

Original Article

Erythroderma: a clinicopathological study of 102 cases

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Abstract *Background* Erythroderma may result from different causes and is a rare skin disorder.

Objective Our objective has been to determine the frequency of erythroderma in our environment, its causes and patient evolution.

Patients and methods We reviewed the clinical features and laboratory profile of 102 patients diagnosed with erythroderma who were treated in our department over a 10 years period (1993-2003).

Results The mean age at diagnosis was 48.6 years with male to female ratio of 1.9:1. The most common causative factors were dermatoses (55.9%), followed by drug reactions (29.4%), idiopathic cause (11.8%) and malignancies (2.9%). Apart from erythema that was present in all patients, itching was the most common complaint (64.7%), followed by generalized scaling (56.8%) and chills (30.3%). Most common physical findings were generalized scaling, onychopathy, pitting edema and lymphadenopathy. Skin biopsy was performed in 54 cases with clinical correlation in 36 patients (66.4%).

Conclusions As in other series, preexisting dermatoses were the most common cause of erythroderma. Many of skin biopsies of idiopathic erythrodermic patients showed drug reactions without any history of drug consumption, showing that drugs were suspected as the cause in many patients that were overlooked by them. Drug-related erythroderma had the best prognosis.

Key words

Erythroderma, psoriasis, eczema, drugs.

Introduction

Erythroderma or exfoliative dermatitis, first described by Hebra in 1868, is an inflammatory disorder in which erythema and scaling occur in a generalized distribution involving more than 90% of the body surface.¹ It doesn't have age or sex

limitation and males are affected two to three time more than females.^{1,2} Fever, chills, alopecia and onychopathy are the most common primary complaints.¹

Erythroderma is a serious disease and sometimes is fatal, without treatment. The main cause of death in these patients is metabolic and electrolyte disturbances.²

Several etiologies are known to cause erythroderma such as: skin disorders, drugs and malignancies; however, the cause in

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many cases remains unknown. Severity of the disease, prognosis and treatment are dependent on the etiology, hence determination of incidence of these etiologies has an important impact in a rapid and effective approach to the patients. The incidence of erythroderma is low, hence a prospective study is time consuming. Therefore, by a retrospective study, we reviewed the erythrodermic patients that were admitted in dermatology ward during 10 years period (1993-2003). We searched about the most common presentation of erythroderma and the sensitivity of skin biopsy in the diagnosis of etiologies. Understanding the major etiologies and the most common presentation of this disease in our community can help us with regard to better management and to reduce the complications of this disease.

Patients and methods

We reviewed the records of all patients admitted in the hospital since 1993 to 2003 and a total of 102 cases were finally analyzed. In data analysis we noted these points: sex and age of the patients at the time of admission, positive points in history that helped us in determining the cause of erythroderma such as (positive family history, positive history of previous skin diseases, positive history of drug consumption etc.), the main clinical presentation and physical findings, para-clinical evaluations: WBC count, eosinophilia, ESR, total serum protein, serum albumin level, hemoglobin and hematocrit level, skin and lymph node biopsy. Specimens of skin biopsies of the selected patients were reviewed by a dermatopathologist, in order to determine

Table 1 The most common physical findings in erythrodermic patients (n=102)

<i>Physical findings</i>	<i>n (%)</i>
Generalized scaling	58 (56.8)
Lymphadenopathy	19 (18.6)
Alopecia	10 (9.8)
Pitting edema	28 (27.4)
Onychopathy	37 (36.2)
Palmoplantar hyperkeratosis	9 (8.8)
Hepatomegaly	1 (0.9)
Splenomegaly	1 (0.9)
Fever	5 (4.9)

the importance of skin biopsy in the diagnosis of the etiology and its correlation with clinical findings.

Results

The average age of our patients was 48.6 years, with the youngest patient 3 years old and the oldest 82 years. 67 patients (65.7%) were males and 35 patients (34.3%) were females and male/female ratio was 1.9/1. Itching the most complaint was recorded in 64.7% of cases (66 patients). Other common complaints included generalized scaling (56.8%), chills (30.3%) and fever (5%). Hepatosplenomegaly was seen only in one patient who was a case of Sézary syndrome. Onycholysis and subungual hyperkeratosis were the most common nail changes (**Table 1**).

Laboratory findings associated with respective etiologies are presented in **Table 2**.

Skin biopsy was performed in 54 cases (52.9%) with clinical correlation in 36 patients (66.4%). Skin biopsy was not performed in the rest of cases because the cause of erythroderma was obvious (previous dermatoses or treatment with

Table 2 Laboratory findings according to etiology (n=102)

Laboratory findings	Psoriasis	Idiopathic	Dermatoses	Malignancy	Drug-induced	Total number
ESR>30mm/hr	7	4	5	3	2	21
Elevated WBC >11000	4	6	10	1	9	30
Anemia HCT<35	1	1	3	—	—	5
Eosinophilia>440	4	3	11	2	8	28
Low level of serum protein	—	3	3	—	—	6

Table 3 Histological findings associated with etiology.

Etiology	Psoriasis	Subacute or chronic dermatitis	Contact dermatitis	Seborrheic dermatitis	Drug eruption	Pityriasis lichenoid	Non specific	Total
Contact dermatitis	-	-	2	-	-	-	-	2
Seborrheic dermatitis	-	-	-	-	-	-	-	3
Atopic dermatitis	-	18	-	-	-	-	-	23
Psoriasis	8	-	-	-	-	-	-	10
Drug-induced	-	1	-	-	5	-	-	9
Idiopathic	-	-	-	-	4	-	3	7

some drugs in erythroderma) or the patients refused to do biopsy (**Table 3**). Of six lymph node biopsies performed, 4 cases showed dermatopathic lymphadenopathy and one case each revealed cutaneous lymphoma and malignant T-cell lymphoma, respectively.

The patients were categorized into four groups according to etiology as shown in **Table 4**. The majority of the patients with erythroderma due to carbamazepine consumption were known cases of seizure disorders and the remaining used it for pain relief and neuralgia or were cases of psychiatric problems.

The mean duration of hospital stay in males was 13.2 days and in females was 11.8 days. Longest duration was seen in patients with psoriasis (40 days) and the shortest duration

was recorded in three patients who were cases of allergic contact dermatitis, seborrheic dermatitis and drug eruption with 2 days hospital stay.

One patient died due to sepsis who was a case of chronic renal failure on multiple drug therapy and developed drug-induced erythroderma.

The group associated with best prognosis was that related to drugs.

Discussion

As in other series we found that erythroderma predominantly occurs in adults and has a male predominance. The mean age at onset was slightly higher than in most other series.^{3,4,5,6,7} The major symptoms are non specific and pruritus is common, usually

Table 4 Diagnostic entities causing erythroderma (n=102).

	<i>n</i>	<i>Total</i>
<i>Preexisting dermatoses</i>		57 (55.9%)
Psoriasis	20	
Seborrheic dermatitis	3	
Contact dermatitis	6	
Pityriasis rubra pilaris	1	
Atopic dermatitis	26	
Scabies	1	
<i>Drug reaction</i>		30 (29.4%)
Allopurinol	1	
Carbamazepin	17	
Penicillin	4	
Phenytoin	1	
Phenobarbital	1	
Ceftriaxone	1	
Cefixime	1	
Amitriptyline	2	
Unknown drug	2	
<i>Malignancy</i>		3 (2.9%)
Cutaneous lymphoma	2	
Sézary syndrome	1	
<i>Idiopathic</i>	12	12 (11.8%)

unrelated to any specific diagnostic group as noted in previous series.^{1,5,8} A subjective chilly sensation independent to recorded temperature is common due to excessive vasodilatation and heat loss.⁹ In other series^{4,6,7,10} generalized lymphadenopathy is reported in the majority of patients, but in our study lymphadenopathy was detected in only 19 patients that might have been missed by examiner.

The use of laboratory evaluation to differentiate the various causes of erythroderma has generally not been helpful in all series.^{1,3,5,8,11} In our series, skin biopsy although helpful in only 66.4% of the cases in which it was performed, remains a useful investigation.

In this and previous series^{4,5,6} lymph node biopsy usually demonstrates dermatopathic changes. No liver biopsy was done in our series, but in a previous study⁵ histologic examination of liver detected only non-specific parenchyma changes. Onycholysis and subungual hyperkeratosis were the most common nail changes in our series but in one study¹ shining nails and Beau's lines were recorded as the commonest changes. The main cause of erythroderma in our series was exacerbation of preexisting dermatoses also described by others.^{1,3,5,6,7,8,12}

Atopic dermatitis was the most common underlying cause which may be due to high exposure to sensitizers. It is in accordance with the other studies that were done in different countries,^{5,6,7} but in one study in Pakistan¹ and a study in Iran⁸ psoriasis was recorded as the common preexisting cause. In contrast to our study, drug reactions were rarely the cause of erythroderma in two other series^{1,6} [Table 5].

The most common drug that lead to erythroderma was carbamazepine (17 of 30 cases) as described earlier.⁸ However, in other series^{5,6,7} carbamazepine has been mentioned as a less frequent cause of erythroderma.

In the previous series^{4,6} skin biopsies of idiopathic erythroderma showed nonspecific changes but in our study, 4 of 7 specimens showed drug reactions, without history of any drug consumption in the weeks before the presentation of disease. It shows that drugs should be suspected as the cause in many patients with idiopathic erythroderma as notified by King *et al.*⁵

Table 5 Comparison of the etiologic groups with four previous series. Given figures show the relative incidence (%).

Conditions	Hasan and Janson (1983)	Sehgal et al. (1986)	Botella-Estrada et al. (1994)	Akhyani et al. (2005)	Present study
Preexisting dermatoses	42	52.5	62.5	59.8	55.9
Drug reactions	22	24.7	16	21.6	29.4
Malignancies	4	0	12.5	11.3	2.9
Undetermined	32	22.5	9	7.2	12

In our study the mortality rate as a whole patients was 0.98% which is low in comparison to other studies in which it has been between 11% and 64%. This low mortality rate is partly explained by the short follow up period and by the relatively low prevalence of lymphoma and malignancy in this group.

Overall the prognosis of patients with erythroderma is quite good, although mortality of up to 64% has been reported in one study.¹³

References

1. Pal S, Haroon T.S. Erythroderma: A clinicoetiologic study of 90 cases. *Int J Dermatol* 1998; **37**: 104-7
2. Burton JL, Holden CA. Eczema, lichenification and prurigo. In: *Textbook of Dermatology*, 6th edn. London: Blackwell Science; 1998. p. 629-80.
3. Sigurdsson V, Toonstra J, Vanvolten WA. Idiopathic erythroderma: A follow-up study of 28 patients. *J Dermatol* 1997; **194**: 98-101.
4. Abrahams IMC, Carthy JT, Sanders SL. 101 cases of exfoliative dermatitis. *Arch Dermatol* 1963; **87**: 96-101.
5. King LE, Dufrense RG, Lovett GL et al. Erythroderma: review of 82 cases. *Southern Med J* 1986; **79**: 1210-5.
6. Sigurdsson V, Toonstra J, Hezemans M et al. Erythroderma: a clinical and follow-up study of 102 patients, with special emphasis of survival. *J Am Acad Dermatol* 1996; **35**: 53-7.
7. Botella-Estrada R, Sanmartin O, Oliver V et al. Erythroderma: A clinicopathological study of 56 cases. *Arch Dermatol* 1994; **130**: 1503-7.
8. Akhyani M, Ghodsi ZS, Toosi S et al. Erythroderma a clinical study of 97 cases. *BMC Dermatology* 2005; **5**: 5.
9. Brice KA, Bettley FR. Skin water loss and accidental hypothermia in psoriasis, ichthyosis and erythroderma. *Br Med J* 1967; **4**: 195-8.
10. Odom RB, James WD, Berger TG. *Andrews' Disease of the Skin: Clinical Dermatology*, 9th edn. Philadelphia: WB Saunders; 2000.
11. Nicolis GD, Helwing WB. Exfoliative dermatitis. A clinicopathologic study of 135 cases. *Arch Dermatol* 1973; **108**: 788-97.
12. Sehgal VN, Srivastava G. Exfoliative dermatitis: A prospective study of 80 patients. *Dermatologica* 1986; **173**: 278-84.
13. Hasan T, Jansen CT. Erythroderma: A follow-up of 50 cases. *J Am Acad Dermatol* 1983; **8**: 836-40.
14. Millan EM. Monoclonal antibodies and cutaneous T-cell lymphoma, theoretical and practical correlation. *J Am Acad Dermatol* 1984; **12**: 102-14.

