be persistent, but occasionally may resolve with post-inflammatory hyperpigmentation.Linear LP lesions are usually only a few centimeters in length, but long, narrow linear lesions extending along the whole length of a limb may occur. If the LP lesions extend to the end of a digit, the nail is often affected.

Vitiligo is a multifactorial disorder that occasionally occurs in a linear distribution. The lesions tend to be broad bands, patches or blocks, corresponding more to dermatomes than Blaschko's lines, perhaps in keeping with a neuronal pathogenesis. The neuronal abnormality could be mosaic or, alternatively, there could be a clonal susceptibility of melanocytes to neuronal or other influences. Compared with symmetric vitiligo, the linear type is earlier in onset, less likely to spread to other areas

of the body, and less frequently associated with other autoimmune diseases.[12]

Linear morphea occurs as a linear band, usually with a single unilateral lesion. The lower extremities are most often involved, followed, in frequency of occurrence, by the upper extremities, frontal area of the head, and anterior thorax.[20] The female to-male ratio is 4:1. Linear scleroderma tends to affect children and adolescents. Whether linear morphea follows Blaschko's lines is controversial.In a detailed review, Bolognia et al could not find a single case of linear morphea following Blaschko's lines.[10] In many cases of linear morphea, it is not clear whether the distribution is segmental, dermatomal or following Blaschko'slines.[10]

ILVEN is probably due to mosaicism for a dominant mutation, as yet unidentified, which would be lethal if it affected all cells and is 'rescued' by mosaicism.[21] Although the lesions may be present at birth, the majority of ILVEN appear during infancy and childhood. They are characterized by pruritus, which may be intense. The lesions are linear, most commonly on a limb, and comprise eczematous or psoriasiform papules. There is a slight preference for the left side.[22] Occasionally, ILVEN is bilateral and widespread. Nail dystrophy may occur when the nail fold is affected. ILVEN can be distinguished from true nevoid psoriasis by pruritus and lack of response to anti-psoriatic treatments.[23]

The term hypomelanosis of Ito is a description rather than a diagnosis. The phenotype of this multisystem disorder is highly variable, except in the skin, where it always presents as hypopigmentation following Blaschko's lines.[12] HI is the third most common neurocutaneous disorder, after neurofibromatosis and tuberous sclerosis.[24] It is diagnosed in 1 in 8000–10,000 general pediatric patients and 1 in 790 pediatric dermatology patients.[25] There have been a few reports of familial HI, but the majority of cases are sporadic. Chromosomal mosaicism can be identified in the blood in about a third of patients. The mosaic karyotype anomalies reported include a variety of defects of chromosome structure and number and can affect autosomes or X chromosomes.[12] However, no consensus exists about the identity of the HI gene. In contrast to the variable systemic manifestations, the consistency of the skin appearance is remarkable. Clinically, lesions appear at birth or infancy as asymmetric, whorled or streaked lesions in a marble cake pattern along the lines of Blaschko, occurring on any part of the body. The hypopigmented streaks can be unilateral or bilateral. Less often the distribution is patchy sparing the palms and soles. Lesions usually appear by 1 year of age in 77% of patients and an initial increase in extent of involvement can be followed by a gradual repigmentation.[26] There is also evidence for somatic mosaicism in the eye in which striated and mottled hypopigmentation of the fundus and iris can occur in a pattern similar to that in X- linked ocular albinism.[27]

Congenital abnormalities, mental retardation, and seizures are the most commonly associated conditions, as reported in the medical literature. Cerebral malformations may occur, and visual impairment may be cortical in nature.[28] Glomerulocystic kidney disease has been reported.[29]Other anomalies include cleft palate, hemi hypertrophy of limbs, hand and/or foot abnormalities, nail abnormalities, hypotonia, teeth abnormalities, hair anomalies, face and/or skull anomalies. The associated anomaly rate is approximately 30%.[12]

Nevus depigmentosis is a localized area of depigmented skin and occurs in 1 in 50–75 individuals.[12] The name is a misnomer as the areas of leukoderma are actually hypomelanotic not amelanotic. This circumscribed area of hypopigmentation is congenital but may not be apparent at birth. There are three clinical variants- isolated, segmental and systematized.The commonest variant is the single, isolated circumscribed, rounded lesion. Segmental and systematized forms are very rare and may resemble hypomelanosis of Ito. Block-like areas of hypopigmentation that respect the midline can also be seen.Most lesions measure a few centimeters in diameter and have irregular but well-defined borders. Hairs within the hypopigmented macules are usually depigmented.The cutaneous findings in nevus depigmentosus are identical to those in HI but are fixed and usually more limited in distribution; and more importantly extracutaneous associations are lacking.[12] However, neurological abnormalities such as seizures have been rarely reported.[30]

Kalter et al.[31]characterized linear and whorled naevoid hypermelanosis as follows: (1) onset within a few weeks of birth and then progressing for 1–2 years before stabilizing; (2) linear and whorled nevoid hyperpigmentation following Blaschko's lines without preceding bullae or verrucae; (3) hyperpigmented areas with increased pigmentation of the basal layer and prominence of melanocytes without incontinence of pigment; (4) sporadic male and female incidence; (5) sparing of mucous membranes, eyes, palms and soles; and (6) possible associations with congenital anomalies. The exact pathogenesis is not known. Somatic mosaicism that develops during embryogenesis appears to be the underlying etiology.[12] The trunk, extremities, neck, face and genitalia are the typical sites affected.[32–34] The pigmentation tends to persist indefinitely.[12] Associated systemic abnormalities include atrial septal defect, dextrocardia, deafness and neurological and musculoskeletal defects.[10]

This study has a few limitations. Genetic work up was not done in this study due to a lack of facilities at our center. Additionally, only a small number of patients were included who underwent biopsy; therefore, the generalizability of the results may be limited.

In conclusion, the importance of correlation of linear lesions with their histopathological features in dermatology

cannot be over emphasized. Accurate and appropriate correlation can help in choosing the appropriate line of management. Our results highlight this discrepancy and add to the knowledge base on linear dermatoses.

5. Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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None.

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