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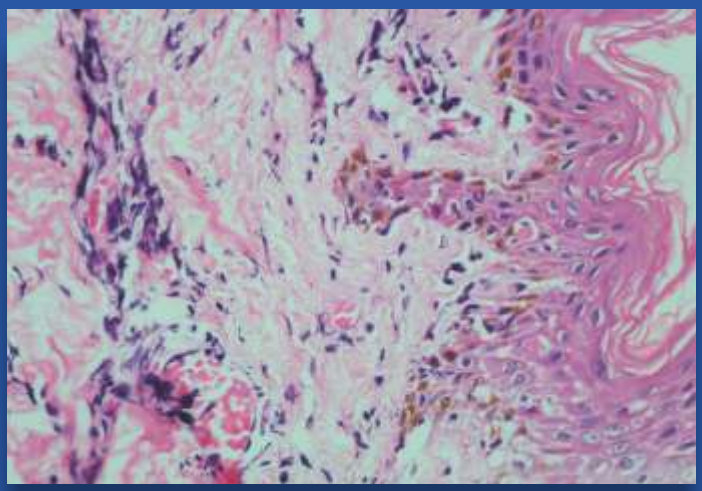

HIGHLIGHTS

A Study To Estimate Serum Vitamin D Level In Alopecia Areata

Chrysalis sign– A new dermoscopicentity in the diagnosis of angiolymphoid hyperplasia with eosinophilia

A skin coloured plaque over back - Mimesis of benign tumours

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FROM THE DESK OF EDITOR

Dear Readers !

Last year was very good for our journal since we got indexed in Index Copernicus. We have completed two years and now present you the first issue of volume 3. I thank all the authors for sending us their articles and also the reviewers for sparing their time to check the articles. We the editorial team would leave no stone unturned to provide you best quality articles. In this time of COVID pandemic we have also included a letter on this issue for you.

Thank You !!!

Dr. Dinesh Mathur
Editor

ORIGINAL ARTICLE
Year: 2020 • Volume: 3 • Issue: 1 • Page: 01-04

A CLINCO-DEMOGRAPHIC PERSPECTIVE ON MALE ANDROGENETIC ALOPECIA WITH IMPLICATIONS ON QUALITY OF LIFE IN AN INDIAN ARMED FORCES COHORT

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Abstract
Introduction: Androgenetic alopecia (AGA) is the most common form of alopecia in men and women. AGA affects approximately 50% of the male population and by the age of 20 years, over 90% of men demonstrate some degree of AGA. The present work was conducted with the aim of studying the clinical profile and quality of life in these patients. **Materials & Methods:** This hospital-based analytical study was conducted at the Dermatology department of a tertiary care hospital in Mumbai for duration of 2 years. Consecutive type of non-probability sampling was used for selection of study subjects after taking prior written informed consent. A total of 150 patients with AGA were selected on basis of eligibility criteria and informed consent. Detailed history, examination and relevant examination was carried out for all patients. Based on examination findings, patients were graded into one of the seven grades as per the Norwood Hamilton Scale. Patient's Self-Satisfaction was evaluated by the Male Androgenetic Alopecia Quality-of-Life Questionnaire. **Results:** Mean age of study subjects was 36.78 years while age of onset was 33.93 years. Most common presentation was insidious hair loss (68%) from fronto-temporal region (56.7%). Thinning of hair was seen in 84% cases while light coloured hair were present in 10.7% cases. Family history of baldness was given by 66% cases. About 2/3rd cases were in Norwood Hamilton grade II or III while remaining 1/3rd had significant frontal and vertex hair loss (i.e. Types IV–VI). A strong association was observed between age, duration of disease and its severity with quality of life ($p<0.05$). **Conclusion:** AGA can be a source of significant psychological distress to the affected patient especially at a younger age. It is thus important that physicians should consider the psychosocial impact of AGA on patient's lives during treatment.
Key words: Androgenetic Alopecia,demographic, Norwood Hamilton Scaling, Quality of Life.

Introduction

Androgenetic alopecia (AGA) is the most common type of hair loss [1]. It is characterized by progressive thinning of the scalp hair and a reduction in hair density and diameter [2,3]. Male AGA presents with a typical pattern of bitemporal and frontal recession of the hair line or vertex thinning which gradually extends anteriorly [4-7]. By the age of 20 over 90% of men demonstrate some degree of AGA. The prevalence increases with age, from 30% for men in their 30s to 50% for men in their 50s[5].

The development and occurrence of AGA depends on an interaction of endocrine factors and genetic predisposition. It is an androgen-related condition in genetically predisposed individuals and can be seen as a genetically pre-determined event[8].

As hair is an important component of identity and self-image, patients with androgenetic alopecia (AGA) may experience a distorted body image and negative feelings of social disadvantages [9-13]. Notably, even clinically imperceptible hair loss has been correlated with a decreased quality of life (QoL) [14,15]. Furthermore, Reid et al. [14] demonstrated that patients may rate their hair loss as more severe than dermatologists. Consequently, understanding the psychosocial concern and QoL of patients with AGA has become a matter of great concern.

The aim of this study was to investigate the clinical profile of patients presenting with AGA and effect of various factors on their quality of life.

Materials and Method

This hospital-based analytical study was conducted at Department of Dermatology of a tertiary care hospital in Mumbai for duration of 2 years.

Eligibility criteria:

1. Male patients in the age group of 20-50 years with AGA stage II-VI Hamilton-Norwood classification.
2. Those giving informed consent for participation in study.

Sampling Technique & Sample Size

Consecutive type of non-probability sampling was used for selection of study subjects after taking prior written informed consent. A total of 150 patients coming to our hospital with newly diagnosed Androgenetic Alopecia (AGA) were selected on basis of eligibility criteria.

Methodology

All patients were subjected to detailed history including:

- Demographic history - name, age, sex, address, contact number, marital status, occupation.
- Disease history - age of onset of hair loss, duration of hair

loss, pattern of baldness (fronto-temporal, vertical, generalized, patchy etc.), course of hair loss (acute or insidious), thinning, history of short light-coloured hair, associations (Itching / Flaking / Scaling/ Rash)

- Treatment history (5% Minoxidil / Finasteride / Dutasteride / Hair Growth Serums / Hair Restoration Surgeries)
- Family history of AGA
- Other Medical and Surgical history (to rule out differentials and associations with AGA)

Detailed examination was done for patterned/ patchy hair loss, generalized or focal, extent of hair loss, evidence of wispy, depigmented Vellus hair, and patients were graded into one of the seven grades as per the Norwood Hamilton Scaling (NHS). A set of basic investigations was conducted on patients to rule out other causes of baldness and for pre-operative work-up.

Patient's Self-Satisfaction was evaluated by the Male Androgenetic Alopecia Quality-of-Life (QoL) Questionnaire, assessing Emotional, Social and Functional outcomes of therapy.

Statistical Analysis

All statistical calculations were done using computer programs Microsoft Excel 2007 (Microsoft Corporation, NY, USA) and SPSS (Statistical Package for the Social Science; SPSS Inc., Chicago, IL, USA) version 21.

Results

Mean age of study subjects was 36.78 years while age of onset was 33.93 years (Table 1). Out of total 150 cases of AGA, 80.7% were married while 14% were unmarried. Most common presentation was insidious hair loss (68%) from fronto-temporal region (56.7%). Thinning of hair was seen in 84% cases while light coloured hair were present in 10.7% cases. Past history of treatment was positive in 62% cases with minoxidil being the most common treatment taken (43.3%) followed by finasteride (11.3%) and growth serum (7.3%). Family history of baldness was given by 66% cases (Table 2). Based on the examination findings, about 2/3rd cases were in Norwood Hamilton grade II or III while remaining 1/3rd had significant frontal and vertex hair loss (i.e. Types IV–VI) (Table 3). Mean Quality of life of study subjects was 36.7 +/- 11.1 A strong association was observed between age, duration of disease and its severity with quality of life ($p<0.05$). Quality of life was lower in younger patients (<30 years), those with greater duration (> 5 years) and increased severity (Hamilton grade IV-VI) (Table 4).

Table 1. Baseline variables among study group

Variable	N	Mean	SD
Age (years)	150	36.78	7.50
Age of Onset (years)	150	33.93	5.70
Duration (years)	150	2.85	2.30

Discussion

In our study, mean age of subjects was 36.78 years while age of onset was 33.93 years. Age distribution in present study was in accordance with previous studies [16-18]. In the study by Shankar DK, including 1005 subjects, 58% prevalence of AGA was seen in males aged 30-50 years [16]. In a study by Norwood et al., almost all patients have an onset prior to 40 years [17]. In a multinational study by Cash et al. investigating men with male pattern hair loss, 96% of participants across various countries were of age group 25–49 years and reported they were at least somewhat concerned about their hair loss, and 75% were concerned to extremely concerned [18].

In the present study, majority of patients had gradual onset of hair loss that was comparable to previous studies. In a study by Hamilton JB, both males and females with androgenetic alopecia had gradual transition from large, thick, pigmented terminal hairs to thinner, shorter, indeterminate hairs and finally to short,

Table 2. Baseline variables among study group

Variable	N	%
Marital Status		
Maried	121	80.7%
Unmarried	21	14.0%
Widowed/ Divorced	8	5.3%
Pattern of Baldness		
Fronto-temporal	85	56.7%
Central	25	16.7%
Generalized	40	26.7%
Course of Hair Loss		
Acute	48	32.0%
Insidious	102	68.0%
Personal History		
Thinning of hair	126	84.0%
Short, light coloured hair	16	10.7%
Treatment History		
Finasteride	17	11.3%
Hair growth Serum	11	7.3%
Minoxidil	65	43.3%
None	57	38.0%
Family History		
Positive	99	66.0%
Negative	51	34.0%

Table 3. Distribution of subjects as per Norwood Hamilton Grading

Norwood Hamilton Grading	Mean	SD
II	81	54.0%
III	23	15.3%
IV	7	4.7%
V	26	17.3%
V	13	8.7%
Total	150	100%

Table 4. Association of Quality of life with clinical features

Quality of Life comparison (Mean - 36.76 +/- 11.1)				
Variables		**Mean**	**SD**	**p- value**
Age (years)	< 30	31.24	10.30	<0.05
	> 30	40.32	11.21	
Duration (years)	< 5	39.41	10.89	<0.05
	> 5	33.03	11.19	
Norwood Hamilton Grade	I-III	39.12	10.33	<0.05
	IV-VI	30.28	12.12	

wispy, non-pigmented vellus hairs in the involved areas [5]. Rushton DH et al. found an average rate of hair loss of about 5% per year [19].

In our study, it was found that 84% of patients gave history of thinning of hair. Short, light coloured hair was observed infrequently. The reason behind these observations could be because of the progressive nature of androgenetic alopecia. Similar findings have been described by other authors in the literature. Paus R et al, and Plerard–Franchimont C et al. described that in AGA, the duration of anagen phase gradually decreases and that of telogen phase increases. As the duration of anagen phase determines the hair length, the maximum length of the new anagen hair becomes shorter than that of its predecessor, leading to miniaturization and eventually a bald appearance [20,21].

About 2/3rd cases were in Norwood Hamilton grade II or III while remaining 1/3rd had significant frontal and vertex hair loss (i.e. Types IV–VI). Norwood observed in his study that higher percentage of patients in Norwood type 1-3 in age groups 19-49 years. Type 4-7 being common over 60 years of age [17]. A study by Grover S et al in an Indian population had type II as the commonest presentation of AGA [22]. Similarly another study by Segal VN et al, in an Indian population had type II and III as the commonest presentation [23]. While a Chinese study by Wang et al., had type IV as the commonest type [24], and the Korean study by Paik et al., had type III as the commonest type [25].

In the present study, family history of alopecia was observed in two third of the subjects. Family history plays an important role in the onset of AGA, which is believed to be influenced by genetic factors. However, the exact mode of inheritance has not been well characterized. Although there are some reports regarding the prevalence of AGA in male paternal family members, reports regarding the maternal side are rare [26]. A very strong correlation in incidence was found in study involving 54 sets of sons and fathers, with 81.5% of balding sons having balding fathers (Hamilton-Norwood scale III or higher) [27,28].

In present study most common treatment taken by study subjects was topical Minoxidil (43.3%) followed by finasteride (11.3%) and hair growth serums (7.3%). The choice of treatment for AGA depends on various factors including efficacy, practicability, risks and costs. Important recommendations stemming from a large meta-analysis are that topical minoxidil lotion, 2% and 5% were the most commonly prescribed medication in clinical practice followed by oral finasteride[29].

Patients with AGA are significantly affected with self-image satisfaction, with potentially adverse psychosocial factors and with negative impact on a patient's QoL [30]. Alopecia has many known psychosocial complications, including depression, low self-esteem, an altered self-image, and less frequent social engagement [30]. Therefore, it has been suggested that physicians should address these psychosocial and QoL issues when treating patients with alopecia[31].

In present study strong association was observed between age, duration of disease and its severity with quality of life. Quality of life was lower in younger patients (<30 years), those with greater duration (> 5 years) and increased severity (Hamilton grade IV-VI). Not surprisingly, patients of younger age and longer durations of AGA had a decreased QoL. Physical appearance is extremely important to most young men, and early onset of hair loss can have a definite negative effect on self-image and self-esteem. Low self-esteem makes life difficult when finding life partners and employment [31].

Various studies have demonstrated that AGA can have a significant negative impact on the quality of life (Qol) of the affected persons [32-37]. A study by Kranz [34] of 160 university students with AGA revealed that the psychological distress due to AGA was not dependent on the age of the patient or stage of baldness. Hair loss affects self-esteem, personal attractiveness and may lead even to depression and other negative effects of life, especially in women. For women affected with AGA the main factors contributing to psychological distress were - inability to style their hair, dissatisfaction with their appearance, concern about the continuing hair loss and concern about others noticing their hair loss [35]. In a study Han et al. investigated the QoL of AGA patients. Their results showed that AGA could harmfully affect the patients' QoL. QoL was more damaged if the patient had severe alopecia, a longer duration of AGA, younger age, had received previous non-medical hair care, and visited the hospital for AGA treatment. They concluded that dermatologists should address these QoL issues when treating patients with alopecia [36].

Conclusion

Androgenetic alopecia is one of the commonest dermatological complaints for which patients seek treatment. The disease is generally presents in mid-thirties with gradual loss of hair from fronto-temporal region. AGA can be a source of significant psychological distress to the affected patient especially at a younger age. It is thus important that physicians should consider the psychosocial impact of AGA on patient's lives during treatment. Patients with low quality of life might need both medical treatment and psychotherapy, as well. Further research is needed to better understand the effects of AGA and to improve treatment on self-image, psychological functioning and quality of life.

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A STUDY TO ESTIMATE SERUM VITAMIN D LEVEL IN ALOPECIA AREATA

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Abstract

Background: Alopecia areata (AA) is an autoimmune disease which is characterized by patchy hair loss and affects any hair bearing area. Low level of vitamin D have been implicated in variety of autoimmune diseases. This study was conducted to assess the level of vitamin D in patients with AA and its correlation with severity, pattern & extent of disease.

Materials & Methods : This is hospital based Case control study including 50 cases with AA and 50 age and sex matched controls. AA cases were grouped according to severity, pattern & extent of disease. Level of vitamin D were assessed and compared to controls. The data was analysed and correlation was derived.

Results: Revealed significant difference between cases and control as regards with 25(OH) vitamin D level. There is also a significant difference between 25(OH) vitamin D status and degree of AA. However, there is non-significant difference in 25(OH) vitamin D status in all participants regarding age and sex of subjects.

Conclusion: Vitamin D deficiency may be one of the factors having a role in etiopathogenesis of AA. Thus, supplementation of vitamin D as a treatment modality may improve the clinical outcome of AA.

Keywords – Alopecia areata, SALT Score, Vitamin D.

Introduction

Alopecia Areata (AA) is an autoimmune disease which is characterised by hair loss and can affect any hair bearing area. AA present with different clinical manifestations varying from reversible patchy hair loss to complete baldness

or complete body hair loss. It affects approximately 1-2 % of general population[1].

AA is an organ specific autoimmune disease characterized by T cell infiltrates and cytokine production around anagen hair follicle.[2,3] .

AA has association with human leukocyte antigen – class I & Class II and known to occur with various autoimmune disorders such as Diabetes mellitus, Thyroiditis, vitiligo, SLE, rheumatoid arthritis [4,5]. Thus, AA is considered as hair follicle –specific autoimmune disease, triggered environment factors in genetically susceptible individuals [1,3] .

Vitamin D is synthesized in the epidermal keratinocytes under effect of UV-B lights (290–315 nm) or ingested in diet and dietary supplements [6]. Vitamin D was found to have immune-regulatory effects. 1,25-Dihydroxy vitamin D3 (1,25(OH)2 D3) which is the active form of vitamin D, is one of the regulators of both innate and adaptive immune responses as it modulates immune function and activities of both T-lymphocytes and B-lymphocytes.[7]

Vitamin D receptors (VDR) expression in epidermal keratinocytes and the mesenchymal dermal papilla cells were detected [8]. Expression of the VDR in keratinocytes is necessary for preservation of the normal hair cycle [9]. Lack of the VDR is associated with reduced epidermal differentiation and hair follicle growth.[10]

The increase incidence of AA let us search more for possible etiological factors. In view of these points, we designed this study to find the association between Vitamin D level and AA.

Methodology

This is case control study performed in outpatient department in Department of Dermatology, Venereology and Leprosy, JLN Medical college and Attached group of Hospitals, Ajmer , Rajasthan after taking approval from ethical committee. The study enrolled 100 subjects of either sex with different groups. Patients of AA of different age and sex attending the Department of Dermatology, Venereology and Leprosy were enrolled in study (Cases) group 'A'. Any patient taking vitamin D supplementation, iron preparations, or calcium (Ca) supplementations, topical vitamin D3 analog were excluded from the study. Patients with any associated disease that alter the blood 25(OH)D level as vitiligo, psoriasis, SLE, renal disease, liver disease, were also excluded from this study.

In control group (Group B) age and sex matched individuals without AA who were suffering from minor ailments were included in this study.

Clinical assessment of the degree of alopecia areata was determined by Alopecia SALT score.

Serum 25(OH)D concentration, the major circulating form of vitamin D, was measured using commercial Enzyme-Linked Immunosorbent Assay (ELISA) kits (Immunodiagnostic Systems Limited, Bolden, UK). Range of Vitamin D level is

Deficient (0-30 ng/ml)

Insufficient (31-39 ng/ml)

Normal (40-80 ng/ml)

RESULTS

Mean age of study group was 21.44 ± 12.301 years whereas of control group was 27.28 ± 10.859 years. In this study there were 32(64%) males and 18(36%) females while in control group 33(66%) males and 17(34%) females. Both age and sex are significantly different.

Table 1. Socio- demographic characters of control and cases.

Variable	Cases		Control		Test	P-Value
*Age						
Range	4-61		9-59		t=2.56	0.013
Mean±SD	21.44 ± 12.301		27.28 ± 10.859			Sig.
*Sex	N	%	N	%	χ2 value	
Male	32	64	33	66	χ2=5.120	0.048
Female	18	36	17	34	χ2=3.920	Sig.

When the level of serum 25(OH)D was compared in both the groups i.e. it was found that there were 30(60%) patient have deficient vitamin D level, 5(10%) have insufficient while 12(24%) have normal and 3(6%) have toxic level of vitamin D in case group whereas in control group 19(38%) have deficient level, 28(14%) have insufficient , 16 (32%) and 1(2%) have toxic level of vitamin D. Vitamin D deficiency is observed more in case group compared to control & difference is statistically significant.

Table 2. Comparison between cases and control regarding serum 25(OH)D status and level

Serum Vitamin D	Cases(n=50)	Control(n=50)
Deficient (0-30)	30 (60%)	19(38%)
Insufficient (31-39)	05 (10%)	28(14%)
Normal(40-80)	12 (24%)	16(32%)
Toxic (> 150)	03 (6%)	1(2%)

Study group shows Maximum 44 (88%) patients were suffering from Patchy pattern of hair loss, followed by 3 (6%) patients were suffering from A Totalis. 2 (4%) patients were suffering from Ophiasis. 1 (2%) patient was suffering from A Universalis.

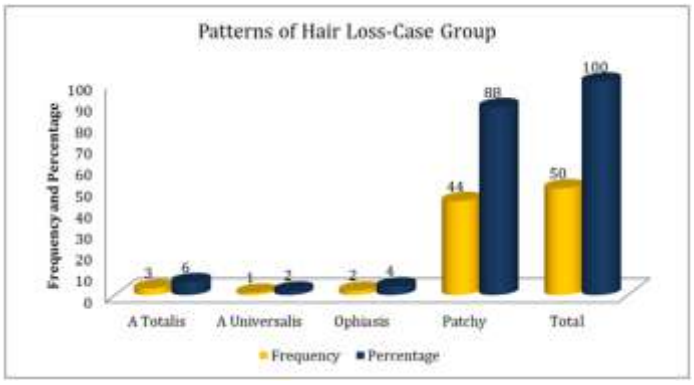

Figure 1 : Pattern of hair loss – Cases group

In our study There were 50 patients in both control and cases groups. The Mean of vitamin D levels in control group 36.40±17.94 . The Mean of vitamin D levels in control group was 29.23±21.74. The relationship between Control and Cases Groups regarding Vitamin D Levels was significant being the value < 0.05. The correlation between Control and Cases Groups regarding Vitamin D Levels was -0.197.

Table 3. Relationship between Control and Cases Groups regarding Vitamin D Levels

Control Group			Cases Group		
Number of Patients	Vitamin D Level (ng/ml) Mean ± SD	Range	Number of Patients	Vitamin D Level (ng/ml) Mean ± SD	Range
50	36.40±17.94φ	79.5	50	29.23±21.74φ	134.0
p-value			<0.05*		
Correlation			-0.197		

In our study of severity of hair loss is assessed by SALT Score. Present study shows association of SALT with mean serum vitamin D levels was significant(P< 0.05). Patients with higher SALT (S4 &S5) Had very low levels of vitamin D level.

Table 4. Relationship between Vitamin D Level and Severity of Hair Loss

SALT Score	Number of Patients (%)	Vitamin D Level (ng/ml) Mean ± SD	p-value
S0	0 (0)	0	<0.05*
S1	31 (62)	33.89±30.40	
S2	11 (22)	38.99±28.84	
S3	3 (6)	36.03±36.97	
S4	1 (2)	25.5	
S5	4 (8)	23.89±14.67	

In our study There were 3 (6%) patients were suffering from A Totalis, 1 (2%) patient was suffering from A Universalis, 2 (4%) patients were suffering from Ophiasis and 44 (88%) patients were suffering from Patchy pattern of hair loss. The pattern of hair loss was having the mean of 3.74 ± 0.777. On the other hand, vitamin D level is showing the mean of 29.23 ± 29.26. The Relationship between Patterns of Hair Loss and Vitamin D Levels is significant being the value less than the critical value of 0.05 and negatively correlated (r= -0.036)

Table 5. Relationship between Patterns of Hair Loss and Vitamin D Levels

Statistics	Pattern of Hair Loss				Vitamin D Level
	A Totalis	*A Universalis*	*Ophiasis*	*Patchy*	
Frequency	3	1	2	44	-
Mean ± SD	3.74±0.777				29.23±29.26
Correlation	-0.036				
p-value	<0.05*				

Discussion

Alopecia Areata is an autoimmune disease having various triggering factors. Vitamin D is prohormone which is synthesised by skin & regulate various immune response. It is hypothesised that development of hair follicle depends on Vitamin D receptors by controlling Vitamin D receptor expression.

We conducted study in patients Age ranged from 4-61 years with mean 21.44 ± 12.30. Maximum no of patients i.e 17 (34%) were in age group 21-30 followed by 14 (28%) in 11-20 which was comparable to study conducted by Panda et al 16 where max no of patients is 32 (44%) were in age group 21-30 followed by 21 (29 %) in 11-20.

In our study male patient were 64 % . The male to female ratio was (1.77: 1) . Similar male predominance ratio was also found

by Yilmaz N et al 8 (2:1), Cerman et al 12 (1.87: 1), Attawa EM et al 14 (1.87:1) and Nassiri et al 17(2.1:1).

Our results were almost equal to the study conducted by Yilmaz et al 8 in which severity of AA cases (According to SALT Score) was seen in decreasing trend as follows S1(71.4%) cases were in S1 grade then , 14.2% in S2, 7.4% in S3 , 4.7 % in S4 % and 2.5 % in S5 grade. This is similar to our study with maximum patients had S1 grade (n=31, 62 %) , followed by S2 (n=11, 22%) , S3 (n=3, 6 %), S4 (n=1, 2%), S5 (n=4, 8 %).

We noted that, the mean serum vitamin D level was significantly lower as compared to controls (29.23± 21.74 Vs 36.40± 17.94 ng/ml P< 0.05). Our results were parallel with study made by Yilmaz N et al 8, Cerman AA et al 12, Mahamid M et al 13 , Attawa EM et al 14, EL -Mongy NN et al 11 and Bhat YJ et al 15 found similar significant lower levels of serum vitamin D in Alopecia areata patients. The mean serum vitamin D levels were low in all studies as we observed.

Cerman AA et al 12 , Attawa EM et al 14 and Bhat YJ et al 15, found a significant inverse correlation between SALT score and serum vitamin D levels Similarly as we found in our study.

Bakry et al 18 in their study showed a significant inverse correlation with pattern of disease. In our study we noticed similar correlation.

These variations in mean serum vitamin D level may be due to studies being conducted in different geographical areas along with degree of sunlight exposure and dietary intake of vitamin D.

Conclusion

Vitamin D is deficient in more number of patients of Alopecia areata. Vitamin D level is also inversely correlated with Severity, extent & pattern of the disease. Hence Vitamin D deficiency may be one of the factors involved in etiopathogenesis of Alopecia areata or may be the exacerbating factors. Further clinical studies are required to confirm the role of vitamin D as a therapeutic agent in Alopecia areata.

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ORIGINAL ARTICLE
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CLINICO-EPIDEMIOLOGICAL PROFILE OF PATIENTS WITH LICHEN STRIATUS: A RETROSPECTIVE STUDY FROM A TERTIARY CARE CENTRE IN NORTH INDIA

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Abstract

Background: Lichen striatus (LS) is a benign, self-limited, linear, inflammatory dermatosis of unknown etiology that usually affects children. Most of the literature on lichen striatus has appeared as case reports or isolated case series from India.

Material and Methods: Clinical records of children with LS, who attended the Department of dermatology, at our tertiary care centre, from July 2013 to December 2017, were analyzed.

Results: Twenty nine patients were analyzed. Mean age was 6.5 years, ranging from 1.6 year to 12 years. We found 17 (58.62%) male and 12 (41.38%) female cases with slight male preponderance. Nine (31.03%) patients were less than 3 year, 13 (44.82%) patients from 3-6 year age group, 7 (24.13%) patients from 6-12 year age group . Most common site was upper limbs 11 (37.93%) cases followed by trunk 7 (24.13%). Most common complaint was hypo pigmentation in 22 (75.86%) children by the parents. Nail involvement was seen in 3 (10.34%) boys.

Conclusion: In this large case study, lichen striatus appears as a disease of young children with a male predilection. The disease appears statistically more often in the cold seasons, particularly in the winter. The most frequently involved sites are the limbs, while nail involvement is very rare.

Keywords: Lichen striatus, Linear inflammatory dermatosis

Introduction

Lichen striatus (LS) is an asymptomatic self-limited skin disease of unknown etiology which was first described by Senear and Caro in 1941.[1] LS is characterized by erythematous or brownish papules with a flattened surface that are frequently scaly in appearance and occasionally display vesicles. The lesions are usually solitary and unilateral and have a linear distribution following Blaschko's lines, usually on the extremities.

Atypical forms with multiple and bilateral lesions have been described. Onset is usually sudden, with the disease progressing over days or weeks and slowly decreasing spontaneously until the papules resolve within 6–24 months, leaving a transitory residual hypopigmentation, especially in patients with a dark complexion.

The inflammatory phase is not always detected, and hypopigmentation may be the first manifestation. The higher incidence during spring and summer, along with the existence of familiar clustering, suggest that viral infections could be an elicitation factor. Other possible precipitating factors may include cutaneous injury, trauma, hypersensitivity, or other as yet unspecified factors.

Material and methods

Medical records and photographs were used in a retrospective study of 29 children affected by lichen striatus who were seen at the Department of Dermatology, Government Medical College, Kota, India, between July 2013 and December 2017. We studied the gender, age, and family history of the patients as well as the season of onset, morphology, distribution, extent, duration, itching, residual hypopigmentation and treatment of lichen striatus and any associated symptoms or diseases.

Results

Twenty nine patients were analyzed (Table 1). Mean age was 6.5 years, ranging from 1.6 year to 12 years. We found 17 (58.62%) male and 12 (41.38%) female cases with slight male preponderance. Nine (31.03%) patients were less than 3 year, 13 (44.82%) patients from 3-6 year age group, 7 (24.13%) patients from 6-12 year age group. Most common site was upper limbs 11 (37.93%) cases followed by trunk 7 (24.13%) (figure-1). Most common complaint was hypo pigmentation in 22 (75.86%) children by the parents. Nail involvement was seen in 3 (10.34%) boys.

Morphology

Based on the morphology of the lesions, we identified the following three clinical patterns:

Lichen striatus albus, with hypopigmented macules and/or papules with only a few typical lichenoid pink papules in 22(75.86%) patients. Typical lichen striatus, with pink, red, or flesh colored, flat-topped, lichenoid papules was found in 4(13.79%) patients.

Table 1. Comparison of Visual analogue scale (VAS) in both the study groups at different time interval.

Clinical profile	Number of patients	Percentage(%)
Age (Years)	(Mean 6.5 years; range 16 months -12 years)	
1-3	9	31.03
3-6	13	44.82
6-12	7	24.13
Sex (M:F)	17:12	
Family history of LS	None	
history of atopy	8	27.58
Season of onset		
Winter	8	27.58
Spring	7	24.13
Summer	6	20.68
Autumn	8	27.58
Distribution		
Single linear/curved band	22	75.86
Multilinear band	6	24.13
Localised oval patch	1	3.44
Involved sites		
Single leg	4	13.79
Single lower limb with buttock	2	6.89
Single arm	3	10.34
Single arm with shoulder	4	13.79
Single Hand	1	3.44
Only head/neck/face area	4	13.79
Only trunk	4	13.79
Back with shoulder	3	10.34
Only forearm	3	10.34
Penis	1	3.44
Morphology		13.79
Typical Lichen striatus	4	75.86
Lichen striatus albus	22	10.34
Nail Lichen striatus	3 (2 finger and one toe nail)	
Associated diseases		
Atopy (atopic dermatitis allergic rhinitis allergic conjunctivitis,asthma)	8	27.58
P. alba	2	6.89
History of trauma	None	
History of viral fever	None	

Nail lichen striatus, which includes onychodystrophy, thinning, longitudinal ridging and splitting, fraying, and onycholysis restricted to the lateral portion of the affected nail, or more rarely, to the medial portion. Nail lichen striatus was seen in three (10.34%) boys, ages 9 and 11 years (figure-2). In all patients of nail lichen striatus, cutaneous involvement was also seen.

Distribution of lesions was unilateral in all patients with 19 (65.55%) on the right side and 10 (34.48%) on the left side of the body. Thirteen patients (44.82%) had pruritus and 16 (55.17%) were asymptomatic. Eight patients (27.58%) had a history of atopy.

All of the 29 patients analyzed in regard to seasonality, the appearance of lesions occurred in 7 (24.13%) during spring months, in 6 (20.68%) in the summer, in 8 (27.58%) in autumn and in 8 (27.58%) in winter.

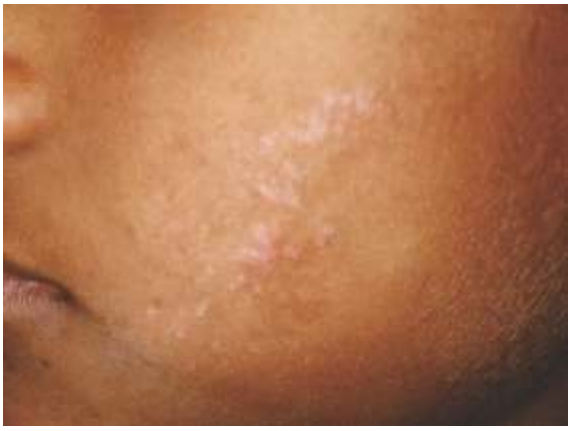

Figure 1a : Showing distribution of lichen striatus lesions ; 1a- over face

Figure 1b : lichen striatus lesions over penisLtd

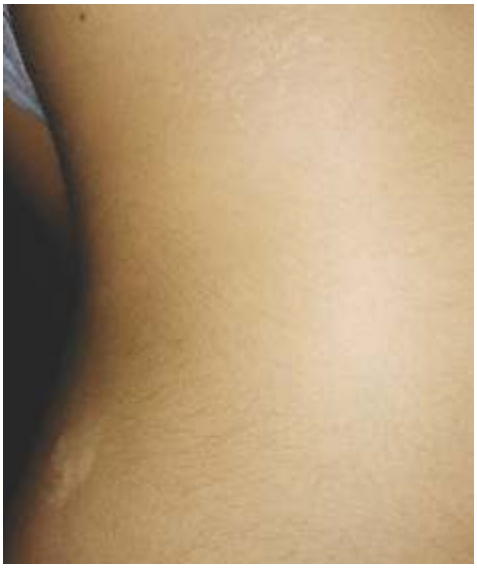

Figure 1c : lichen striatus lesions over trunk

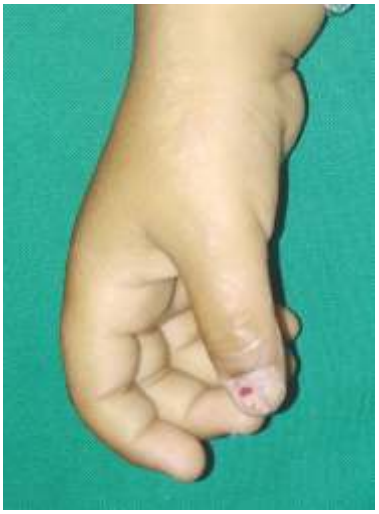

Figure 2a : Variants of lichen striatus ;2a -linear LS

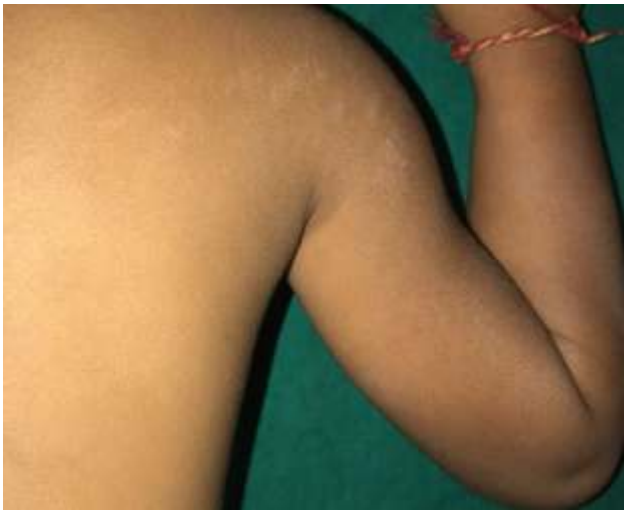

Figure 2b : : Hypopigmented variant of lichen striatus

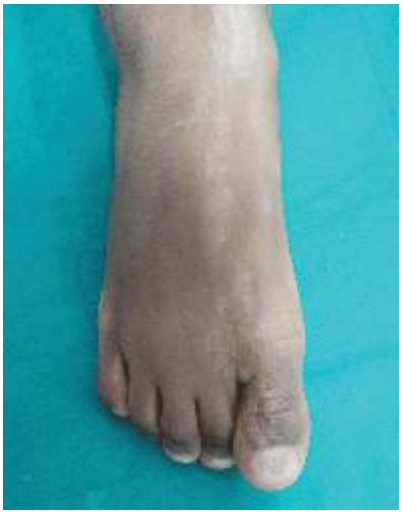

Figure 3 : Nail lichen striatus

Table 2. Review of the various studies of lichen striatus

Series of cases by author/year	Number of cases	Mean age (years) (range)	Gender M/F	Seasons of presentation (%)	Location (%)	Mean duration of Episode	Itch (%)	Residual hypopigmentation (%)
Our study	29	5.06 (16m to 12 yr)	19:12 (3:2)	Spring (24.13) Summer(20.68) Autumn(27.58) Winter (27.58)				
Kennedy et al5	61	2.98	1:2	Spring and summer (71)	EE 77, IE>SE	NC	NC	NC
Sittart et al8	53	2.5 y (1–40 m)	1:2.3	Spring and summer (75)	EE 92	NC	NC	NC
Taniguchi et al18	89	29 d–14 y)	1:3	Spring (22.4) Summer (34.2) Autumn (22.4) Winter (21)	EE 86.5 IE>SE	NC	34	NC
Patrizi et al19	115	4 y + 5m (1 m–13 y)	1:2	Spring (24) Summer (4) Autumn(23) Winter (48)	EE 62 IE>SE	6 m	11	28.57, 3.8 hyper

Discussion

The analysis of 29 patients with LS found a male:female ratio of 1.41:1 showing a clear preponderance of the disease in boys, in contrast to the findings of other studies.[2,3,4,5.6] Taieb et al 7 found an equal incidence in both sexes. The ages of our patients ranged from 1.6 to 12 years, the same age interval as has been reported in other series 2,3,4,7 Review of the various studies is shown in table 2.

As in the study of Kennedy and Rogers 5 and Sittart et al 8, who reported an incidence peak in preschool children, we also found 13(44.82%) cases of LS in 3 to 6 years age group.

In some reported series there was mostly involvement of the upper limbs 3,6,7. Kennedy and Rogers 5 found lesions on the superior limbs 1.7 times more often, as was also seen by Ruiz-Maldonado et al.2 In our study, 11(40.7%) of 29 patients had lesions on the upper limb and 6 had lesions on the lower limbs. This means that almost half of the instances occurred on the upper limbs; this location is 2 times more common than occurrence on the legs 4 (13.79%).Other locations were also affected, such as the trunk (thorax, abdomen, and dorsum) and neck. In all patients, the lesions followed a pattern corresponding to the lines of Blaschko.

Lichen striatus is a dermatosis of unilateral manifestation in the majority of cases.[2,5] In our study, all of the patients had unilateral lesions. The right side was involved more than left and the side involved was independent of gender. Bilateral lesions were described in few case reports.[9-13] Bilateral lesions were not found in our patients.

In addition, 16 (55.17%) of our patients were asymptomatic. This finding is in agreement with the series of Ruiz- Maldonado et al[2], who described 80% of patients without associated symptoms. Thirteen (44.82%) patients in our study had pruritus

and this feature was independent of age. A relevant finding was that all of the atopic patients were 8 (27.58%) had pruritus .

Toda et al[14] , in a study of 26 patients with LS, found that 84.6% had atopy, suggesting that a personal or family history of atopy could favor the appearance of the disease. Di Lernia et al[15], studying a group of 19 children, found that 53% had atopy, a much lower percentage than that observed by Toda et al.[14] In our study, only 8 (27.58%) patients have personal history of atopy. These findings do not allow the association of LS with atopy, since this number is very close to the incidence of atopy in the general po[pu]lation, which is 15% .[16]

Kennedy et al[5] observed an increased incidence of LS in spring and summer, and suggested a possible viral origin. Significant differences in regard to the season of the year were not found in our study. In our study most of the patients 16 (55.16%) appeared in winter and autumn while 13 cases appeared in other rest of the seasons.

Differential diagnosis of LS may include different linear papular eruptions such as linear psoriasis, linear Darier's disease, linear lichen planus, linear porokeratosis and inflammatory linear verrucous epidermal nevus (ILVEN). Hypopigmented lesions may be confused with linear vitiligo or nevoid hypomelanosis (the so-called hypomelanosis of Ito). Of these entities, ILVEN may be the most difficult to differentiate. In contrast to LS, ILVEN is always pruritic and it does not regress spontaneously but instead undergoes periods of exacerbation with periods of improvement.[17]

Conclusion

In conclusion, through this study we found that LS is a disease that predominately affects boys between 1.5 and 14 years of age, involving mostly the upper limbs unilaterally, with right side being affected more than left side and with a pattern following the lines of Blaschko.

Most of the children did not show any symptoms but when symptomatic, the most common finding was pruritus.

Asymptomatic lesions were twice as common as pruritic ones. Atopy was not a striking feature, as its proportion was similar to that found in the general population. Nevertheless, although the existence of atopy or pruritus was infrequent, they tended to be seen together in many patients. The majority of atopic patients presented with pruritic lesions and this may suggest that the sensitive skin of these children is more susceptible to the development of exacerbated reactions in LS lesions.

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LETTER TO EDITOR
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A CASE OF PITYRIASIS LICHENOIDES ET VARIOLIFORMIS ACUTA (PLEVA) LASTING FOR 22 YEARS-A SUBACUTE VARIANT?

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Sir,

Pityriasis Lichenoides et Varioliformis acuta (PLEVA) is an acute variant of Pityriasis Lichenoides which encompasses a spectrum of clinical presentations ranging from rapidly progressing severe ulceronecrotic variant with systemic manifestations and a high mortality rate (Febrile ulceronecrotic Mucha Haberman Disease- FUMHD) to a more chronic manifestation consisting of benign-appearing scaly papules of Pityriasis Lichenoides Chronica (PLC).[1]

It is generally accepted that PLEVA and PLC represent two ends of a continuous spectrum, and therefore it is not uncommon to observe both acute and chronic lesions in the same patient, as well as lesions at intermediate stages between PLEVA and PLC. The maximum duration of PLEVA lasting in any person has been reported as long as 08 years.[2] Herein we report a case where it lasted for nearly 22 years.

A 25-year-old female presented to our center with complaints of recurrent crops of red raised lesions over her body, occurring at a frequency of once a week and then resolving spontaneously over a period of 10 to 15 days since the age of three years. There was no history of any pain or itching over the lesions. No history of fever or malaise associated with the occurrence of lesions. There was no history of weight loss, evening rise of temperature or night sweats. The patient denied any similar lesions in family members.

General and systemic examination was essentially normal. Dermatological examination revealed the involvement of face, trunk, and lower limbs in the form of few discrete erythematous papules, few vesiculopustules, with some papules showing a central necrotic eschar or crust, measuring 0.5 cm – 2 cm in diameter [Figures 1 & 2]. Examination of mucosa, palms, and soles was within normal limits.

Relevant hematological and biochemical parameters were within normal limits.

Histopathology showed characteristic features of necrotic keratinocytes with extravasation of RBC's into the epidermis. Papillary dermis was edematous and the capillaries were congested with RBCs. Superficial and the deep dermis showed endothelial swelling with foci of fibrinoid necrosis within the capillaries[Figure 3]. Perivascular chronic inflammatory infiltrates of lymphocytes was seen. Immunohistochemistry was

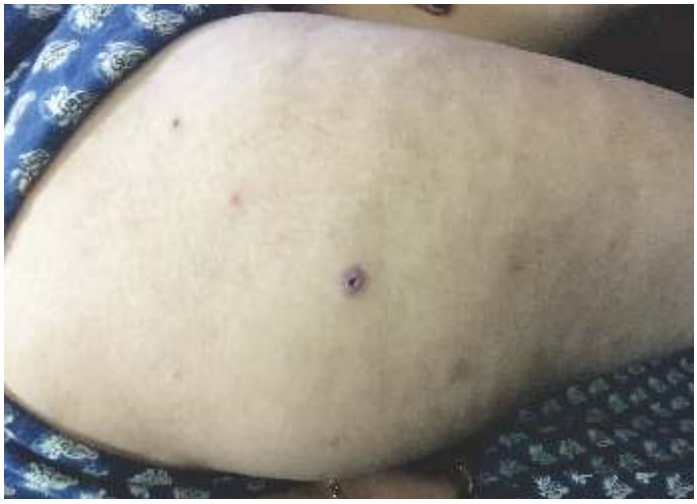

Figure 1 : Presence of few discrete erythematous papules, few with necrotic centre over the thigh

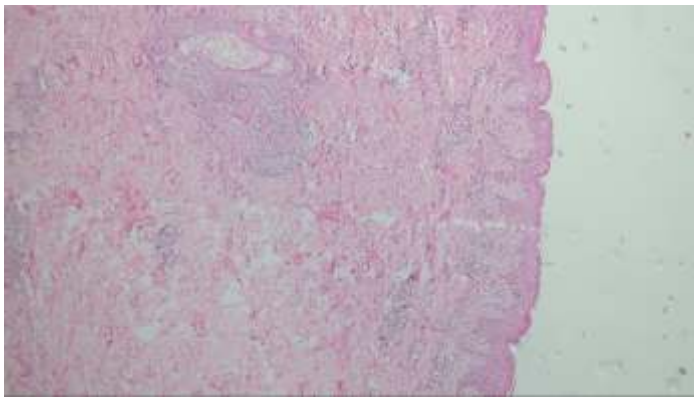

Figure2 : H&E(100x)- scanner view showing predominant interface dermatitis with vacuolar degeneration and edematous papillary dermis

positive for CD3+,CD7+and CD8+ and CD 30- [Figure 4]. Based on the above clinical, histopathological and immunohistochemistry a diagnosis of PLEVA was made.

The patient was managed with Cap Doxycycline 100mg twice daily along with 0.1% mometasone cream applied twice daily with which she has shown an excellent response in the form of no new lesions and regression of older lesions.

PLEVA is a disease of young people and children with inflammatory and usually diffuse erythematous papules of rapid onset which

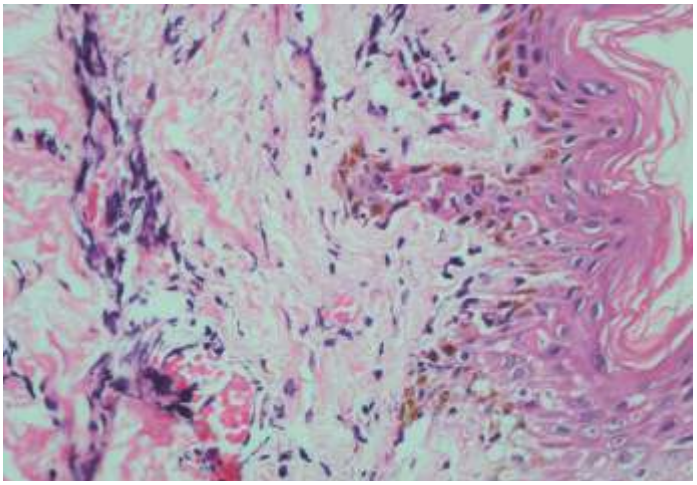

Figure 3 : H & E (200x)Necrotic keratinocytes with extravasation of RBCs into the epidermis with superficial and deep dermis showing endothelial swelling with foci of fibrinoid necrosis within the capillaries. Papillary dermis was edematous and capillaries were congested with RBCs.

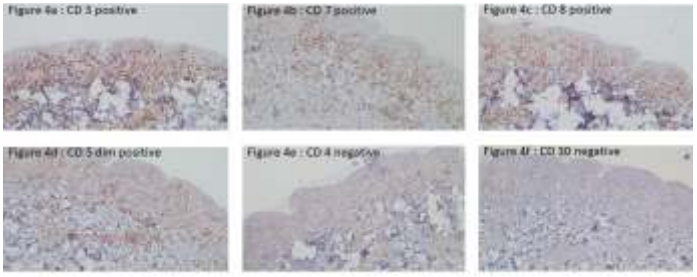

Figure 4(a -f) : IHC (200x) showed positivity for CD3+, CD7+, CD8+ and negative for CD 30-

evolve to form papules with a central punctum. The papule evolves to become vesicopustular, undergoes hemorrhagic necrosis and becomes ulcerated with overlying crusts. These lesions spontaneously heal to leave behind varioliform or smallpox-like scars and post-inflammatory hyperpigmentation and hypopigmentation. 1,2 Though the distribution of the lesions can be generalized, the trunk, flexors, and extremities are commonly involved.

The etiology of the disease still remains unknown though antibodies against Toxoplasma gondii, cytomegalovirus, parvovirus, adenovirus and Epstein Barr virus have been demonstrated in some studies. 1-3 T-cell dyscrasias and immune complex deposition have also been cited as probable causes in the literature.

The differential diagnosis for PLEVA includes lymphomatoid papulosis, arthropod bite reactions, varicella, Gianotti-Crosti syndrome, erythema multiforme, pityriasis rosea, guttate psoriasis, vasculitis, and secondary syphilis. Differentiation between PLEVA and lymphamatoid papulosis is important as patients with lymphomatoid papulosis may develop systemic lymphoma. The presence of large CD30+ atypical lymphoid cells is the hallmark of lymphomatoid papulosis and clinically, the papules of lymphomatoid papulosis may develop into nodules, tumors, and large plaques.[4]

Cinical management of PLEVA is difficult due to exact pathology being unknown. Medications used till date include tetracycline, erythromycin, methotrexate, calciferol, chinoline and acridine derivatives, cyclosporine, intravenous gamma globulin, and retinoids.5 Systemic corticosteroids may have a role in severe cases of PLEVA. Topical corticosteroids and topical tacrolimus are helpful in symptomatic cases, but they do not alter the course of the disease. Despite a lack of randomized controlled trials, oral tetracycline and erythromycin have been prescribed most often in case series.

The existing literature outlines the mean duration of this rare disease to be 1.6 year with an episodic course, the frequency of which varies greatly from individual to individual.[2] The longest duration mentioned in literature is 8 years. Our case highlights a case PLEVA with a disease duration of 22 years with a subacute course which till now is the longest reported duration for the disease in literature.

We present this case to stress upon this rare presentation of PLEVA which can exist for years without any systemic manifestations and resolves spontaneously leaving behind post inflammatory hyperpigmentation. To give it a new nomenclature as "Subacute PLEVA" would not be wrong.

How to cite this article:

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LETTER TO EDITOR
Year: 2020 • Volume: 3 • Issue: 1 • Page: 14

POHL PINKUS LIKE CONSTRICTIONS IN LICHEN PLANO PILARIS ON DERMOSCOPY

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Sir,

A 32 year old female presented with complete loss of hair over the scalp since 5 years (Figure 1). Treatment taken in the past was not effective and she still complained of new lesions with loss of hair. Skin of the alopecic patch involving almost the entire scalp sparing only the peripheral rim of hair line throughout, appeared shiny and atrophic and hair pull test from the periphery of the alopecic patch was positive.

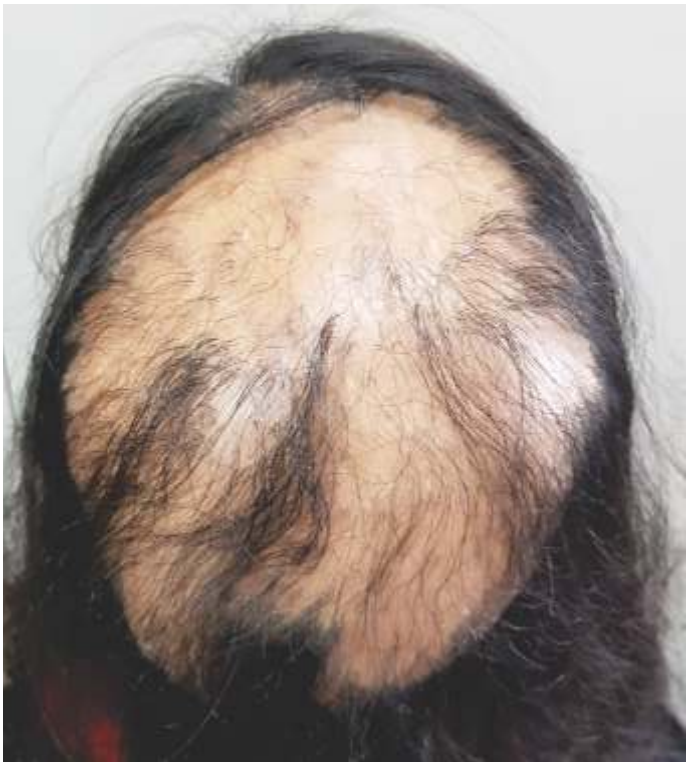

Figure 1: Loss of hair involving 85% scalp area

Dermoscopic examination (Heine dermatoscope) revealed diminished follicular ostia, tubular perifollicular scaling and peripilar casts. Perifollicular blue-grey dots- target sign, suggestive of melanin incontinence were present1. White areas indicating fibrosis in the interfollicular area were seen. Interestingly, all hair in the affected area demonstrated varying thickness throughout the length of the hair shaft (Figure 2).

Figure 2: Dermoscopic picture-(Heine dermatoscope under 50x magnification polarized mode) Yellow arrow-peripilar cast, Red arrow- blue gray dots, Green arrow-white areas, Blue arrow-Pohl Pinkus like constrictions

Multiple pohlpinkus like constrictions were seen throughout the hair shaft probably suggestive of the multiple episodes of trauma that the hair experienced each time there was a flare of the disease. Integrin expression has a role in cell cell adhesion , epidermal differentiation and migration , its altered expression is seen in active lesions of lichen planus pilaris and probably the reason for pohlpinkus like constrictions here. Trichogram revelaed floppy sock appearance due to roughened cuticle. To conclude, lichen planopilaris is the most common cause of cicatricial alopecia. It frequently affects middle-aged females with irregular areas of atrophic alopecia on the scalp. Dermoscopy can act as a useful non-invasive aid in diagnosing lichen planopilaris .

How to cite this article:
Agarwal S, Dhurat R, Surve R, Daruwalla S, Sharma A. Pohl pinkus like constrictions in Lichen Plano Pilaris on dermoscopy. JDA Indian Journal of Clinical Dermatology 2020;3: 14

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LETTER TO EDITOR
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CHRYSALIS SIGN– A NEW DERMOSCOPICENTITY IN THE DIAGNOSIS OF ANGIOLYMPHOID HYPERPLASIA WITH EOSINOPHILIA

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Sir,

Dermoscopy plays a very important role in differentiating this benign, vasoproliferative disorder from ominous diseases such as Kaposi's sarcoma, Kimura's disease and cutaneous metastases. Dermoscopic signs, reported herein, and in a seven-case-series by Padilla et al include linear vessels, red dots, red lacunae, ulceration over a pale red background, when ALHE is examined under polarizing dermoscopy. In our observation of three cases, we found another sign – the Chrysalis sign, which is an established sign in a few conditions, viz., basal cellcarcinoma, melanomas, dermatofibromas and scar tissue. But this has not been reported in conjunction with ALHE.

In our cases, the site of presentation varied from the centro-facial region, to the concha of the ear and scalp. (Figure-1) Ulceration, with or without frank hemorrhage wa snoted in two cases. All three cases showed a central, yellow structureless area with chrysalis-like, white streaks on a homogenous red background with few linear and dotted vessels. This chrysalides pattern is seen, particularly, as short, white orthogonal and parallel streaks, akin to the structural framework of the pupa, an intermediate stage between larval and adult life in a butterfly or a moth, and hence the nosological analogy. It is seen exclusively on polarized dermoscopy due to refringence from the hypertrophic or disoriented collagen in the dermis. (Figure-1)Fig 1

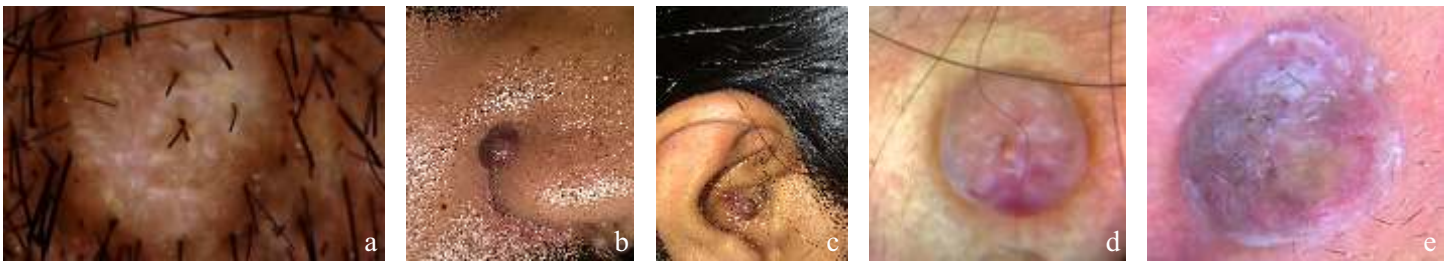

Figure 1 a,b,c,d,e: a. Multiple grouped skin coloured to erythematous papules to nodules over scalp in third case whichisdescribed by polarizeddermoscopy of the lesions over the scalp; with contact plate; 50x magnification [Dinolite AMZ413ZT], b. Papulo-nodularlesion over the ala of leftnostril, in the second case, c. Twoerythematous nodules over the right concha, in the first case, d. Polarizeddermoscopy of the right conchallesionshowing diagonal and orthogonal linesarrangedperpendicularly – Chrysalis-like structures, using immersion oil as a contact medium; 50x magnification [Dinolite AMZ413ZT], e. Polarizeddermoscopy of the lesion over the right nostril, with contact plate; 12x magnification [Dinolite AMZ413ZT]

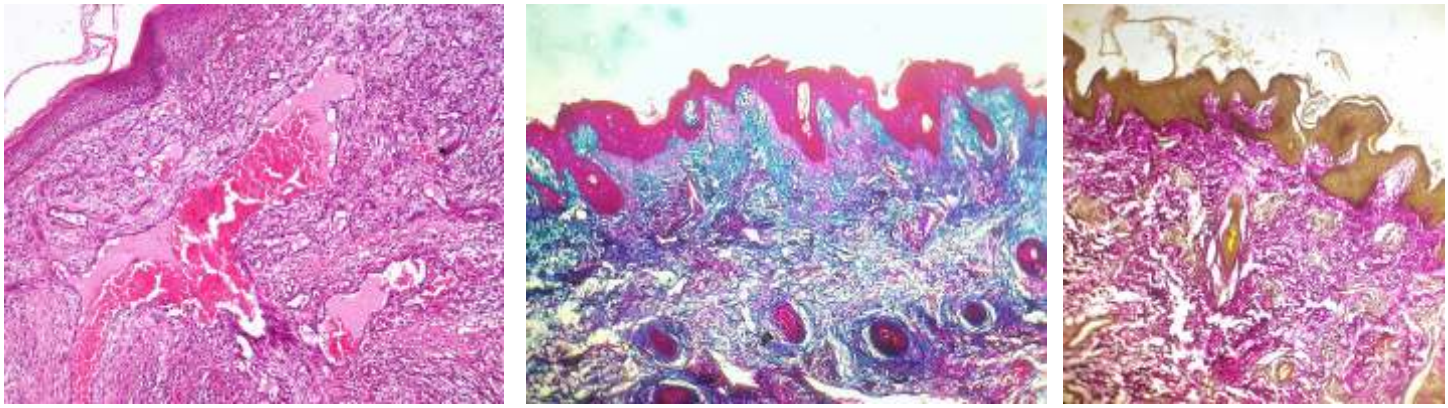

Figure 2 a,b,c : a. Multiple proliferating bloodvessels, with plump endothelial cells, with abundant extravasation of RBCs and few eosinophils in the dermis and haphazardly arranged collagen fibres ; 20x Hematoxylin-Eosin ,b. Haphazardly arranged collagen fibres and bundles were noted, in both vertical and horizontal orientations; 10x Masson trichome histochemical stain and on, c. Verhoeff-Van Gieson histochemical stain 10x

All three cases underwent a skin biopsy to confirm the diagnosis. Hematoxylin and eosinstaining revealed multiple proliferating blood vessels with abundant extravasation of RBCs and a few eosinophils in the dermis. These changes were consistent with ALHE. Haphazardly arranged collagenfibers and bundles were noted, in both vertical and horizontal orientations, which was confirmed on staining with Verhoeff Van-Geison and Masson's trichrome (Figure-2). This disorientation contributes to the Chrysalis-like pattern.

We analyzed the dermoscopic pictures by Lomba et al and noticed features consistent with the Chrysalis sign, and we discussed the same with the principal author. This sign should be added to the dermoscopic constellation of ALHE, to further knowledge on this enigmaticentity.

How to cite this article:

Sharma S, Dhurat S, Vishwanath T, Agarwal S, Jasrania D, Sharma R. Chrysalis sign– A new dermoscopicentity in the diagnosis of angiolymphoid hyperplasia with eosinophilia. JDA Indian Journal of Clinical Dermatology 2020;3:15-16

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CASE REPORT
Year: 2019 • Volume: 2 • Issue: 3 • Page: 17-19

SKIN BLISTERING ASSOCIATED WITH SEVERE SCARRING AND PHOTOSENSITIVITY AFFECTING TWO SIBLINGS - KINDLER SYNDROME OR DYSTROPHIC EPIDERMOLYSIS BULLOSA? – A CASE REPORT

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Abstract

Kindler syndrome is an autosomalrecessive inherited condition characterized by acral bullae, progressive poikiloderma, photosensitivity along with mucosal involvement. Kindler syndrome and dystrophic epidermolysis bullosa, both can have similar clinical presentation and it may be difficult to differentiate them from each other, especially in neonatal period. We are reporting here the case oftwo siblings having photosensitivity with poikilodermatous changes, acral blistering and mucosal involvement, features clinically consistent with Kindler syndrome.

Key words: Skin blistering,Photosensitivity,Dystrophic epidermolysis bullosa,Kindler syndrome.

Introduction

Dystrophic Epidermolysis Bullosa (DEB) and Kindler syndrome are genetic blistering disorders which are characterized by traumatic blistering, which usually starts at birth or in infancy. The fragility of skin results in blisters appearing at trauma prone sites and these lesions usually heal with secondary changes like scarring, milia formation and nail changes. Dystrophic epidermolysis bullosa has both dominant and recessive pattern of inheritance and is characterized by a defect in collagen 7 protein which causes defect in the sublaminadensa, while Kindler Syndrome has recessive pattern of inheritance and is characterized by defect in Kindlin 1. In addition to the fragile skin, teeth and nail changes seen in DEB, Kindler syndrome has progressive poikiloderma, photosensitivity and mucosal inflammation also.[1,2]

Case Report

A 23-year-old female borne out of non-consanguineous marriage presented to our out-patient department with history of clear fluid filled lesions rupturing to leave behind crusted raw areas all over the bodysincethe age of 3 months, more over areas prone to trauma, like knees, elbows and dorsa of feet and hands. She also gave history of photosensitivity with burning sensation on exposure to sun and fluid filled lesions developing at the sites of sun exposure. There was also a history of similar complaints in her younger sister. On examination, patient had multiple well defined irregular shaped erosions with overlying crust present over both shoulders, elbows and knees. [Figure 1] There were multiple areas of atrophic scarring over both elbows, knees, dorsa of hands and feet with mild poikilodermatous changes.

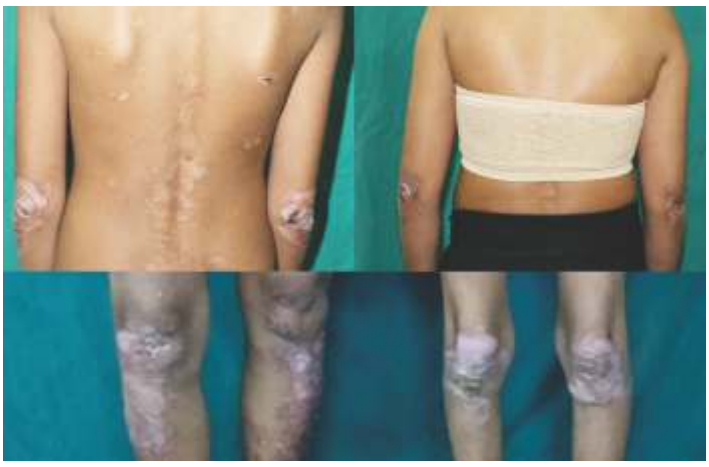

Figure 1 : Multiple well defined irregular shaped erosions with overlying crust present over bilateral elbow and knee

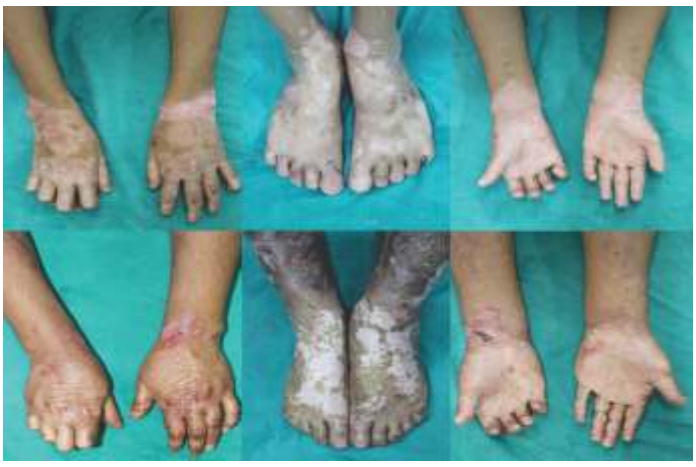

Figure 2 : Atrophic scarring over both elbows, knees, dorsa of hands and feetwith mild poikilodermatous changes, clawing of both hands and anonychia over bilateral fingers and toes

Figure 3 : Oral cavity showing multiple erosions with overlying slough over the hard palate and cheilitis

There was contracture with resultant immobile clawing of both hands and anonychia over bilateral fingers and toes. [Figure 2] In the oralcavity, there were multiple small erosions with overlying white slough over the hard palate and bilateral buccal mucosa; along with cheilitis. [Figure 3] She also had multiple, well defined, and flat topped, dark brown to violaceous atrophic papules and some plaques, predominantly over the forehead, upper chest and back and milia were present over both helix of ear. [Figure 4] The teeth were normal and there were no lesions on the scalp.

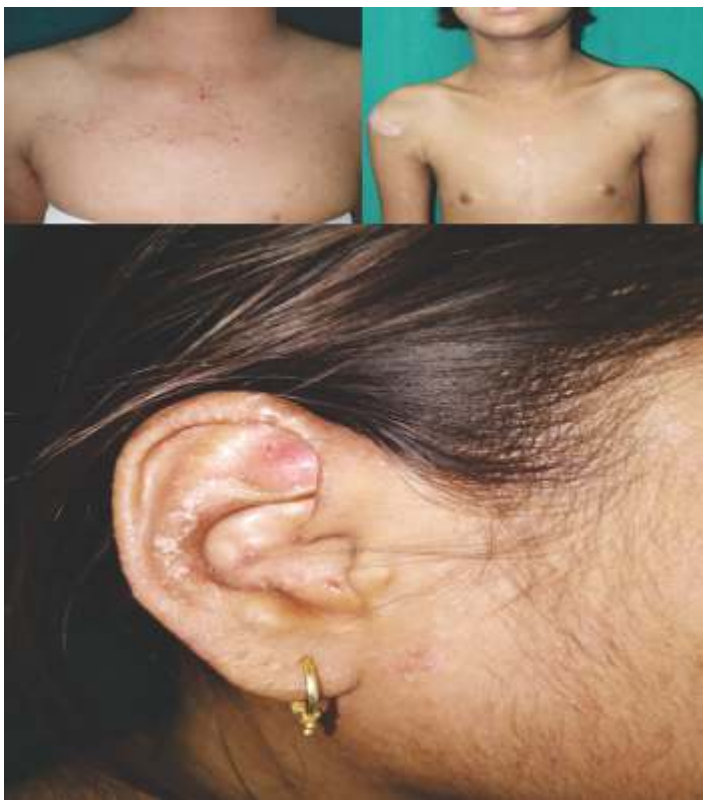

Figure 4 : 4Well defined dark brown to violaceous atrophic papules and plaques, over upper chest and back; milia over helix of ear

Routine hematological investigations were normal and a shave biopsy was sent from one of the lesions over her chest. Histopathological examination showed basal cell degeneration and split formation at multiple level at dermo epidermal junction along with melanophages in the dermis on hematoxylin and eosin stain (H&E). [Figure 5] Electron microscopy and genetic analysis could not be done because of resource constraints.

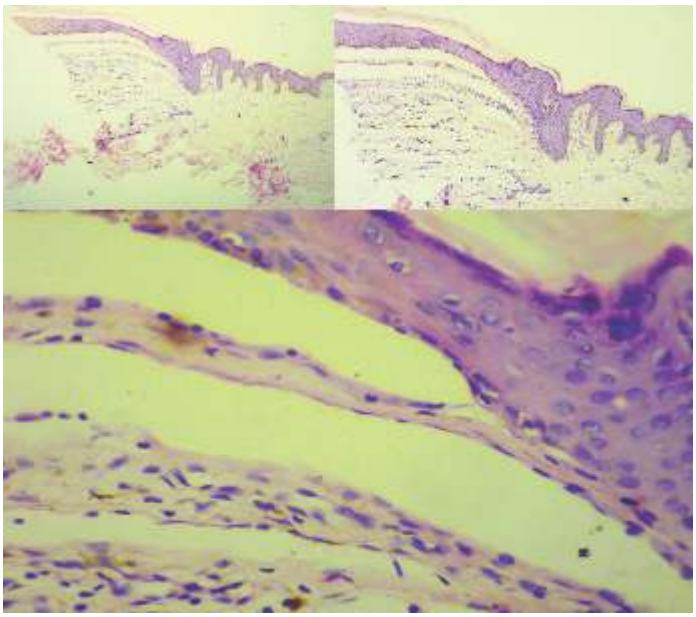

Figure 5 : Basal cell degeneration with split at multiple level at dermo epidermal junction along with melanophages in the dermis. (H&E stain 40x,100x,400x)

Discussion

Kindler syndrome was first described in 1954 by Theresa Kindler in a child with acral blistering, pigmentary changes and photosensitivity. The KIND1 gene encodes the protein Kindlin 1 which connects the actin cytoskeleton to the extracellular matrix. Recently, a novel mutation in FERMT1 gene has also been discovered.[3]Both dystrophic epidermolysis bullosa and Kindler syndrome can have similar clinical presentation and it may be difficult to differentiate them from each other, especially in neonatal period. There are many features which may help in differentiation, but the definite diagnosis can however be made after molecular studies only [Table 1].[4]

In Kindler syndrome, photosensitivity and acral blistering decrease with age, and appearance of poikiloderma and cutaneous atrophy tend to gradually worsen. Atrophic changes which typically appear as cigarette paper like wrinkled skin are most prominent over the sun exposed areas, most commonly site being the dorsal aspect of the hands and feet but may become generalized by adolescence. Mucosal involvement is common in both and may present as ectropion, corneal erosions, gingival inflammation, periodontal disease and scarring of the external urethral meatus. In Kindler syndrome, additional oral mucosal

findings include advanced periodontal bone loss, and leukokeratosis of buccal mucosa, trismus and a form of desquamative gingivitis.[5] Other less common features include ichthyosis, palmoplantar hyperkeratosis, light colored hair, pseudoainhum, nail changes including dystrophy and long and thick cuticle of nail,and an increased susceptibility to the development of squamous cell carcinoma.[6-9] The hands may develop pseudosyndactyly, similar to some cases of dystrophic EB. Chronic colitis may complicate the cases of Kindler syndrome sometimes.

The initial diagnosis relies upon careful clinical examination, family history and establishing the level of blister formation. Traditionally, transmission electron microscopy and immunofluorescence microscopy using a panel of antibodies against the candidate proteins implicated in EB are the preferred methods. The main objectives of skin biopsy are first to establish the level of blistering or tissue separation and, second, to search for other clues that may be indicative of the underlying disorder and therefore helpful in the diagnosis. Histopathologic examination alone cannot distinguish between dystrophic epidermolysis bullosa and Kindler syndrome. Kindler syndrome, shows variable plane of cleavage or duplicated lamina densa as compared to specific cleavage planes in epidermolysis bullosa. In addition, there may be epidermal atrophy, basal layer vacuolization, variable epidermal melanin content, dermal melanophages and capillary dilatation, which is more prominent in older patients when the poikiloderma sets in.[10] Immunostaining with anti-kindlin 1 antibody shows decreased staining of the epidermis in KS.[11]

Diagnostic criteria have been proposed by Fischer et al. and the presence of four major criteria makes a diagnosis of KS. The major criteria include:[12]

1. Acral blistering beginning in infancy

2. Progressive poikiloderma,

3. Cutaneous atrophy

4. Photosensitivity,

5. Fragility and/or swelling of gums.

The minor criteria include syndactyly and involvement of other mucosae. Additional features including palmoplantar keratoderma, ectropion, pseudoainhum, oral leucokeratosis, squamous cell carcinoma, onychodystrophy, skeletal abnormalities, and dental problems may be seen. The diagnosis is "confirmed" if four major criteria are fulfilled, as seen in our patient. The presence of three major and two minor criteria makes a "probable" diagnosis, while presence of two major and two minor/additional features makes the diagnosis "possible".

We have presented this case because of considerable overlap between these two conditions, and rarity of Kindler syndrome; it is important to diagnose Kindler syndrome because photo protectionto prevent any cutaneous malignancy needs to be advised to the patient along with management akin to dystrophic EB.

How to cite this article:
Agarwal P, Jagati A, Arwindebrar, Saikia S. Skin Blistering Associated With Severe Scarring And Photosensitivity Affecting Two Siblings - Kindler Syndrome Or Dystrophic Epidermolysis Bullosa? – A Case Report. JDA Indian Journal of Clinical Dermatology 2020;3:17-19

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CASE REPORT
Year: 2020 • Volume: 3 • Issue: 1 • Page: 20-21

A SKIN COLOURED PLAQUE OVER BACK - MIMESIS OF BENIGN TUMOURS

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Abstract

Fibroepithelioma of Pinkus (FEP) is an uncommon tumour in Indian population. FEP clinically resembles a variety of benign skin tumors which are not routinely excised or biopsied. Diagnostic confirmation is necessary becausethe nature of this tumor remains a subject of debate.

Key words: Fibroepithelioma of Pinkus (FEP)

Introduction

Fibroepithelioma of Pinkus (FEP) is rare and often misdiagnosed clinically as it imitates multiple benign skin tumours. Some authors argue that the relative rareness of this tumour is mainly because of its misdiagnosis. Differential diagnosis in case of FEP is dermal nevus, fibroma, acrochordon, and seborrheic keratosis, which are not routinely excised or biopsied. Dermoscopy and confocal microscopy have emerged as non-invasive tools but histopathology remains the gold standard for diagnosis.

Case report

An 89-year-old male, presented with an asymptomatic skin colored to reddishlesion over back for 6 years. The Lesion had slowly grown to present size over a period of 6years.

Examination revealed a solitary, non-tender skin coloredplaque of size 4×3 cm with non-tender central erythematous nodule.Surrounding the plaque there was a rim of brown-black hyperpigmentation present. [Figure1]

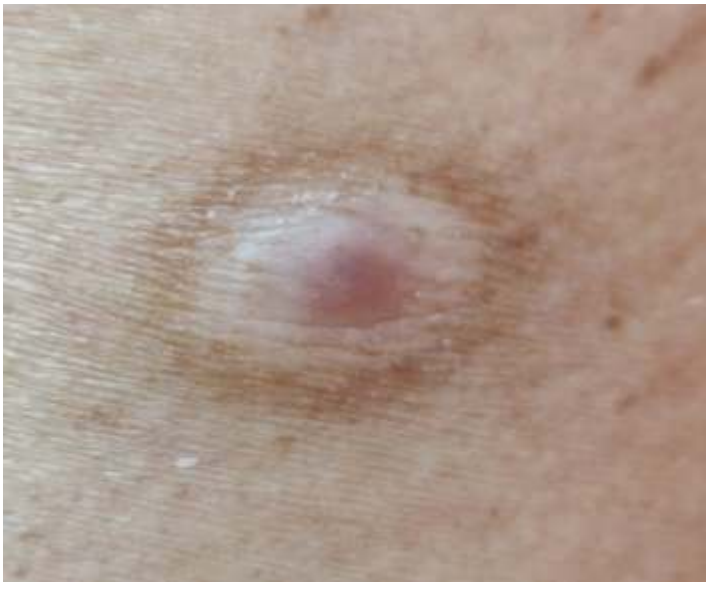

Figure 1 : A solitary, non-tender skin coloured plaque of size 4×3 cm with non-tender central erythematous nodule with a rim of hyperpigmentation.

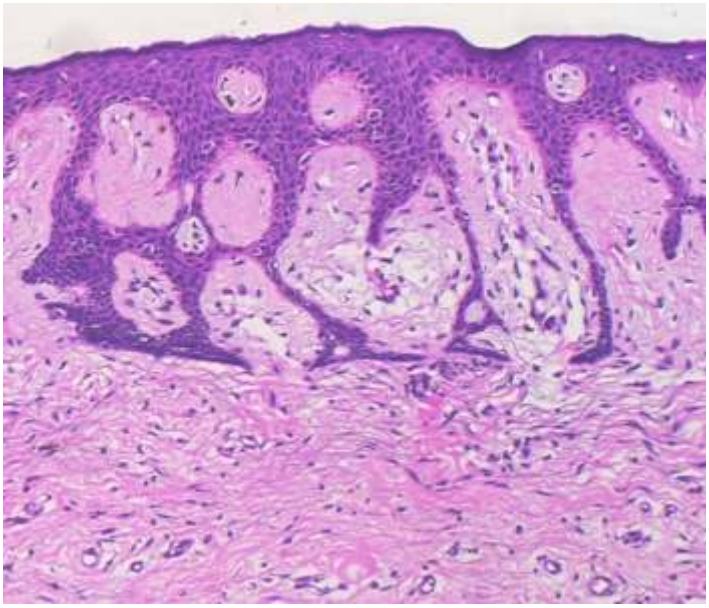

Figure 2 : Epithelial neoplasm that shows a connection with surface epidermis at several places.

A clinical differential of appendegeal tumor, intradermal nevi was kept in mindand punch biopsy was takenfor further confirmation.

Histopathological examination revealed epithelial neoplasm that shows a connection with surface epidermis at several places [Figure2]. The tumour islands were made up of small round cells that show uniformly stained and dark nuclei no nucleoli.The neoplastic cells were arranged in a proliferation of thinned out rete ridges with small bulbous ends that show hair germ like appearance[Figure3].The surrounding stroma shows fibroplasia [Figire4]. There were no clefts between the stoma and the epithelium.

Discussion

Fibroepithelioma of Pinkus was first described by Hermann Pinkus. It presents as flesh-colored to a pink solitary, sessile or pedunculated papule, plaque or nodule on the trunk often resembling a fibroma, intradermal nevus, seborrhoeic keratosis, achrochordon. But large pedunculated, polypoid or ulcerated

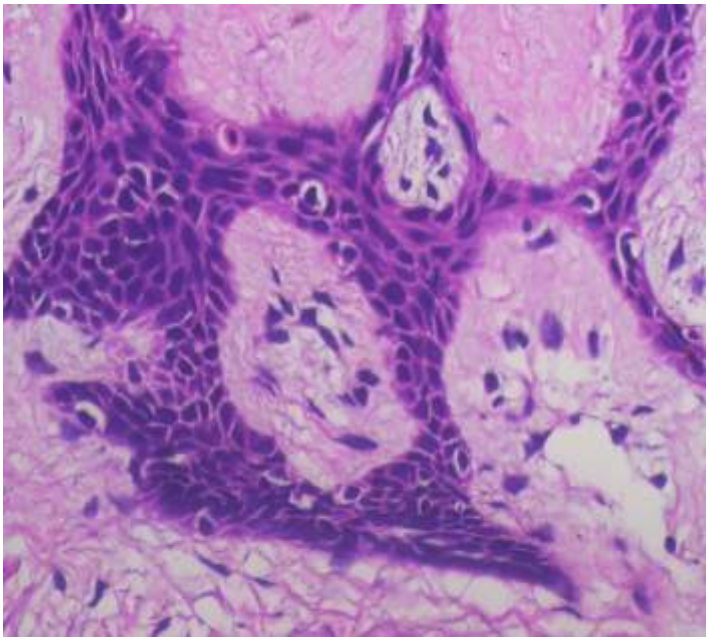

Figure 3 : The neoplastic cells are arranged in a proliferation of thinned out rete ridges with small bulbous ends that show hair germ like appearance.

cases have also been reported . It is commonly reported to occur in adults aged 40–60 years. However, a few cases in the pediatric population have been reported .The strong predilection for the lumbar region and for the female gender differentiates FEP from other BCC subtypes that usually arise on sun-exposed areas andpredominantly in males .

Dermoscopy of FEP demonstrates white streaks and fine arborizing vessels with occasionally dotted vessels. When pigmented, features of FEP also include structure-less gray-brown areas and blue-gray dots. The hallmark of confocal microscopy is a fenestrated pattern demonstrating dark "holes" that correspond to the fibrous stroma surrounding tumour strands .

The gold standard diagnostic test for FEP is a skinbiopsy for histopathologic examination. It consists of basaloid cells arranged in cords and columns, which anastomose with each other. Stroma surrounding these cords and columns is thick and rich with fibrocytes. These columns and cords may show palisading and follicular germ-like structures formation. Many of the cords and columns show connections with the surface epidermis .

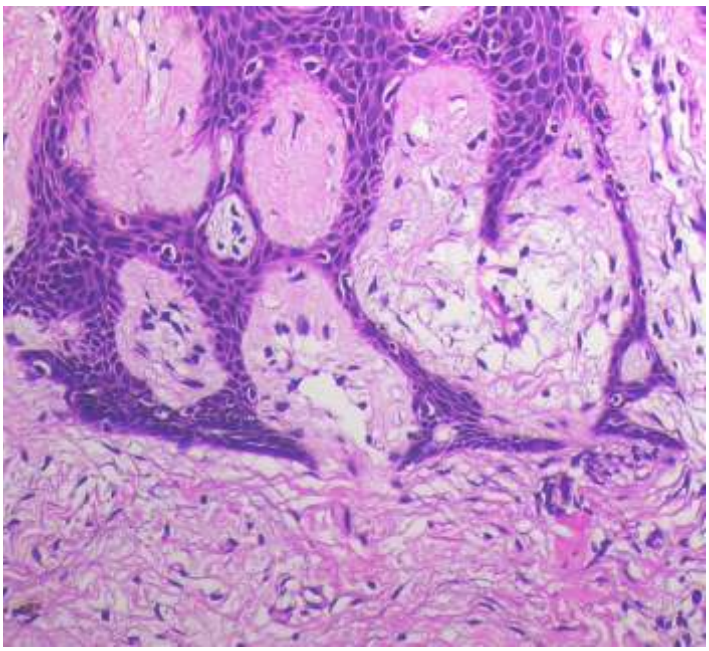

Figure 4 : The surrounding stroma shows fibroplasia.

Hermann Pinkus initially classified it as a premalignant fibroepithelial tumor .Later on, FEP was classified as a subtype of basal cell carcinoma, although some consider FEPas a variant of trichoblastoma . Some consider it as a benign and unusual counterpart of BCC, while others think it a malignant neoplasm .Finding such as expression of androgen receptor is shared by BCC and FEP, which is minimal in trichoblastomas; whereas FEP and trichoblastomas display Merkel cells, which are usually absentin BCC . In another study, authors found that tumour-specific type of epithelial hyperplasia in FEP that is positive for the stem cell marker PHLDA1 and present a unifying concept of FEP as a subtype of BCC.

Recent research highlights the fact that bcc and trichoepithelioma share common cell origin,which is the epithelial stem cells of the hair follicle. So it is proposed that FEP might be a trichoblastic tumor intermediate between trichoepithelioma and bcc .

There are case reports showing an association of nodular bcc with FEP supporting its malignant nature . So its classification still remains controversial.

Local excision of FEP is recommended treatment of choice and is almost universally curative without recurrence. Our patient 89-year old male also presented as an asymptomatic plaque with central nodule over back. Dermoscopy or confocal microscopy was not done in our case but histopathological examination did demonstrate classic histopathologic features for FEP (Figure 2,3,4). The morphological resemblance to benign skin tumours is one of the cause for its misdiagnosis and relative rareness. In the light of controversial malignant nature of FEP,early diagnosis and curative treatment with complete local excision are mandatory.

How to cite this article:
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CASE REPORT
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A CASE OF ACCIDENTAL METHOTREXATE TOXICITY PRESENTING AS MUCOCUTANEOUS ULCERATION

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Abstract

Methotrexate is a commonly used drug in treatment of psoriasis. However, it has a very steep safety profile and, therefore, its usage requires following strict guidelines. Here we are describing a case of acute methotrexate toxicity occurring in a patient of chronic plaque psoriasis following inappropriate self-medication of the drug leading to development of ulceration in psoriatic plaques, mucositis and bone marrow suppression treated with folinic acid.

Key words: Methotrexate toxicity, acute toxicity, psoriasis, skin ulcer.

Introduction

Methotrexate (MTX), formerly named as Amethopterin,is an antiprolierative and immunosuppressive drug which inhibits mitosis. [1,2]After entering the cell, MTX acts as a Dihydrofolate (DHF) reductase inhibitor.[3] This enzyme coverts DHF to tetrahydrofolate (THF). Its inhibition leads to DNA inhibition. It is used at higher doses (0.5-12g/m2) as an anticancer drugwhile at lower concentrations (1-40 mg/m2) it is effective in inflammatory diseases like Rheumatoid Arthritis and Psoriasis to name a few.[4] There are various reports of chronic long-term use of the drug associated hepatic and pulmonary toxicity.[5] But there is still a dearth of reported cases on acute MTX toxicity presenting as cutaneous erosions and mucositis complicated by bone marrow suppression.We are reporting this case for the rarity of its occurrence in literature.

Case Report

A female patient aged 29-years, with an 18-months history of plaque-type psoriasis involving bilateral lower extremities, buttocks, trunks and elbows, was taking MTX for 7 months. But instead of physician prescribed three consecutive dosages of 5mg at interval of 12 h per week, the patient accidentally started consuming 5mg MTX once a day every day throughout the week.Fifteen days later the patient presented to the emergency department with hemorrhagic erosions and crusting over preexisting plaques of psoriasis, with painful mucosal lesions chiefly involving the lower lip. Patient also had fever with malaise.

On admission patient was conscious with respiratory rate 30/minute, pulse 96/minute, blood pressure 142/90mmHg and temperature 99˚F. On physical examination around 10% of the body surface area was involved with presence of ulcerated psoriatic plaques over bilateral lower limbs (figure 1), buttocks (figure 2) and elbows. Patient also had hemorrhagic mucositis of oral mucosa chiefly involving the lower lip (figure 3). There were no positive findings on examination of other organs.

On laboratory investigation the patient's red blood cell count was 3.25µL, hemoglobin was 9 g/dL, white blood cell count was 450/µL and the platelet count was 9,000/µL. Biochemical investigations revealed blood urea nitrogen 32.0 mg/dL, serum creatinine 0.9mg/dL, total bilirubin 2.0mg/dL, direct bilirubin 1.4mg/dL, alkaline phosphatase 180 U/L, SGOT 200IU/L and SGPT 250 IU/L. Patient's bleeding time, clotting time and prothrombin time were within normal limits.

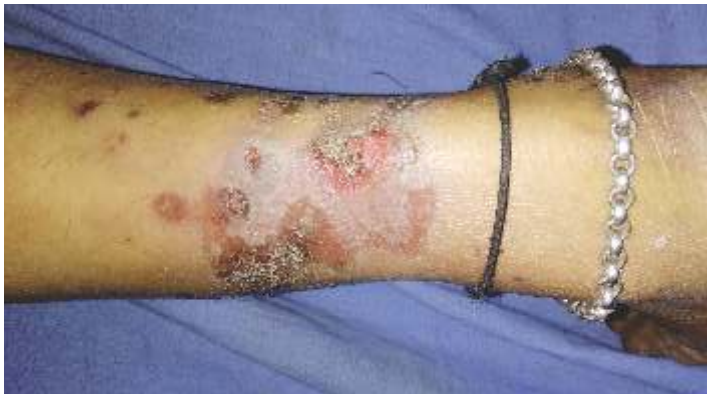

Figure 1 : Ulceration of psoriatic plaques over lower limb

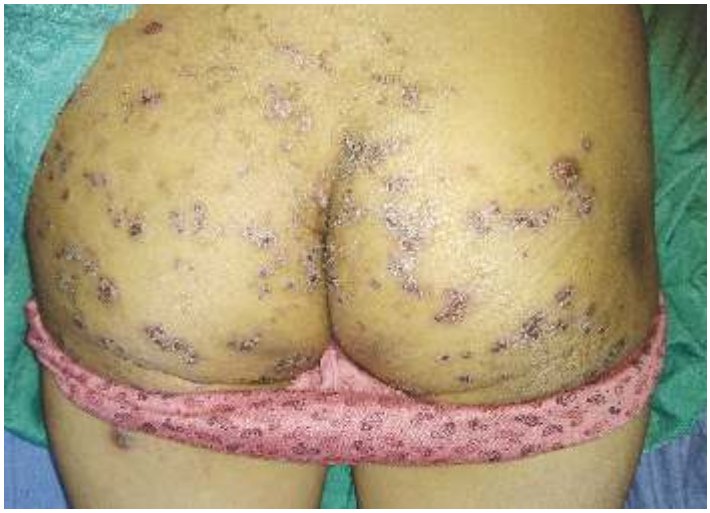

Figure 2 : Ulceration of psoriatic plaques over buttocks

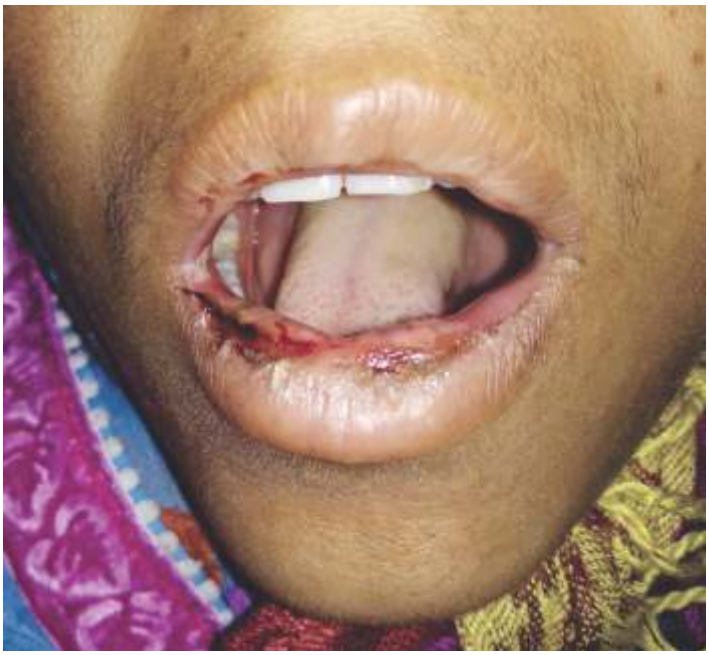

Figure 3 : Ulceration over labial mucosa

The patient was diagnosed with acute MTX toxicity. MTX was stopped immediately and intravenous folinic acid (as MTX antidote) in the dose of 15 mg was given 6 hourly (1mg/kg) on first day followed by 10 mg 6 hourly until counts reverted back to normal. Patient's intravenous fluid intake was maintained optimally according tourine output and antibiotics were started empirically. The eroded psoriatic plaques were taken care of with topical antibiotics. Mucositis was dealt with a suspension prepared according to Pandya's formula consisting of a mix of 5 mL each of syrup prednisolone (5mg/ml), syrup Gelusil and syrup Benadryl. The patient was asked to rise the mouth with this suspension for 2 minutes before spitting it out.[7] This was repeated 3-4 times a day until oral lesions started resolving. Patient's urine output started improving after 24 hours and at 7th day patient's antibiotics were stopped in absence of any active infective foci. Patient's vitals were repeatedly reported everyday.

Discussion

MTX toxicity has been reported to occur in both idiosyncratic and dose-dependent manners. Overdosing of MTX is considered to be the most common cause of acute MTX toxicity [6] It can appear with varying manifestations ranging from vomiting, nausea and diarrhea to pancytopenia, myelosuppression, pulmonary, hepatic and renal dysfunction, cutaneous ulcerations, mucositis, stomatitis as well as gastrointestinal ulcerations.[7] Amongst all of these, ulceration of pre-existing lesions is considered as one of the early signs of impending MTX toxicity in Psoriasis patients. [8] The cells observed to be affected the most by MTX are those with high turn over like epidermal, gastrointestinal and bone marrow cells. The effect of MTX on these cells is dose dependent. Drugs like NSAIDs decrease renal elimination and tubular secretion of MTX MTXwhile trimethoprim/sulfamethoxazole contributes of cytotoxicity caused by MTX since trimethoprim is also an antifolatereducatase inhibitor.MTXconcertation in blood and urine can be measured by radioimmunoassay.A confirmative diagnosis can be reached by histological analysis of affected sites.[2] In an ideal setup, any patient presenting with MTX toxicity should be kept in an intensive care unit with reverse barrier nursing. [6] The drug should be stopped as soon as possible followed by prompt treatment with intravenous Folinic Acid (Leucovorin).[2]Lucovorin bypasses the enzyme dihydrofolate reductase and salvages normal bone marrow cell division. The dose of Lucovorin should be titered according to serum MTX level.[6] In case of significant neutropenia granulocyte colony stimulating factors should be used. Thrombocytopenia is treated with packed red cells of platelets transfusion. Other supportive measures include intravenous fluids,antiemetics and urine alkalizers. In case of impaired renal function Glucarpidase, in the dose of 50 U/kg body weight as a single intravenous injection, can be used to convert MTX into its inactive metabolites.[9] Hemodialysis and hemoperfusion might be required is serious cases of toxicity. The use of corticosteroids in MTX toxicity is debatable since there is a scarcity of large trials suggestive its efficacy and it may also lead to superinfections.[10] The eroded plaques incase of MTX toxicity in psoriasis mostly require just supportive care and heal in 1-2 weeks.Mucositis is treated by proper oral care and topical coating agents like Sucralfate suspension. Specific mixture consisting of a mix of 5 mL each of syrup prednisolone (5mg/ml), syrup Gelusil and syrup Benadryl, also known as Pandya's formula, is also used for rinsing.[6]

Eventhough, if not diagnosed and treated timely, MTX toxicty can be lifethreatening, prompt treatment of patients in ICU and emergency department can reduce mortailty rates. Proper patient councelling before presciribing MTX can all together eliminate such accidents.

How to cite this article:
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CASE REPORT
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MILIA EN PLAQUE

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Abstract

Discrete milia are common whereas milia en plaque is rarely encountered entity. Althoughreason for postauricular localization in not yet understood, repeated trauma could be predisposing factor.

Key words: Milia , Retroauricular, Middle age

Introduction

Milia are subepidermal keratin cysts that can be seen in all age groups from neonate to elderly. Milia can be primary or secondary to various skin disorders. It frequently appears over cheeks and eyelids.[1] Although discrete milia are common, milia en plaque is rarely encountered. Here, we are reporting two patients where milia en plaque found on retro-auricular skin.

Case 1

A 32-year old male, farmer by occupation, presented to dermatology OPD with multiple skin coloured lesions on retro-auricular skin, present for last three years. Patient denied use of eye glasses, any topical cream or any possible trauma. Clinical examination revealed 3 x 2 cm size plaque composed of multiple yellowish papules on retroauricular skin (Figure 1a,b). Fig 1a,b

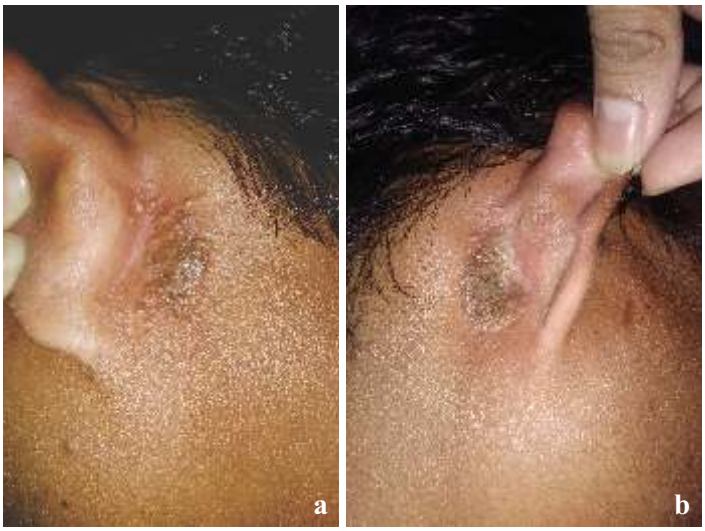

Figure 1a : Erythematous plaque studded with multiple shiny yellow colored milia over left retroauricular area.
1b : Milia en plaque at right retroauricular space

Histopathological examination revealed multiple concentric lamellar keratin filled cysts lined by keratinized stratified squamous epithelium in dermis (Figure 2a). Mature or fully developed cysts were lined by single layer whereas evolving cyst lined by multiple layers. Peri-cystic and peri-appendegeal mononuclear cell infiltration was noted[1] (Figure 2b). Diagnosis of milia en plaque was made and patient was asked for local application of tretinoin 0.025% cream. Patient was however loss to follow up.

Case 2

A 35-year-old woman, presented with closely aggregated milia on left retroauricular skin (Figure 3). Lesions were present for last six months. She gave history of wearing large ear-ring for 1 years. Cutaneous Examination showed 2x2 cm skin colored plaque composed of multiple milia with shiny yellowish surface. Patient did not give consent for histopathological examination. On the basis of morphological examination, diagnosis of milia en plaque was made.

Discussion

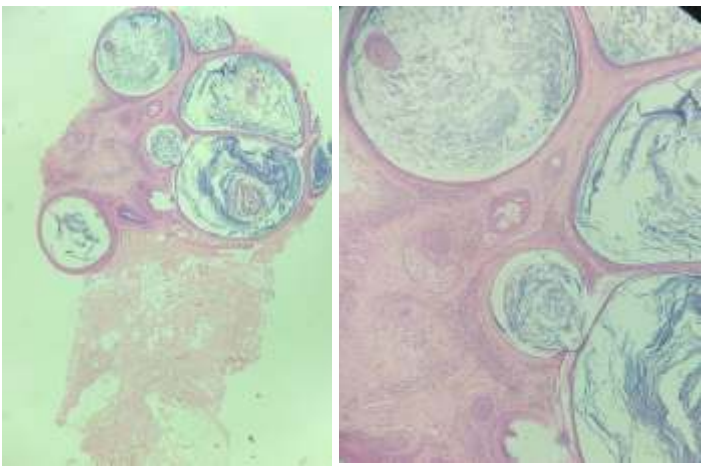

Figure 2a : 10x view of HPE of milia en plaque with multiple lamellar keratin filled cyst in superficial and deep dermis.
2b : 40x view of multiple cyst lined by single or multiple layer with peri-cystic and peri-appendegeal mononuclear cell infiltration

Milia can be considered primary appearing spontaneously, or secondary, caused by skin disease, trauma, or medications.

Primary milia are small epidermoid cyst, 1-4mm in diameter , arise from lowest part of infundibulum of vellus hair follicle at the level of sebaceous gland. They are fixed and persistent. Primary milia may occur congenitally or shortly after birth in up to 50% newborns. Favourable sites are the face, especially nose, scalp, proximal extremities and upper trunk. They resolve over weeks.[2]

Secondary milia arises from eccrine duct or hair follicle, in order

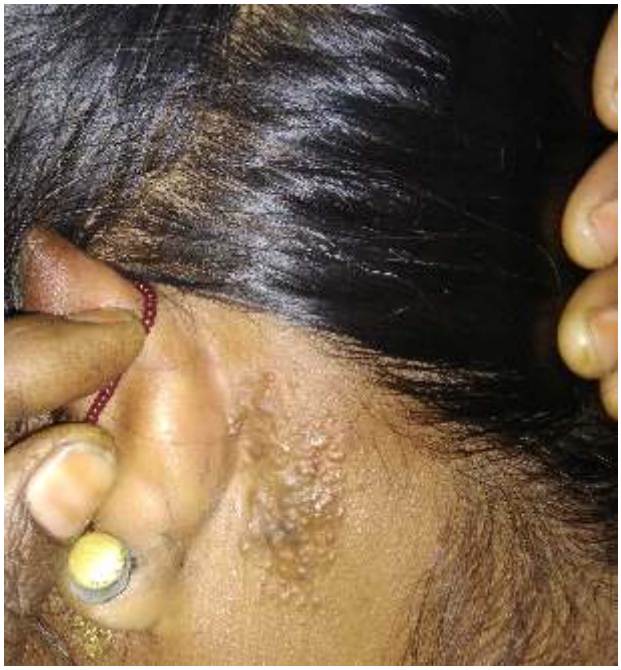

Figure 3 : Skincolored plaque with grouped milia forming milia en plaque over left retroauricular area.

to re-epithelise the eroded epidermis. They are transient and spontaneously disappear. Milia on palate are called Epstein pearls. Secondary milia can develop as a result of blistering skin disease, they also tend to occur after trauma such as dermabrasion, chemical peeling, ablative laser, skin grafting and radiotherapy[2]. Long term use of topical corticosteroids and occlusive moisturizer can cause milia. Cyclosporine and 5-FU have been associated with development of milia.[3]

Milia en plaque is an inflammatory variant of milia. It shows predilection for the head and neck area, especially periauricular and periorbital area[4].This condition is more common in middle aged women.[2]

In both our cases lesions of Milia en plaque were present on retro-auricular skin.

First case mentioned here, had no prior risk factors like pre-existing skin lesions or any chronic trauma. It is possible that in second patient, history of wearing large ear rings might have acted as precipitating factor.

Milia en plaque is not common and reported only sparingly in literature. This prompted us to report these cases.

How to cite this article:
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