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Original Article

A clinicoepidemilogical study on icthyosis in a district hospital

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Abstract

Objective To study the demography and clinical spectrum of icthyosis in children below twelve years of age.

Methods A prospective randomized hospital based study was conducted between June 2013 and June 2015 in a district hospital in North India. The children were thoroughly examined and routine investigations were carried out.

Results In two years duration, 2523 children coming to the dermatology OPD were screened for icthyosis.

Conclusion There were 21 patients (70%) of icthyosis vulgaris, 3 patients(10%) of X linked recessive icthyosis, 2 patients (6.66%) of lamellar icthyosis, 2 patients (6.66%) of non bullous icthyosiform erythroderma, 1 patient (3.33%) of bullous icthyosiform erythroderma and 1 patient (3.33%) of Netherton syndrome. Palmoplantar keratoderma was seen in 5 patients(16.66%), impairment of sweating was seen in 6 patients (20%), keratosis pilaris was seen in 3 patients (10%), atopic diathesis was seen in 3 patients (10%), colloidon membrane was seen in 2 patients (6.66%) and eye involvement in the form of corneal opacity was seen in 1 patient (3.33%).

Key words
Icthyosis; Syndrome; Lamellar erythroderma; Palmoplantar; Non bullous; Colloidon.

Introduction

Icthyosis is a disorder of cornification. The word icthyosis is derived from Greek word 'icthys' which means fish. There are various types of icthyosis including icthyosis vulgaris, X linked recessive icthyosis, lamellar icthyosis and various icthyosiform syndromes like bullous icthyosiform syndrome and non bullous icthyosiform syndrome.[1,2] Icthyosis can be hereditary or acquired. Acquired icthyosis is rare and it usually appears in adulthood. The commonest type of icthyosis is icthyosis vulgaris and it is autosomal dominant. Here scaling is mostly present on the extensor surface of extremities and is absent on the flexor surfaces. X linked recessive icthyosis involves mainly flexures.[3] In lamellar icthyosis, the skin is covered with large, thick and dark plate like scales. In congenital icthyosiform erythroderma, the scales are finer and white in colour as compared to lamellar icthyosis, but erythroderma is present.[4] The severest form of icthyosis is harlequin icthyosis in which the neonate has a shell of plate like scales with contraction abnormalities like eyes, ears and mouth.

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Methods

A prospective randomized hospital based study was conducted between June 2013 and June

2015 in a district hospital in North India. The children were thoroughly examined clinically including examination of hair, nails and palms and soles. Routine investigations were carried out. The specialized investigations like skin biopsy were carried out as and where required. Prior permission of hospital ethical committee was taken for the study. A written informed consent was taken from the parents of the children before the study

Results

The results were collected, tabulated and the results were analyzed statistically using chi square test.

Discussion

In two years duration, 2523 children coming to the dermatology OPD were screened for icthyosis and its variants and syndromes and thirty four children were selected. Out of these four children, whose parents refused to be photographed and were not willing to sign the consent form were excluded from our study. Regarding the age distribution **(Table 1)** of patients, 13 patients (43.33%) were below 5 years of age, 12 patients (40%) were between 6-9 years of age and 5 patients (16.66%) were between 10-12 years of age. Our study is in accordance with a study on Ghosh *et al.* in which maximum number of patients presented in the age group of 0-10 years (39; 36.8%).[3] There were 22 males and 8 females and male:female was 2.75:1. Most of the patients in our study were males as most of the patients were from poor socioeconomic strata, where female children are usually ignored and no treatment is sought for them.

Regarding the clinical types **(Table 2)**, out of total 30 children, there were 21 patients (70%) of icthyosis vulgaris **(Figure 1)**, 3 patients (10%) of X linked recessive icthyosis, 2 patients (6.66%) of lamellar icthyosis, 2 patients (6.66%) of non bullous icthyosiform erythroderma **(Figure 2)**, 1 patient (3.33%) of bullous icthyosiform erythroderma **(Figure 3)** and 1 patient (3.33%) of Netherton syndrome.

Table 1 Age distribution of patients

Sr. no.	*Age distribution*	*No of patients(%)*
1	0 – 5 years	13 (43.33%)
2	6 – 9 years	12 (40%)
3	10 – 12 years	5 (16.66%)

Table 2 Clinical variants of icthyosis

Sr. no.	*Clinical type*	*No of patients(%)*
1	Icthyosis vulgaris	21 (70%)
2	X linked recessive icthyosis	3 (10%)
3	lamellar icthyosis	2 (6.66%)
4	Non bullous icthyosiform erythroderma	2 (6.66%)
5	Bullous icthyosiform erythroderma	1 (3.33%)
6	Netherton syndrome	1 (3.33%)

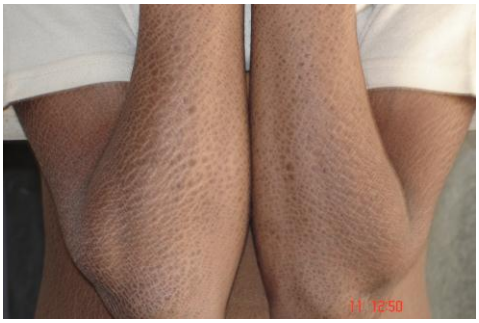

Figure 1 Icthyosis vulgaris in a 11 years old male child.

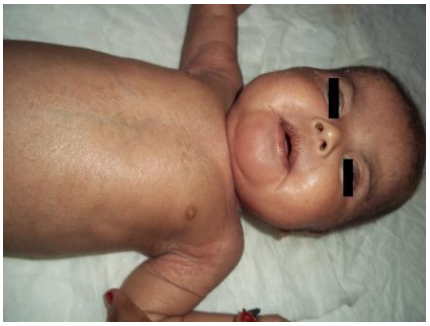

Figure 2 Eight months old male child with Non Bullous icthyosiform erythroderma.

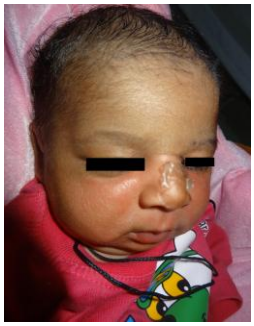

Figure 3 Ten months old male child with Bullous icthyosiform erythroderma.

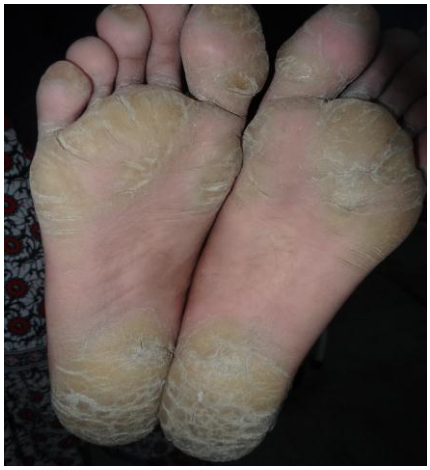

Figure 4 Palmoplantar keratoderma in 12 years old child with icthyosis vulgaris.

Table 3 Clinical spectrum of icthyosis.

Sr. no.	*Clinical spectrum*	*No of patients(%)*
1	Palmoplantar keratoderma	5 (16.66%)
2	Impairment of sweating	6 (20%)
3	Keratosis Pilaris	3 (10%)
4	Atopic diathesis	3 (10%)
5	Colloidon membrane	2 (6.66%)
6	Eye involvement	1 (3.33%)

So, icthyosis vulgaris was the commonest clinical type seen in our study. Family history of icthyosis was positive in 6 patients (20%). Consanguinity was present in 5 patients (16.66%). In a study by Sivaya devi *et al;* out of 64 patients with congenital ichthyosis, ichthyosis vulgaris constituted 72% of cases followed by lamellar ichthyosis 14%. Non-bullous ichthyosiform erythroderma (NBIE) constituted 6% followed by bullous ichthyosiform erythroderma (BIE).[5] So, our results were comparative with other studies.

So, icthyosis vulgaris was the commonest clinical type seen in our study. There is mutations in protein filaggrin in these patients.[6] Icthyosis vulgaris is also known as retention hyperkeratosis. The prognosis of hereditary icthyosis is excellent with many patients experiencing clinical improvement with age.[7-9] In our study, all the cases were of hereditary icthyosis vulgaris. It is typically absent at birth and appears in most of the patients during the first year of life. Acquired icthyosis vulgaris is rare. In most of our patients of icthyosis vulgaris, the symptoms improved during the summer months.

About the clinical spectrum **(Table 3)** of icthyosis, palmoplantar keratoderma **(Figure 4)** was seen in 5 patients (16.66%), impairment of sweating was seen in 6 patients (20%), keratosis pilaris was seen in 3 patients (10%), atopic diathesis was seen in 3 patients (10%), colloid on membrane was seen in 2 patients (6.66%) and eye involvement in the form of corneal opacity was seen in 1 patient (3.33%).

Histopathology of icthyosis vulgaris was done in some children who were willing for it. It showed compact hyperkeratosis in stratum corneum in most of the biopsies.[10] Follicular plugging was seen in those patients in which there was associated follicular plugging. In most of the patients, the granular layer was absent or one layer thick. Dermal infiltration was absent in most of the patients.

Conclusion

This study is important to look into the insights of the children with icthyosis. Since inheritance plays a very important role in these conditions, the parents of the affected children need proper genetic counseling especially in cases of children born out of consanguineous marriage.

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