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Original Research Article

A clinico-epidemiological study of various keratinization disorders

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ABSTRACT

Background: There is a vast spectrum of disorders with basic defect in the process of keratinization. There are various associations (genetic, autoimmune and environmental) with different keratinization disorders. The aims and objectives of this study to study the epidemiology, clinical features and associations in various keratinization disorders.
Methods: A retrospective observational study of 500 patients was done in a tertiary care center. Detailed history was taken and clinical examination was done. Investigations and skin biopsy were performed when needed.
Results: In our study of 500 cases of keratinizing disorders, there were 269 (53.8%) cases of psoriasis, 132 (26.4%) cases of palmoplantar keratoderma, 22 (4%) cases of phrynoderma, 19 (3.8%) cases of ichthyosis, 13 (2.6%) cases of acanthosis nigricans, 11 (2.2%) cases of porokeratosis, 7 (1.4%) cases of Darier's disease, 3 (0.6%) Cases of pityriasis rubra pilaris, 2 (0.4%) cases each of pachyonychia congenita and erythron keratoderma. The most common age group affected was 51-60 years (19.6%). Males to female ratio was 1.13:1. Chronic plaque psoriasis (43.51%) was the most common variant of psoriasis. Psoriasis vulgaris (75%) was the most common cause of erythroderma. Histopathological findings in all patients whose biopsy was taken was consistent with clinical diagnosis. Non trans gradient (97.75%) was the most common type of palmoplantar keratoderma. Ichthyosis vulgaris (47.38%) was the most common type of ichthyosis.
Conclusions: Heredity plays an important role in keratinization disorders. Also, various comorbidities have been associated with different keratinization disorders. Hence, we need to look for these factors while evaluating the patients of keratinization disorders.

Keywords: Keratinization disorders, Psoriasis, Palmoplantar keratoderma, Comorbidities

INTRODUCTION

The term 'disorders of keratinization' refers to broad spectrum of skin disorders where there is abnormal differentiation of the epidermis and/or appendages, often with aberrant formation of the cornified envelop. These disorders may have associated with epidermal fragility, and there is clinical and pathological overlap with inherited and acquired blistering disorders.[1]

Epidemiological studies revealed that a distinct group of disease is quite frequently associated with psoriasis, e.g. Arthritis, colitis, diabetes and hypertension.[2] However, there is scanty data regarding the demographic profile and clinical features of patients with congenital Icthyosis in india.[3]

Aims and objectives

The aim of this study, the epidemiology, clinical features and associations in various keratinization disorders.

The objectives of this study were to find out various keratinization disorders in the sample taken in the study,

to compare the results of present study with various other studies and to find the presence of various comorbidities in keratinization disorders.

METHODS

A retrospective observational study of 500 patients of keratinization disorders attending the outpatient department of dermatology, venereology and leprosy, PDU Government Medical College and Hospital, Rajkot during year December 2017 to November 2019.

With informed consent, Patients personal data like age, sex, marital status, education were collected, detailed history was taken including duration of the disease, joint pain, smoking, alcohol intake, tobacco consumption, concomitant illnesses, concomitant drug intake, consanguinity and family history of similar disease. Thorough clinical examination was done.

Investigations included complete blood count (hemoglobin, total count, differential count, platelet count), C reactive protein, erythrocyte sedimentation rate, urine routine and microscopic examination, liver function tests (S. bilirubin, alanine transferase, aspartate transferase levels), renal function tests (blood urea, serum creatinine), serum total protein and albumin, serum calcium, blood sugar estimation, thyroid function tests (T3, T4, TSH levels), fasting lipid profile, ultrasound of abdomen. In clinically difficult cases, biopsy was done for diagnosis. Specific investigations included: electrocardiogram (ECG) in patients with history of cardiovascular risk factors; chest X-ray with history of respiratory complaints and referred to physician if abnormal; X-ray of involved joint, rheumatoid factor and S. uric acid with history of joint pain. In patients of ichthyosis; ultrasound of abdomen, audiometry to access hearing loss and neurologic checkup was done to rule out systemic involvement.

Data was collected and results (tables and graphical representation) were obtained by using MS excel version 2016.

RESULTS

The maximum number of patients at the time of presentation belonged to the age group of 51-60 years (19.6%) (Table 1). Male: female ratio was found to be 1.13:1.

Out of 500 patients of keratinization disorders, psoriasis (54%) was the most common type of keratinization disorder followed by palmoplantar keratoderma (26.4%) (Figure 1).

In 289 patients of psoriasis, maximum number of patients belonged to 51-60 years age group (24.6%), followed by 41-50 years (23.2%) (Table 2), with male: female ratio was 1.5:1. Out of all the variants of psoriasis, chronic plaque psoriasis was the most common clinical type affecting 75.8% patients followed by scalp psoriasis 6.6%, palmoplantar psoriasis 4.5%, guttate psoriasis 2.8%, inverse psoriasis 2.4% and pustular psoriasis 1% (Figure 2).

Table 1: Age wise distribution of keratinization disorders.

Age (in years)	No. of cases	Percentage
<1	4	0.8
1-10	30	6
11-20	45	9
21-30	85	17
31-40	93	18.6
41-50	94	18.8
51-60	98	19.6
>60	51	10.2
Total	500	100

Table 2: Age wise distribution of psoriasis disease of present study (2016) (n=289).

Age (in years)	No. of patients (%)
<1	0
1-10	8 (2.7)
11-20	15 (5.2)
21-30	42 (14.5)
31-40	55 (19)
41-50	67 (23.2)
51-60	71 (24.6)
>61	31 (10.7)
Total	289 (100)

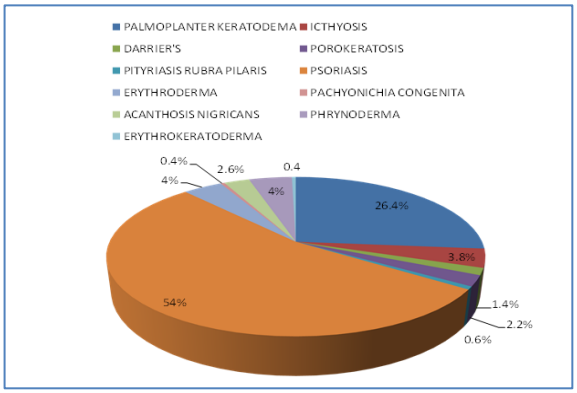

Figure 1: Overall pattern of various keratinization disorders.

Amongst systemic comorbidities associated with psoriasis, we found that maximum number of patients were obese (36.68%) followed by hypertension (31.81%) (Table 3).

All the nail changes in psoriasis patients, we found pitting (39%) was most common followed by longitudinal ridges 21% (Figure 3).

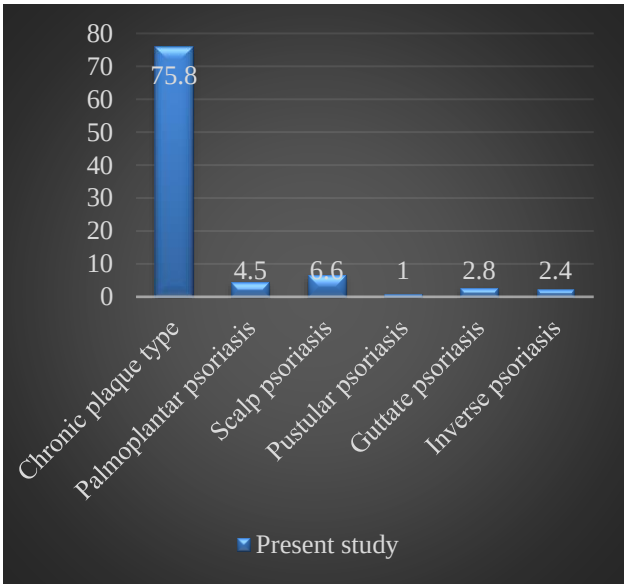

Figure 2: Distribution according to variants of psoriasis.

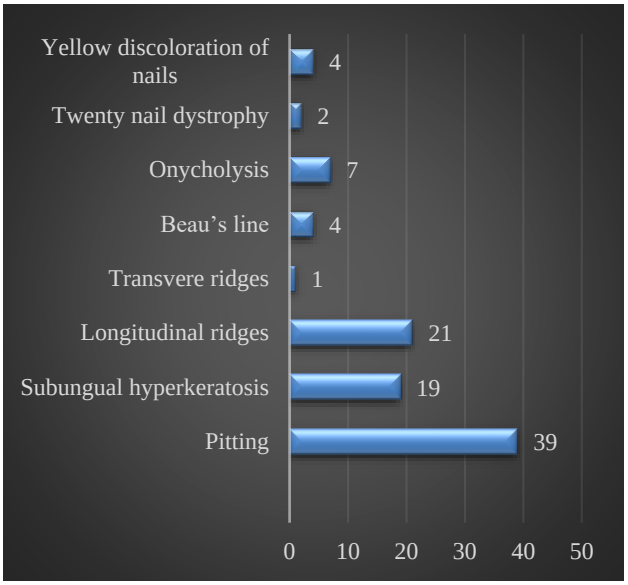

Figure 3: Nail changes in psoriasis.

Out of 132 patients of palmoplantar keratoderma, majority of patients (24.26%) belonged to 41-50 years age group with male: female ratio was 0.69:1. Out of all the types of keratoderma, acquired type (98.5%) was the most common type and 2 patients of congenital type (0.5%), 1 each of punctate keratoderma and mal de malada syndrome (0.75%).

Out of 19 patients of Icthyosis, 89.47% were male and 10.52% were female; Icthyosis vulgaris was the most common clinical type affecting 47.38 % followed by Non bullous Icthosi form erythroderma in 15.78% (Figure 4). Amongst 7 patients of Darier's disease 6 had positive family history.

Table 3: Distribution of patients according to associated miscellaneous systemic disease and co-morbidities (n=289).

Systemic disease	%
Hyperthyroidism	3
Hypothyroidism	3
COPD, bronchial asthma	2.5
Psychiatric comorbidity	12
Alcohol dependence	6
Chronic renal failure, renal stones	2
Down's syndrome	0.5
Psoriatic arthropathy	6.3
HIV	1
Tuberculosis	0
Fatty liver/cirrhosis liver*	1.5
NAFLD**	5
Ca breast	0.5
Hepatitis-B	0.5
Dyslipidemia	26.39
Overweight	10.66
Obesity	36.68
Morbid obesity	26.01
Diabetes mellitus	30.54
Hypertension	31.81

*H/O chronic alcoholism present, **NAFLD: non-alcoholic fatty liver disease.

In 22 patients of phrynoderma, most common age group was 1-10 years of age (54.55%) with male to female ratio was 1.2:1.

Amongst 13 patients of acanthosis nigricans, most common age group was 11-20 years (38.46) with male to female ratio was 1.16. In our study, we had two cases of pachyonychia congenita, both male children with ages 6 years and 9 years respectively.

DISCUSSION

On clinico epidemiological features of keratinization disorders, very few studies were done in India. Vijayan et al reported that majority of the patients of psoriasis (23%) belonged to the age group of 50-60 years, while Dogra et al stated most common age group to be 31-40 years.[4,5] The result of the present study was almost similar to the other studies.[6,7] The sex ratio of Vijayan et al was 1.32 which was similar to our study in which it was 1:1.5.[4]

In present study among psoriasis patients, chronic plaque type (75.8%) was the most common variant (Figure 5) followed by scalp psoriasis (6.6%). Previous studies reported plaque type as the commonest type (90.5%) and scalp psoriasis in 81%.[6,7]

Figure 4 (a and b): Ichthyosis vulgaris and collodion baby.

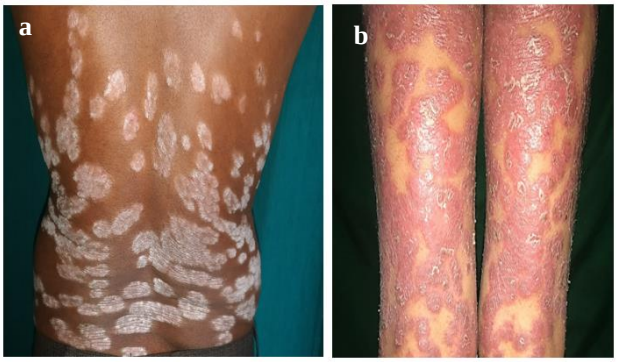

Figure 5 (a and b): Chronic plaque psoriasis.

In our study, HTN (31.8%), diabetes mellitus (30.5%) and obesity (26%) were more common while diabetes mellitus (13%), hypertension (8%) was more common in Alexander et al DM, family history of DM, alcohol addiction and tobacco chewing were more common in Periera et al study.[8,9] Abdominal obesity prevalence varies across different populations. Love et al found that, abdominal obesity was most common component of metabolic syndrome associated with psoriasis followed by hypertriglyceridemia and low levels of HDL cholesterol.[10] While according to German health study, the most common components were HTN and hyperlipidemia.[11]

Several studies have shown that psoriasis is associated with atherogenic dyslipidemia with increased blood levels of total cholesterol, triglycerides, low density lipoprotein (LDL), very low-density lipoprotein, lipoprotein A and low levels of HDL and apolipoprotein B. The association with dyslipidemia was less in our study compared to Pierera et al.[9]

In a study by Gisondi et al, the occurrence of non-alcoholic fatty liver disease was higher in psoriasis patients than in controls.[12] Non-alcoholic fatty liver disease patients in the psoriasis group were more likely to have metabolic syndrome and diabetes than those with psoriasis alone, which was not seen in our study.

The most common nail change seen in psoriasis was pitting (39%), this was in comparison to that seen in Puri et al and Bedi et al study where the most common nail changes seen was pitting 70% and 50% respectively.[13-14] However, the second most common change seen was onycholysis in Bedi et al (22.0%) and Puri et al (52.0%) but in our study it was longitudinal ridges (21%).[14,13]

Out of 132 patients of keratoderma, most common age group was 41-50 years (24.26%) with male: female ratio being 0.69:1 which was less as compared to Murthy et al 1.65:1.[15]Amongst them the most common variant was acquired type of palmoplantar keratoderma (Figure 6).

Figure 6: Palmopalntar keratoderma.

Figure 7: Darrier's disease showing dirty, warty papules involving seborrheic area.

Amongst patients of erythroderma most commonly affected age group was 31-40 years. Such finding was in accordance with mean age group of Akhyani et al and other studies.[16-19] Amongst them most common cause was psoriasis vulgaris (75%) which showed similar result with Akhyani et al.[16]

Amongst 7 patients of Darier's disease, 6 had positive family history. Except the study by Svendsen et al which showed a male to female ratio of 1.6:1, others

epidemiological studies indicated an equal sex distribution of Darier's disease.[20] In present study, male to female ratio was around 1:1 (Figure 7).

Amongst patients of acanthosis nigricans most common age group was 11-20 years of age (38.46%) followed by 41-50 years of age group. Acanthosis nigricans has no known sex predilection. In present study there was no sex predilection.

Figure 8: Pachyonychia congenita showing hypertrophic nail changes and dystrophy of nails.

In our study, we had two cases of pachyonychia congenita, (Figure 8) both male children with 6 years and 9 years of age. Out of them, a 9 years old boy had the features characteristic of pachyonychia congenita type 1 (severe subungual hyperkeratosis, follicular keratotic lesions over extensors and buttocks, focal plantar keratoderma and oral leucokeratosis). Sarojini et al and Sivasundaram et al stated that pachyonychia congenita is an autosomal dominant disorder with high penetrance but of variable expressivity.[21] Jadassohn-Lewandowsky pachyonychia congenita type 1 is the most common type and in our study pachyonychia congenital belong to this type. The other child had only nail changes. There was no family history in any case.

CONCLUSION

Heredity plays an important role in keratinization disorders. Also, various co-morbidities have been associated with different keratinization disorders. Hence, we need to look for these factors while evaluating the patients of keratinization disorders. Paucity in Indian data regarding the subject needs to be addressed by conducting more epidemiological studies with larger sample size, which will help in formulating screening guidelines for Indian population.

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