



## ERYTHRODERMA : A CLINICO – ETIOLOGICAL STUDY IN A TERTIARY HOSPITAL .

### Dermatology

**Pooja Nupur** Senior Resident , Department Of Skin & V.d , Nalanda Medical College & Hospital , Patna.

**Ramawatar Singh\*** Assistant Professor , Department Of Skin & V.d , Nalanda Medical College & Hospital , Patna – \*Corresponding Author.

### ABSTRACT

**Background :** Erythroderma (exfoliative dermatitis) is a cutaneous reaction pattern characterized by generalized erythema and scaling which can be seen in a wide range of cutaneous or systemic diseases. It is important to establish the correct diagnosis whenever possible so that specific therapy may be initiated. The study was undertaken to evaluate the clinical profile and etiology of erythroderma and to correlate clinical diagnosis with histopathological findings.

**Methods :** A retrospective study was conducted in the Department of Skin & V.D , Nalanda Medical College and Hospital , Patna. Fifty (50) cases of erythroderma diagnosed clinically in the past 2 years were studied and their epidemiological, clinical, laboratory & histopathological findings were analyzed.

**Results :** The mean age of presentation was 36 years and male to female (M:F) ratio was 3:2. Apart from erythema and scaling that were present in all patients, pruritis (n=40,80%), peripheral edema (n=10,20%) and fever (n=9,18%) were the most common symptoms at presentation. The most common etiology of exfoliative dermatitis was Drug Reactions (n=20,40%) followed by Atopic Dermatitis (n=10,20%) and Erythrodermic Psoriasis (n=15,30%). Other causes were Mycosis Fungoides, Allergic/Air borne Contact Dermatitis, Lichen Planus, Pityriasis Rubra Pilaris. There was no significant contribution of laboratory data and all the patients were HIV sero negative. The best clinico pathological correlation (100%) was found in Erythrodermic Psoriasis.

**Conclusion :** Erythroderma often obscures the primary lesion. Clinicians should evaluate all cases of erythroderma by eliciting proper clinical history, taking biopsies and performing other ancillary haematological investigations. Hospitalization may be required as the disease carries risk of significant mortality.

### KEYWORDS

Exfoliative Dermatitis , Histopathology , Drug Reaction

### INTRODUCTION

Erythroderma or exfoliative dermatitis is a rare skin disorder characterized by erythema and scaling involving > 90% of body surface area. Erythroderma is not a specific diagnosis but the clinical manifestation of a variety of underlying diseases. The estimated annual incidence is 1 to 2 patients per 100,000 population.<sup>1</sup> It is more common in males and is usually seen in elderly patients however age of onset is primarily related to etiology. Most cases of erythroderma in younger patients are due to atopic dermatitis. Many times erythroderma is the manifestation of an underlying skin disorder like psoriasis, atopic dermatitis, pityriasis rubra pilaris or actinic dermatosis. It may be related to drug intake, idiopathic or due to underlying malignancies. It usually evolves slowly over months to years except for drug reactions which tend to develop acutely. Common clinical symptoms include fever, chills, malaise and pruritis along with peripheral edema, lymphadenopathy and secondary skin infections. It is important to establish correct diagnosis because specific therapy other than corticosteroids or anti-inflammatory may be necessary to improve patient's condition. Erythroderma carries a significant risk of mortality and morbidity and accounts for upto 1% of all dermatological hospital admissions.

There are very few published articles on erythroderma in Indian patients particularly from the Northern India. We aimed to study the clinical profile and etiology of erythroderma and correlate the clinical diagnosis with histopathological findings in this part of the country.

### MATERIAL AND METHODS

This study was carried out retrospectively in the Department of Skin & V.D , Nalanda Medical College and Hospital , Patna. All consecutive cases diagnosed clinically as erythroderma in past 2 years from December 2015 to November 2017 were studied. We analyzed and tabulated their epidemiological, clinical, laboratory and histological data from the hospital records and their relationship to etiology of erythroderma.

### RESULT

The incidence of erythroderma in the Department of Skin & V.D was 0.9%. Fifty (50) cases of erythroderma were studied during the period of 2 years. The male female (M:F) ratio was 3:2. Age of patients ranged from 14 to 78 years. The mean age of presentation was 36 years.

Erythema and scaling were seen in all patients followed by pruritis (n=40, 80%), peripheral edema (n=10, 20%) and fever (n=9, 18%). The onset of these symptoms were gradual taking about 2-3 weeks to develop except in cases of drug reactions where it was acute and patients presented with complaints developing within 24 -48 hours. The most common etiology behind development of erythroderma was drug reaction (n=20, 40%) (Table 1). The offending drugs were not known to the patient in 10% cases where they had taken over the counter drugs for fever or pain. Carbamazepine and Sulfasalazine were other agents causing 5% cases each of drug induced erythroderma (Table 2). This group of erythroderma had a good prognosis with rapid resolution after withdrawal of offending drugs. Atopic dermatitis (n=10, 20%) was seen mostly in people with mean age of presentation being 35 years. Such patients had a personal/ family history of atopy (asthma, allergic rhinitis, atopic dermatitis). Erythrodermic psoriasis (n=15, 30%) was seen in patients with pre existing psoriasis who gave history of irregular use of medications or stoppage of all treatment . Allergic contact dermatitis produced by occupational exposure to chemicals was seen in 1 case. Clinical diagnosis was Air Borne Contact Dermatitis in both these lesions. One case of Lichen planus (LP) and 2 cases of Pityriasis Rubra Pilaris (PRP) were also diagnosed on histopathology. Mild anemia, raised ESR, leukocytosis and hypoalbuminemia were common laboratory findings, however, nothing contributory to finding cause of erythroderma could be established.

Sl. no	Causes	n (%)	Mean age	Pathological features
1	Drug induced	20(40%)	35	Epidermal spongiosis, occasional eosinophils in upper dermis
2	Atopic dermatitis	10(20%)	35	Intercellular edema in epidermis with perivascular infiltrate. Exocytosis and few mast cell.
3	Erythrodermic psoriasis	15(30%)	42	Psoriasiform hyperplasia, parakeratosis and dilated capillaries in dermis
4	Pityriasis rubra pilaris	2(4%)	35	Alternating orthokeratosis and parakeratosis
5	Allergic contact dermatitis	1(2%)	25	Spongiotic epidermis with exocytosis of lymphocytes and few eosinophils

6	Lichen planus	1(2%)	50	Saw-toothed rete ridges with band like inflammatory infiltrate and few civatte bodies.
7	Seborrhoeic dermatosis	1(2%)	30	Spongiosis, irregular acanthosis, inflammatory infiltrate.

**Table 1 : Causes Of Erythroderma**

Serial no.	Drug	No. of cases
1.	Unknown	10 (50%)
2.	Ayurvedic/Homeopathic	3 (15%)
3.	ATT(INH)	3 (15%)
4.	Sodium valproate	2 (10%)
5.	Carbamazepine	1(5%)
6.	Sulfasalazine	1(5%)

**Table 2 : Drugs causing Erythroderma (n=20)****DISCUSSION**

Erythroderma is a rare exfoliative skin disorder, the true incidence of which remains unknown. It is uncommon with an incidence of 1 to 2 per 100,000 population.<sup>1</sup> A prospective study by Sehgal and Srivastava in India determined the incidence to be 35 per 100,000 dermatologic patients,<sup>2</sup> while another study by Hasan and Jansen also estimated the annual incidence to be 1 to 2 per 100,000 population.<sup>3</sup> There is a male predominance in most studies with male to female (M:F) ratio ranging from 2:1 to 4:1 and the mean age between 40 - 60 years.<sup>2</sup> This is similar to our study where the male to female ratio was 1.5:1 with mean age of 36 years. Erythroderma is rarer in children where it accounts for approximately 0.1% of cases attending pediatric dermatology clinics.<sup>4,5</sup> The youngest patient in our study was 14 year male who was diagnosed as a case of erythrodermic psoriasis.

A major challenge lies in determining the etiology of erythroderma. A pre existing dermatosis is the single most common cause of erythroderma as determined by most published series. The most common cause of these dermatosis in Spanish, Middle East and Indian studies include psoriasis and eczema.<sup>6,7,9-12</sup> The triggers for precipitating erythroderma in psoriatic patients are withdrawal of systemic or topical glucocorticoids, use of systemic medications like lithium and antimalarials, phototherapy burns, infection and systemic illnesses.<sup>13</sup>

Drug reactions leading to erythroderma was seen in 11.2% cases in study by Rym et al and 12% cases by Bandyopadhyay et al, however in our study we found drug reactions in 40% cases.<sup>6,14</sup> No particular association was seen with winter season as also found in study by Hulmani et al.<sup>15</sup> The laboratory findings were non contributory similar to the study by Pal and Haroon.<sup>16</sup> In patients with no history of underlying skin disease and who denies any history of drug intake, the diagnosis is more difficult and such cases are almost always biopsied, although in such cases the histopathology is usually non rewarding and shows subacute or chronic dermatitis with psoriasiform reaction. Stage of the disease should be kept in mind while interpreting skin biopsy. In the acute stage spongiosis and parakeratosis are prominent, while in chronic stage, acanthosis and elongated rete ridges are seen.<sup>16</sup> Despite the clinical presentation of erythroderma being relatively uniform, histopathological characteristics of underlying lesion are usually distinctive.<sup>17</sup> The exact underlying pathogenesis of erythroderma is not clear. It is believed that there may be role of cytokines, cell adhesion molecules including interleukins 1 and 2, intercellular adhesion molecules (ICAM-1) and Tumor Necrosis Factor (TNF).<sup>18</sup> Their interactions lead to a high epidermal turn over rate causing increased mitotic activity and increased absolute number of germinative skin cells. This is also associated with loss of epidermal cells together with superficial loss of protein and folate.<sup>19</sup> The principles of management are to maintain skin moisture, avoid scratching, avoid precipitating factors, applying topical steroids and treating the underlying cause and complications. Once the acute irritated stage of the skin is improved, further treatment can be targeted at the underlying etiology.<sup>22</sup> The outcome is unpredictable in idiopathic erythroderma and the course is marked by multiple exacerbations with prolonged requirement of glucocorticoid therapy. In initial documented series, the recorded death rate due to erythroderma varied from 18 to 64%.<sup>2</sup> No mortality was reported in our hospital and this supports Hasan and Jansen's view that erythroderma does not pose a significant risk to the patient's life.<sup>3</sup>

**CONCLUSION**

Erythroderma can be caused by a variety of underlying dermatosis, infections and systemic diseases. Since clinical features are identical irrespective of the underlying cause, and laboratory findings pointing to a specific diagnosis may not be rewarding, the clinician should take a detailed history supported by ancillary investigations. However for definite clinic pathological correlation, multiple skin biopsies may be needed followed by close follow up and repeated evaluation. Despite the homogeneity of clinical expression of erythroderma, diagnostic histopathological features of the underlying disease are retained in most cases. Hospitalization is needed for initial evaluation and treatment. Prognosis of erythroderma is dependent on underlying etiology.

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