

Original Research Article

Study on linear dermatoses

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ABSTRACT

Background: The objective of the study was to study the incidence, age and sex distribution, association, distribution, clinical presentation, histopathological correlation of linear dermatoses at the skin Outpatient Department.

Methods: A prospective study was conducted over a period of 1 year in 90 patients, attending the Department of Dermatology, at a tertiary care hospital, Chennai. Data collection, clinical examination and skin biopsy were recorded and analyzed statistically. Inclusion criteria were all patient with linear dermatoses. Exclusion criteria were Koebner phenomenon.

Results: The incidence of linear dermatoses in our outpatient department- 0.2% per year. Among the linear dermatoses, lichen striatus was found to be the most common. The other dermatoses following Blaschko's lines, in the descending order of frequency seen in this study were linear epidermal nevus, linear lichen planus, linear morphea, inflammatory linear verrucous epidermal nevus, segmental vitiligo, hypomelanosis of ito, linear lichenoid dermatitis, linear psoriasis, segmental neurofibromatosis, linear whorled nevoid hypermelanosis, incontinentia pigmenti, nevus depigmentosus, linear porokeratosis. Female preponderance was noted. Majority of patients showed unilateral distribution mostly on the extremities.

Conclusions: Most of the linear lesions are arranged along Blaschko's line. The importance of histopathological correlation is obvious. Cases which were clinically diagnosed as lichen striatus, showed histopathological features of psoriasis and linear epidermal verrucous nevus. One case diagnosed clinically as epidermal nevus was found to be super imposed by psoriasis histologically. Another case clinically diagnosed as linear psoriasis clinically, was found to be linear porokeratosis on histopathology. Very few associations were noted.

Keywords: Blaschko's lines, Lichen striatus, Lichen planus

INTRODUCTION

The term mosaicism in genetics indicates persons with cells of different genotypes. Genetic mosaicism classically appears as Blaschko's lines in the skin. The pattern may vary according to timing and cell type of mosaicism. Dr. Alfred Blaschko, dermatologist from Berlin, in 1901 stated that epidermal nevi, and some other conditions now known to be mosaic, follow characteristic

lines and whorls on the skin.¹ Lyonization, somatic mutation, half-chromatid mutation, chromosomal non-disjunction or chimerism can result in Blaschko's lines. These lines are comprehensively reviewed by Bologna et al.² Blaschko's lines represents boundaries between populations of mutant and normal cells, and this was first expressed by Douglas Montgomery, who studied several cases of extensive linear epidermal nevi.³ The now widely held idea that Blaschko's lines reflect embryonic

cell migration, but his hypothesis has never been proven. Few drawbacks of these lines were that, Blaschko's reproduced diagram neglected the scalp, mid-face, lateral neck, and genital area and cutaneous mosaicism does not follow Blaschko's lines always. Now the patterns in above areas, as well as the distribution of lesions in the teeth, eyes and oral cavity are established.^{4,5}

The mechanisms or anatomical factors dictating the linearity are given in Table 1.

Most of the linear lesions follow the Blaschko's lines. Patients with linear lesions attending the dermatology outpatient department comprise our study group.

Table 1: Anatomical and causative factors in linear lesions.

Determinant of pattern	Examples
Blood vessels	Thrombophlebitis, monod's disease
Lymphatics	Lymphangitis, sporotrichosis
Dermatome	Herpes zoster, zosteriform naevus, zosteriform darier's disease
Nerve trunks	Leprosy
Developmental, Baschko lines	Pigmentary demarcation line, linea nigra, epidermal naevi, incontinentia pigmenti, hypomelanosis of ito, lichen striatus, linear psoriasis, linear lichen planus
Skin stretching	Striae
Infestation	Scabies, larva migrans
External factors	Phytophotodermatitis, caustics, burns, trauma
Other determinants	Linear scleroderma, senear-caro ridge, dermatomyositis, interstitial granulomatous dermatitis, flagellate pigmentation due to bleomycin.

Aims of study

To study the incidence, various clinical presentation, age and sex incidence, distribution, clinico histopathological correlation, association, of various linear dermatoses.

METHODS

Study population

It is a prospective cohort study done at government general hospital Chennai, Tamil Nadu, India, during the period August 2013 to September 2014. 90 cases of linear dermatoses were included in the study from the patients attending the skin department, government general hospital.

A detailed history including the family history, marital status, trauma, symptoms, duration, distribution, morphology, nail changes were obtained. Skin biopsy along with necessary investigations was done. All the patients were categorized according to their clinical features.

Sample and diagnostic procedure

90 Patients with linear dermatoses included in the study after excluding linear lesion due to koebnerization. All the cases were examined clinically and most of cases underwent biopsy for histological correlation.

CBC, platelet count, urine examination, RFT, LFT, VDRL, HBsAg, and CXR were done. ANA titre for linear morphoea was done. Great care was taken to find out associated skin disorders.

Statistical analysis

The data analysis was performed using descriptive statistics including median, frequency and frequency percentage. Comparisons are made using chi square test using standard equates. Results were reported with $p \leq 0.05$ as the accept level of significance.

RESULTS

Total cases of linear dermatoses in our study was 90 among 44517 new cases attending outpatient, forming incidence of 0.2%. Lichen striatus was seen in 28% followed by linear epidermal naevus in 21%, linear lichen planus in 20% linear morphoea in 10% and others (21%). 68 cases were asymptomatic and reported for cosmetic reason. Intense itching was the main reason to bring the lichen planus and few cases of lichen striatus.

Age and sex distribution of patients with linear dermatoses in this study is shown in Table 2.

The lesions were observed most commonly in the age group between 1 and 30 years, forming male to female ratio of 10:11. 92.3% cases showed unilateral distribution and the remaining 7.7% showed bilateral distribution. 73% cases had lesions mainly over the extremities corresponding to Blaschko's line.

Lichen striatus

25 presented with Lichen striatus in the age group between 11 months and 45 years and forming male to female ratio of 11:14.

Nineteen patients had asymptomatic hypopigmented macules and 4 patients had brown coloured macules and papules and another 2 patients had skin coloured tiny papules distributed mainly over the extremities (Figure 1). Multiple site of distribution was noted in 5 patients on same side. The duration of the lesions ranged from 4 months to 4 years, with an average of 12 months. 4 patients had second and third degree consanguinity and none of the family members had similar problems. Biopsy was done for 15 cases. Nine cases showed chronic dermatitis picture. Two cases showed psoriasiform dermatitis like picture. Three cases showed lichenoid dermatitis. One case showed granulomatous dermatitis picture.

Table 2: Age and sex distribution in this study of 90 patients with linear lesions are shown in (n=90).

Age (years)	Male	Female	Total
1-10	9	14	23
11-20	16	13	29
21-30	13	5	18
31-40	1	4	5
41-50	3	8	11
>50	2	2	4
Total	44	46	90



Figure 1: Lichen striatus involving extremities.

Linear lichen planus

In this study group 18 patients presented with linear LP. The age group ranged between 7 years and 70 years with an average of 30 years, forming a male: female ratio of 1.5:1. 14 cases presented with itching with duration ranging from 1 month to 7 years.

Two patients had prior drug intake (NSAID and other unknown drugs). Two patients were treated for dermatophytosis prior to the onset of lesions.

The lesions were common over the left lower extremities. Two patients had multiple site involvement (trunk and limbs).

All these patients had hyperpigmented, discrete, flat topped papules and plaques of size varying from 0.5 – 1 cm, following Blaschko's lines.

Associations found were Becker's nevus, tinea versicolor, tinea corporis, varicose vein, Freckle, HbsAg, chronic urticaria, androgenic alopecia.



Figure 2: Linear lichen planus with pterygium.

None of them showed any mucous membrane involvement and two patients had nail changes like pterygium and pitting (Figure 2).

Among the 17 biopsy, 15 specimens showed the classical features of lichen planus and in the remaining two, one had the features of lichenoid dermatitis and other had features of lichen plano pigmentosus.

Linear epidermal nevus

In this study group 19 patients presented with linear epidermal nevus in the age group between 5 years and 55 years, forming male to female ratio of 12:7. Fifteen patients were asymptomatic and mainly came for cosmetic reasons and only 4 patients presented with itching. Sixteen patients had verrucous papules and plaques and 3 patients had skin coloured velvety plaques. 14 cases had an interrupted and 5 had continuous pattern.

Most of these lesions were brown to black in color. The duration of the lesions range from 2 years to 25 years. 2 cases had second and third degree consanguineous parentage but no family members had similar problems.

The lesions were present mainly over the head and neck. Multiple site was involved in 8 patients implicating systematized form. Neurological and ophthalmological evaluations were normal for all patients. One patient had unilateral association of gigantism on same side of nevi.

Other associations are lichen simplex chronicus, TBVC, acrochordon, acne, scabies phrynoderma, intertrigo and one patient had psoriasis over the nevi lesion (Figure 3).



Figure 3: Linear verrucous nevus associated with psoriasis.

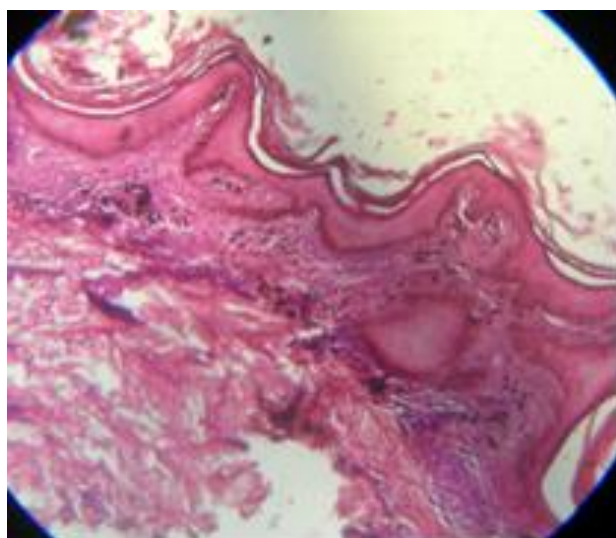


Figure 4: HPE of linear epidermal nevus showing hyperkeratosis, papillomatosis, acanthosis and patchy infiltrates in upper dermis.

Among the 14 biopsies done, 12 showed the features of epidermal verrucous nevus. Two of them showed the features of ILVEN like hyperkeratosis with foci of parakeratosis, moderate acanthosis, elongation and thickening of the rete ridges with a 'psoriasiform' appearance, papillomatosis, and slight spongiosis with exocytosis of lymphocytes (Figure 4).

Linear morphoea

In this study 9 patients of linear morphoea was recorded. Two cases were linear pansclerotic morphea, 3 cases en-coup-de sabre, 3 cases linear morphoea in extremities and trunk, one case was Parry romberg syndrome.

The lesions started in early childhood or in adolescence. The duration of lesions varied from 1 year to 4 years. Eight cases presented without any specific complaints and the remaining one had pain over the plaque. Only one patient had prior history of intramuscular injection over the lesional site. Only two cases were born of second degree consanguinity with no family history of similar illness.

Four patients had lesions over the head and neck (paramedian), Two patients had lesions involving the lower limbs (left side). Remaining two had lesion over upper limb and lower limb respectively, last patient had lesion over trunk, upper and lower limbs. The lesions were skin coloured to brownish, atrophic, indurated plaques of size varying from 3cms to 25cms, following Blaschko's lines. Three cases were found to be fixed to the underlying structures. All of them had hair loss over the plaques. None of them showed either mucous membrane or nail involvement.

On investigation, eosinophilia was seen in 3 patients and ANA was positive in 2 cases (1/10 and 1/40). X-ray, ECG, EEG and Neurological opinion for all the patients was normal. Biopsy was done for 8 patients, features consistent with morphoea.

Linear psoriasis

In this study two patients were presented linear psoriasis. Both cases presented with hypopigmented scaly plaques of 6 months to 2 years of duration, over the upper and lower extremities. There is no evidence of any trauma in preceding lesions. Both had no similar lesions anywhere else. One had pitting over finger nails. Biopsies of both were consistent with psoriasis.

Segmental vitiligo

In this study group 3 cases presented with segmental vitiligo between the age group 8 years and 21 years. One patient had lesions over the upper limb, one had lesion over face and other had lesions over the upper limb, neck and face.

The lesions were isolated macules to patches of size 0.5 cm to 10 cms. All the lesions were unilateral along the lines of Blaschko. The duration of lesions varied from 3 years to 7 years. The lesions are asymptomatic in all. None of them had vitiligo elsewhere. One patient had leucotrichia over the vitiligo patches.

Hypomelanosis of ito

In this study 2 females and 1 male patient presented with hypomelanosis of ito, between the age group 3 months and 14 years (Figure 5). Two patients had lesions over the lower limb and abdomen and the other over upper limb and abdomen. All the lesions were unilateral in distribution, along the lines of Blaschko. The duration of lesions varied from 2 months to 13 years and were asymptomatic. Examination of CNS, Skeletal system and eyes were normal in 2 cases, but one patient is under the treatment for epilepsy with normal neurological investigation.



Figure 5: Hypomelanosis of ito.

Linear lichenoid dermatitis (LLD)

In this study one female (41 years) and one male patient (24 years) presented with LLD. Both the patients had lesions over the lower limb extensor aspect and they were violaceous plaques, without any drug history.

Segmental neurofibroma

50 years old patient who was under the treatment for pemphigus vulgaris presented with segmental NF. Lesions were present in right hypochondria with positive button hole sign.

ILVEN

In this study group 3 patients recorded as ILVEN. Age group was between 9 years to 44 years. Two patients had lesions over the lower limb and the other one presented over the trunk.

The lesions were multiple scaly patches and plaques of size 1 cm to 5 cms. All lesions were unilateral. The duration of the lesions varied from 3 months to 5 years. One patient was diagnosed clinically as lichen striatus, but histopathology showed the features of ILVEN.

Incontinentia pigmenti

10 years old female patient presented with hypopigmented and hyperpigmented atrophic patches over trunk, extremities and thigh since birth, associated with aplasia cutis, right limb hypotonia, limb length discrepancy, hypoplastic external genitalia, and cicatricial alopecia over right side of scalp (Figure 6). Histopathology showed hyperkeratosis, keratotic plugging, spongiosis in some areas. Suprabasal cleft present. Increased pigment in basal layer. Pigment incontinence in the upper dermis with inflammatory infiltrate mainly around the blood vessels (Figure 7).



Figure 6: Incontinentia pigmenti.

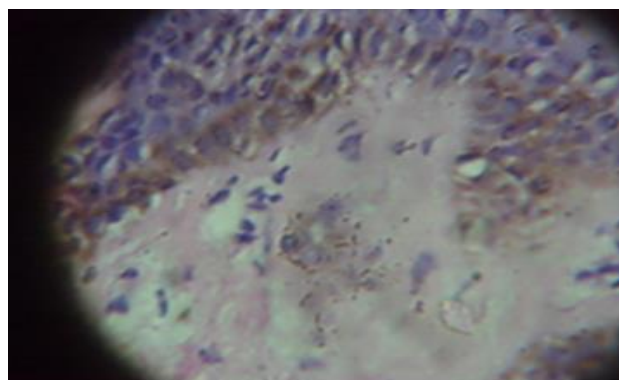


Figure 7: HPE of incontinentia pigmenti in low and high power view showing increased pigment in basal layer and pigment incontinence.

Linear porokeratosis

22 year male born out of 3rd degree consanguinity, presented with hypopigmented lesion in left foot for 5 years duration. It was associated with tinea cruris and longitudinal melanonychia in left thumb. Histopathology showed cornoid lamella, hyperkeratosis, hypogranulosis, acanthosis and lymphocytic infiltrate in upper dermis.

Linear whorled nevoid hypermelanosis

14 years male patient presented with multiple linear and whorled hyperpigmented lesion all over the body (Figure

8). Echo, X-ray, EEG was normal. Biopsy showed increase pigmentation in basal layer.



Figure 8: Linear and whorled nevoid hypermelanosis.

Nevus depigmentosus

40 years presented with hypopigmented patch in right side of face since birth.

DISCUSSION

In this study of 90 cases with a linear distribution of the lesions which did not exhibit Koebner's phenomenon, none of the cases seemed to follow the linearity determined by the Nerves, vascular or lymphatic structure and it has been suggested that these lesions develop in the lines of Blaschko.

Hence the various nevoid and acquired conditions which are supposed to follow the Lines of Blaschko, which are thought to be due to a form of human mosaicism were included in this study.

Lichen striatus

Lichen striatus formed the majority of cases in this study. The condition is said to occur commonly in the age group of 5–15 years whereas in our study, majority of patients were in the age group of between 3–20 years. It was more commonly observed in females in this study group with a Male: Female ratio 11:14 as also been documented by Hauber et al.⁶

The lesions are normally asymptomatic, with occasional pruritus in the study group as described in the literature. Most of the patients had lesions over the extremities, but few patients had lesions also over the trunk as recorded in the literature, which complied with the variable sites of expression. All the patients had unilateral distribution. Two of the patients showed nail changes among this group, although changes in the form of longitudinal

ridging, subungual hyperkeratosis, splitting and onycholysis have been documented.

Atopy was found to be associated with lichen striatus in 80% of patients, although none in this study group had personal or family history of atopy but had few coincidental associations, which were not documented so far.

Histopathological examination was consistent with the variable histological pictures as described in the literature. There were no systemic abnormalities noted in any of the lichen striatus patients in this study.

Linear lichen planus

It forms the next common condition. Most of the patients were in the age group between 11–30 years. In this study, a slight male preponderance was seen in contrary to the literature.⁷

There was no history of contact with any chemicals or trauma but 2 patients had history of intake of NSAID and another 2 were incidentally associated with freckle and Becker's naevus.

Two patients showed multiple linear lesions following the lines of Blaschko, but no immune compromised state was noted as shown in literature. On histopathological examination, 16 out of 18 lesions showed the classical features of lichen planus as described in the literature and one picture showed normal epidermis, basal cell degeneration, superficial mononuclear cell infiltrate in upper dermis, colloid bodies and pigment incontinence which was fit into the features of lichenoid dermatitis. One case was diagnosed clinically as lichen planus but biopsy showed features of lichen planus pigmentosus (LPP).⁸

But in this case it had unusual presentation like: Unilateral distribution, linear pattern, in unexposed areas-lower limb. HbsAg seropositivity was found in one of the patient, which has not been reported in literature. Some of other associated features may be coincidental.

Linear epidermal nevus

A male preponderance was seen in this study, in contrary to equal sex incidence given in the literature.⁹ All the patients mainly came for cosmetic reason. No family history was recorded. In majority of our patients, the lesions were since birth, but in few the lesions became apparent later in life. In this age group 5 patients had lesions involving trunk and extremities, implicating systemized form of verrucous epidermal nevus.

Linear morphoea

In this study nine cases of linear morphoea were recorded, among them 7 were less than 20 years of age. Generally, the peak incidence of this condition is between

20 and 30 years of the age group. Out of the 9 cases, 1 was male and 8 were female patients forming a male:female ratio 1:8 correlating with the female preponderance of this condition as recorded in the literature.^{10,11}

Except one who had prior history of intramuscular injection over the lesional site, all others (8) there was no history of any provocative factors like trauma or drug intake prior to the onset of lesion. There was no history of similar lesion in the family members. Most of the patients had an asymptomatic atrophic plaques except one who had pain over the plaque. Most of the patients (4 out of 9) had lesions over the head and neck region among which one case was diagnosed as Parry Romberg syndrome and 5 patients had lesions over the lower limb, which is the commonest site of involvement shown in literature.¹²

Linear psoriasis

Among the two patients, one had fine, regular pitting over the finger nails. On histopathological examination, the lesion showed a characteristic feature of psoriasis, with which, the clinical diagnosis was revised from lichen striatus to linear psoriasis.

Second patient was presented with asymptomatic, erythematous plaques over the extremity. On histopathological examination, it was found to have hypergranulosis and orthokeratosis, alternating with absent granular layer and parakeratosis and had other characteristic features of psoriasis.

Segmental vitiligo

One had lesions over the face and neck region corresponding to the dermatomes rather than Blaschko's lines, perhaps in keeping with a neuronal etiological theory of vitiligo and it could be also clonal susceptibility of melanocytes to neurons.

CONCLUSION

The incidence of linear dermatoses in our dermatology outpatient department, tertiary care centre, Chennai during the period of one year was 0.2%. Among the linear dermatoses, lichen striatus was found to be more common. The other dermatoses following Blaschko's lines, in the descending order of frequency seen in this study were linear epidermal nevus, linear lichen planus, linear morphea, inflammatory linear verrucous epidermal nevus, segmental vitiligo, hypomelanosis of ito, linear lichenoid dermatitis, linear psoriasis, etc. In this study, on the whole, female preponderance was noted. Most of the patient showed unilateral distribution, more often on the extremities. The importance of histopathological correlation is very obvious. Cases which were clinically diagnosed as lichen striatus showed histopathological features of psoriasis and linear epidermal verrucous nevus, one case diagnosed clinically

as epidermal nevus was found to be super imposed by psoriasis histologically. Another case which was diagnosed as linear psoriasis clinically was found to be linear porokeratosis on histopathology. It shows the importance of histopathology which ultimately changes the management in any given condition. Very few associations were noted such as, cases of lichen planus which were associated with Becker's nevus, Varicose vein, Freckles, HbsAg seropositivity, chronic urticaria, androgenic alopecia. Incontinentia pigmenti was associated with aplasia cutis, right limb hypotonia, limb length discrepancy, hypoplastic external genitalia, cicatricial alopecia over right side of scalp. One of the epidermal nevus patient was associated with ipsilateral gigantism.

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Ethical approval: The study was approved by the institutional ethics committee

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