International Journal of Dermatology, Venereology and Leprosy Sciences

-ISSN: 2664-942X
P-ISSN: 2664-9411
www.dermatologypaper.com/
Derma 2018; 1(1): 23-25
Received: 18-11-2017
Accepted: 22-12-2017

Dr. Amit Mishra
Department of Dermatology, SGT Medical College Hospital & Research Institute, Haryana, India

Corresponding Author:
Dr. Amit Mishra
Department of Dermatology, SGT Medical College Hospital & Research Institute, Haryana, India

Evaluation of cases of Icthyoses: A clinical study

Dr. Amit Mishra

DOI: https://doi.org/10.33545/26649411.2018.v1.i1a.8

Abstract
Background: Icthyoses are heterogeneous group of disorders. The present study was conducted to evaluate cases of Icthyoses reported to the department.
Materials & Methods: The present study was conducted on 38 cases of congenital Icthyoses reported to the department. A careful clinical examination was done. A punch biopsy was taken from atypical and doubtful cases.
Results: Maximum cases were seen in age group 0-10 years (18) followed by 11-20 years (6), 21-30 years (4), 31-40 years (5), 41-50 years (3) and >50 years (2). The difference was significant ($P< 0.05$). Ichthyosis vulgaris was seen in 14, X□linked recessive ichthyosis in 10, Lamellar ichthyosis in 6, Bullous ichthyosiform erythroderma in 4, Epidermolytic ichthyosis in 3 and Ichthyosis hystrix in 1. Hyperlinear palms were present in 3, hyperlinear soles in 34, keratosis pilaris in 22 and atopy in 17. The difference was significant ($P< 0.05$).
Conclusion: Most common lesion reported was Ichthyosis vulgaris and age group 0-10 years had maximum cases.

Keywords: Ichthyosis vulgaris, Skin, X□linked recessive

Introduction
Icthyoses are heterogeneous group of disorders due to defect in keratinization or cornification with abnormal differentiation and desquamation of epidermis which is clinically characterized by dry-rough skin with scaling over much or the entire body surface. The primary function of the stratum corneum is to provide a barrier to water loss without which terrestrial life is not possible. Defective barrier function leads to increased transepidermal water loss, a characteristic feature of ichthyosis. The terminology and nosology of congenital ichthyosis have India continuously evolved and has led to a confusing medley of different terms and classifications. A number of well-defined ichthyoses have characteristic features and can be reliably diagnosed.
According to the 2009 first consensus classification, ichthyosis is divided into nonsyndromic and syndromic. Harlequin ichthyosis, lamellar ichthyosis (LI), and congenital ichthyosiform erythroderma (CIE) fall under autosomal recessive congenital ichthyosis. The keratinopathic ichthyosis due to keratin mutations includes epidermolytic ichthyosis (EI) and superficial EI. Diagnosis is based on dermatologic evaluation, careful family and medical history, and can be strongly supported by directed morphologic examinations and other special analyses. If available, molecular analyses are suggested to confirm diagnosis, and allow for testing of family members and prenatal diagnosis. The need to offer support through patient associations was also stated. With the exception of HI, which is often lethal, patients with ARCI seem to have a normal life expectancy. The present study was conducted to evaluate cases of Icthyoses reported to the department.

Materials & Methods
The present study was conducted in the department of Dermatology. It comprised of 38 cases of congenital Icthyoses reported to the department. The study protocol was approved from institutional ethical committee. All patients were informed regarding the study and written consent was obtained.
Data pertaining to patients such as name, age, gender etc. was recorded. A careful clinical examination was done. Laboratory investigations including complete blood count, liver function test, renal function test, and lipid profile were done. A punch biopsy was taken from atypical and doubtful cases. Results thus obtained were subjected to statistical analysis. P value less than 0.05 was considered significant ($P< 0.05$).

Results

Table I: Age wise distribution of cases

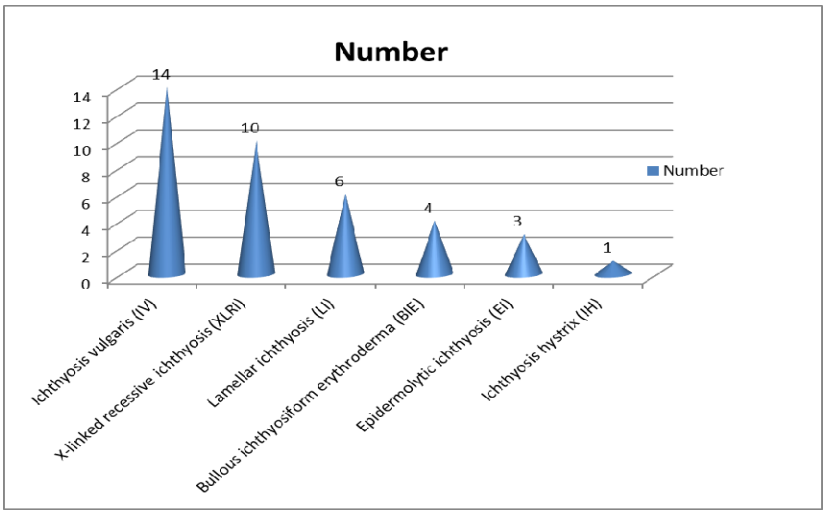
Age group (Years)	Number	P value
0-10	18	0.02
11-20	6	
21-30	4	
31-40	5	
41-50	3	
>50	2	

Table I shows that maximum cases were seen in age group 0-10 years (18) followed by 11-20 years (6), 21-30 years (4), 31-40 years (5), 41-50 years (3) and >50 years (2). The difference was significant ($P< 0.05$).

Table II: Type of lesion

Type	Number	P value
Ichthyosis vulgaris (IV)	14	0.05
X□linked recessive ichthyosis (XLRI)	10	
Lamellar ichthyosis (LI)	6	
Bullous ichthyosiform erythroderma (BIE)	4	
Epidermolytic ichthyosis (EI)	3	
Ichthyosis hystrix (IH)	1	

Table II, graph I shows that Ichthyosis vulgaris was seen in 14, X□linked recessive ichthyosis in 10, Lamellar ichthyosis in 6, Bullous ichthyosiform erythroderma in 4, Epidermolytic ichthyosis in 3 and Ichthyosis hystrix in 1.

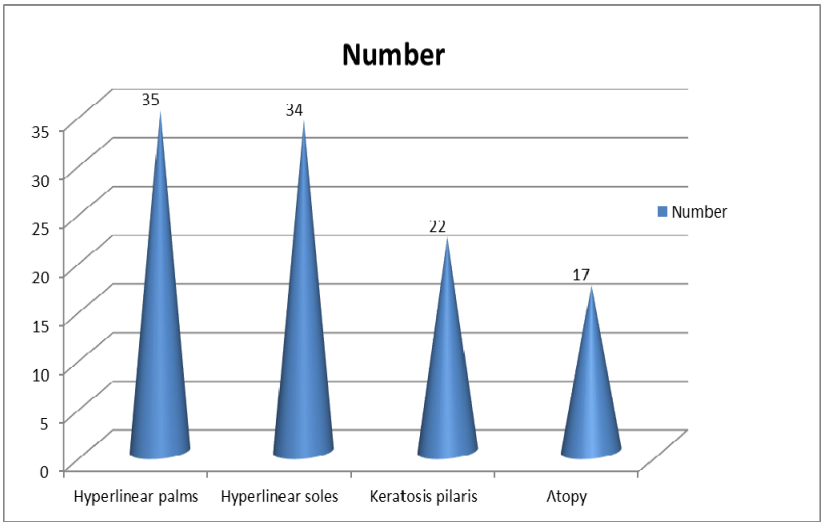


Graph I: Type of lesion

Table III: Clinical features in patients

Clinical features	Number	P value
Hyperlinear palms	35	0.91
Hyperlinear soles	34	
Keratosis pilaris	22	
Atopy	17	

Table III, graph II shows that hyperlinear palms were present in 3, hyperlinear soles in 34, keratosis pilaris in 22 and atopy in 17. The difference was significant ($P< 0.05$).



Graph II: Clinical features in patients

Discussion

In general, determination of whether an ichthyosis is inherited or acquired, presented at birth or later in life, and whether it is limited to the skin or part of multisystem disorder helps in making a diagnosis. Quality and distribution of scales, presence or absence of blistering, erythroderma, and associated abnormalities of skin adnexa are other useful clinical features. A thorough family history is essential for recognizing the inheritance pattern. Establishing a correct clinical diagnosis in a patient with ichthyosis is a prerequisite for making prognostic prediction and therapeutic decision. Recent advances in the molecular genetics have provided tools to categorize ichthyosis on the basis of their underlying genetic defect which helps in offering genetic counseling. The present study was conducted to evaluate cases of Icthyoses reported to the department.

In this study, maximum cases were seen in age group 0-10 years (18) followed by 11-20 years (6), 21-30 years (4), 31-40 years (5), 41-50 years (3) and >50 years (2). Ichthyosis vulgaris was seen in 14, X□linked recessive ichthyosis in 10, Lamellar ichthyosis in 6, Bullous ichthyosiform erythroderma in 4, Epidermolytic ichthyosis in 3 and Ichthyosis hystrix in 1.

Ghosh *et al.* [8] evaluated the epidemiology and clinical characteristics of various types of congenital ichthyoses. Patients were evaluated for epidemiological profile and clinical features. 106 patients of congenital ichthyoses were identified. The most common of the various ichthyoses was ichthyosis vulgaris, followed by lamellar ichthyosis, X□linked recessive ichthyosis. One case of Netherton syndrome and one of ichthyosis hystrix were also identified.

We found that hyperlinear palms were present in 3, hyperlinear soles in 34, keratosis pilaris in 22 and atopy in 17. Wells *et al.* [9] obtained data from 3 incomplete sources and combined them using the capture-recapture method. They identified 144 living patients with ARCI. Of these, 62.5% had classic lamellar ichthyosis and 30.6% had congenital ichthyosiform erythroderma. The age distribution included fewer elderly patients than expected. The prevalence of ARCI in patients younger than 10 years, the best estimate as less subject to bias, was 16.2 cases per million inhabitants. According to the capture-recapture model, 71% of the patients were not being followed up in reference units, 92% did not have a genetic diagnosis, and 78% were not members of the ichthyosis association.

Ichthyosis vulgaris (IV) is relatively mild and easily amenable to emollients. However, the more severe ones like EI (formerly bullous ichthyosiform erythroderma [BIE]) and LI pose difficulties in the treatment of their associated features of fibrous digital bands threatening autoamputation of the fingers, ectropion, and eclabium [10] While the knowledge of these conditions helps treat them by simple means whenever possible and thus avoid unnecessary medications. Furthermore, intensive therapy in the form of high dose oral retinoids can promptly be started to relieve more severe features associated with BIE and LI. Their inheritance pattern is especially important to find the chances of transmission of the disorder to the offspring [11].

Sivaysdevi *et al.* [12] conducted a cross-sectional observational study in which a total of 64 patients were included in this study and relative incidence of different types of ichthyoses was noted. 13 cases of collodion babies were followed and 70% of them developed lamellar ichthyosis. A significant proportion of cases with autosomal recessive inheritance had a history of consanguineous marriage in the parents.

Conclusion

Authors found that most common lesion reported was Ichthyosis vulgaris and age group 0-10 years had maximum cases.

References

1. Sever RJ, Frost P, Weinstein G. Eye changes in ichthyosis. JAMA. 1968; 206:2283-6.
2. Toribio J, Fernández Redondo V, Peteiro C, Zulaica A, Fabeiro JM. Autosomal dominant lamellar ichthyosis. Clin Genet. 1986; 30:122-6.
3. Costagliola C, Fabbrocini G, Illiano GM, Scibelli G, Delfino M. Ocular findings in X□linked ichthyosis: A survey on 38 cases. Ophthalmologica. 1991; 202:152-5.
4. Okano M, Kitano Y, Yoshikawa K, Nakamura T, Matsuzawa Y, Yuasa T *et al.* X□linked ichthyosis and ichthyosis vulgaris: Comparison of their clinical features based on biochemical analysis. Br J Dermatol 1988; 119:777-83.
5. Lykkesfeldt G, Høyer H, Ibsen HH, Brandrup F. Steroid sulphatase deficiency disease. Clin Genet 1985; 28:231-7.
6. Lykkesfeldt G, Høyer H, Lykkesfeldt AE, Skakkebaek NE. Steroid sulphatase deficiency associated with testis cancer. Lancet. 1983; 2:1456.
7. Paige DG, Emilion GG, Bouloux PM, Harper JI. A clinical and genetic study of X□linked recessive ichthyosis and contiguous gene defects. Br J Dermatol. 1994; 131:622-9.
8. Ghosh A, Ahar R, Chatterjee G, Sharma N, Jadhav SA. Clinico-epidemiological study of congenital ichthyosis in a tertiary care center of Eastern India. Indian J Dermatol. 2017; 62:606-11.
9. Wells RS, Kerr CB. Clinical features of autosomal dominant and sex-linked ichthyosis in an English population. Br Med J. 1966; 1:947-50.
10. Traupe H, Fischer J, Oji V. Nonsyndromic types of ichthyoses - An update. J Dtsch Dermatol Ges 2014; 12:109-21.
11. Van Gysel D, Lijnen RL, Moekti SS, de Laat PC, Oranje AP. Collodion baby: A follow-up study of 17 cases. J Eur Acad Dermatol Venereol. 2002; 16:472-5.
12. Sivayadevi P, Karthikeyan R, Anandan H. Congenital Ichthyoses in Pediatric Age Group: A Prospective Study. Int J Sci Stud. 2017; 4(12):143-145.