

Prevalence, Aetiology and Diagnostic Accuracy for Erythroderma in a Tertiary Centre in Portharcourt, Nigeria

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Abstract Background: Erythroderma is a rare, life threatening dermatological emergency with multiple underlying aetiologies. Methods: We reviewed the clinical, laboratory and histologic findings of 22 patients diagnosed with erythroderma, who were managed in the dermatology unit over six years (from 2016 to 2021). Results: Symptoms were scaly lesions, generalized rash, itching, fever, pustules, and on examination there was lichenification. The Male to Female ratio was 1.75:1 with a mean age at diagnosis of 41.6+17.3 years. The causative factors were commonly idiopathic (59 %) and drug-induced (9%). Others were psoriasis, lichen planus, and onychodermatitis. The comorbidities identified were HIV and Chronic kidney disease. Test of agreement between clinical and pathological diagnosis was 62.5%. Conclusion: Erythroderma is more common in men, aetiology was mostly idiopathic Test of agreement between clinical and pathological diagnosis was 62.5, showing biopsies are an important aid in making a definitive diagnosis.

Keywords: Erythroderma, prevalence, aetiology, diagnostic accuracy

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1. Introduction

Erythroderma is also known as exfoliative dermatitis, it is an inflammatory disorder with a striking clinical presentation characterized by generalized erythema and scaling of >80-90% of the body surface area. [1,2]

Other manifestations include diffuse alopecia, keratoderma, nail dystrophy, ectropion, peripheral oedema and lymphadenopathy. [1,3]

It is a rare, life threatening dermatologic emergency. [2,4,5]

There are multiple aetiologies with common causes such as existing dermatoses such as psoriasis, atopic dermatitis, hypersensitivity drug reactions or cutaneous T-cell lymphoma. [6]

Uncommon causes are pityriasis rubra pilaris, ichthyosis, bullous dermatoses, infestations such as scabies, and connective tissue diseases. [1]

There are also other rare causes which include paraneoplastic, inflammatory and neoplastic causes. The list is extensive and continues to increase despite extensive clinical investigations. The underlying cause is usually not found in at least 25% of patients. [1,5] This is termed idiopathic erythroderma.

A meticulous and detailed clinical assessment and pathologic correlation contribute toward making a diagnosis. [1]

Men are more commonly affected, with a male-to-female ratio ranging from approximately 2:1 to 4:1. [1] There is no racial predilection. The condition can occur at any age but is most commonly seen in patients older than 40years with exceptions of atopic dermatitis, seborrheic dermatitis, staphylococcal scalded skin syndrome and hereditary ichthyosis. [7]

The pathways involved in the de novo genesis of erythroderma are complex, and age-related immune senescence is a contributory factor in elderly patients. [8] There is a complex interaction of cytokines, chemokines, and intercellular adhesion molecules.

Significant systemic complications include fluid and electrolyte abnormalities, hypoalbuminemia, thermoregulatory disturbance, cardiac failure, capillary leak syndrome, infection, and death. [8]

There are few studies in the literature and they are mostly retrospective. This study is aimed at looking into the demographics, aetiology and diagnostic accuracy of patients with erythroderma attending the dermatology clinic of the University of Port Harcourt teaching hospital, Port Harcourt Nigeria.

2. Materials and Methods

2.1. Study Area

This study was carried out at the Dermatology Clinic of the University of Port Harcourt teaching hospital. This is a tertiary centre located in an urban town of Port Harcourt and serves the people of Rivers State and referrals from primary and secondary health centres and private hospitals.

2.2. Study Population

The study population included individuals aged 18years and above presenting to UPTH dermatology clinic with erythroderma.

2.3. Sample and Sampling

This was a retrospective, cross-sectional study to review cases of Erythroderma seen at the Dermatology Unit of the University of Port Harcourt Teaching Hospital (UPTH) between 2016 and 2021.

A proforma data collection sheet was used to collate the history of duration and type of eruptions, onset of eruptions, location of initial lesion, itching, pain, fever, presence of scaly lesions, ulcerations, pustular lesions, topical medication use, comorbidities and examination of the integumentary system.

Results of investigations such as retroviral screening, hepatitis B and C, full blood count, erythrocyte sedimentation rate, serum electrolytes, urea and creatinine, urinalysis, liver function tests, Glucose 6 phosphate dehydrogenase deficiency (G6PD assay), fasting blood glucose, fasting lipid profile and skin biopsy was collated.

The study was approved by the ethical committee of the hospital.

Cochrane’s formula was used in the sample size calculation. Using an alpha level of 0.05, an erythroderma

rate of 4.22% from an African study was obtained. [9], with error limit of 0.1, non-response of 10%, a sample size of 22 was deemed appropriate for the study.

The Statistical Package of Social Sciences version 21 was employed in the statistical analysis. Frequencies and proportions were used to summarize the qualitative variables while means and standard deviations were used for quantitative variables in the study. Chi-square statistics were used to compare differences in proportions using the 0.05 significant level.

3. Results

A total of 2,628 new dermatology patients were seen in the dermatology outpatient clinic during the study period, a diagnosis of erythroderma was made in 22 patients, with 14males and 8 females giving a male to female ratio of 1.75:1. The mean age of patients in this study was 41.6 + 17.3years (age range 4-83years).

Table 1 shows the socio-demographic characteristics of the study population with half the population (50%, 11) in the 40-59 age range.

Table 1. Socio-demographic characteristics of the study population

Variables (n=22)	Frequency n	Percentage %
Age category (in years)		
0 – 19	3	13.6
20 – 39	5	22.7
40 – 59	11	50.0
60 – 79	2	9.1
80 – 99	1	4.5
Sex		
Male	14	63.6
Female	8	36.4

The aetiologies identified were Idiopathic, Lichen planus, Psoriasis, Atopic dermatitis, Onchodermatitis, and Drug-induced. The most common cause was idiopathic.

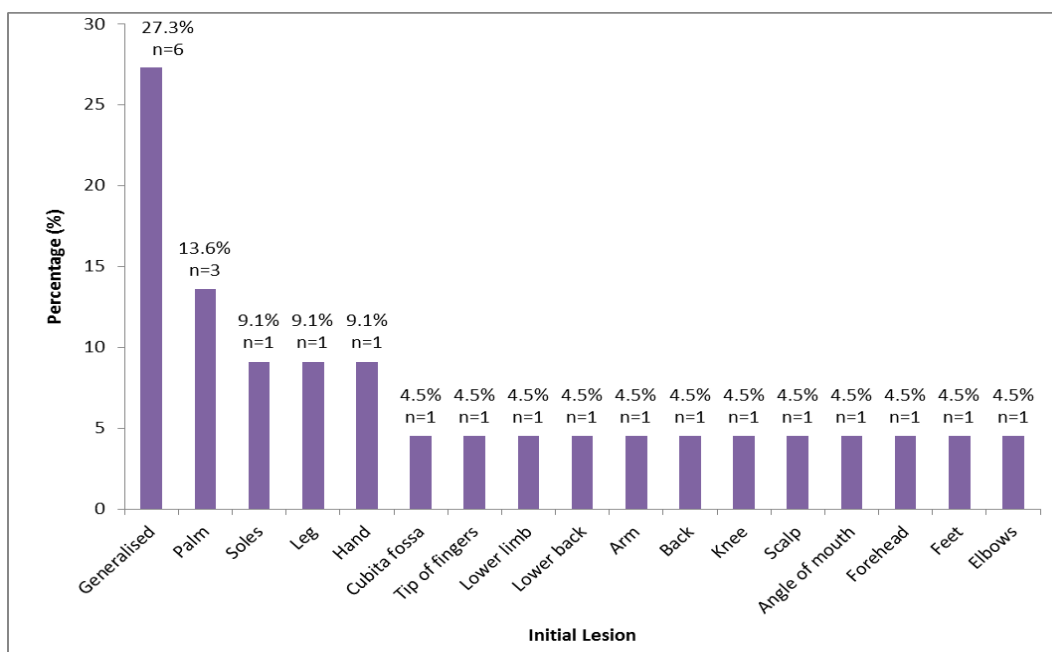


Figure 1. Shows location of initial rash

Figure 1 shows the location of the initial rash which was generalized (27.3%), palm (13.6%), soles (9.1%), leg (9.1%), hand (9.1%), back (4.5%), knee (4.5%), scalp (4.5%), forehead (4.5%), feet (4.5%) and elbows (4.5%) with the generalized presentation being the most common.

Table 2. Duration of dermatological complaints

Duration of Presenting complaints	Frequency n	Percentage %
<1month	1	5
1month – 1year	11	50.0
>1year	10	45
Total	22	100.0

Half (50%) of the population had the duration of complaints from 1month to 1year. However, 95.5% had complaints ranging from 1month to > 1year.

Only one patient was HIV positive using serum antibody tests. All 12 tested were negative for both

Hepatitis B Antigen and Hepatitis C Antibody.

Drug causes (herbal medications) were identified in two patients. The herbal medications were alcohol-based, other ingredients could not be identified.

Sixteen out of twenty-two patients reported itching, fourteen reported scaly lesions, and four had a fever.

The comorbidity identified was Chronic kidney disease on maintenance haemodialysis. There were no patients with hypertension, diabetes mellitus or asthma.

None of the patients had hepatomegaly, peripheral lymph node enlargement or anaemia. However, the erythrocyte sedimentation rate was elevated in two patients, with values of 40 and 62mm/hr.

Liver function tests were within normal limits in four patients who carried out the investigation.

Skin biopsies were done in 10 of the 22 patients and the diagnosis of erythroderma was confirmed in four patients. Skin biopsy was not done in some of the patients due to financial constraints.

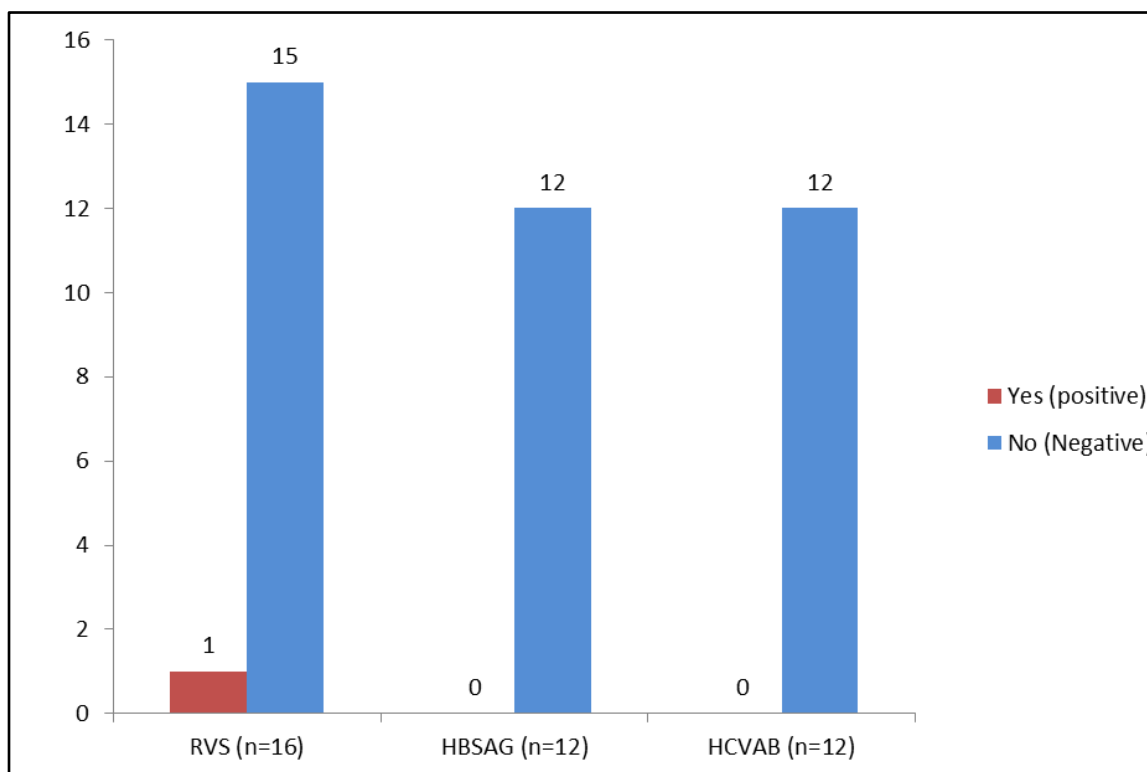


Figure 2. Shows results of serology tests.

3.1. Analysis Outputs

A total of 16 patients with skin lesions who underwent biopsy were involved in the study. Mean age (SD) was 45.6 (15.5) years. Male to Female ratio was 1.3:1.

3.2. Validity Tests for Clinical Diagnosis in Relation to Histological Diagnosis

Table 3. Diagnostic Accuracy of Clinical Diagnosis in Relation to Histological Findings among Patients with Erythroderma

		Histology diagnosis (Gold Standard)		Total
		Erythroderma	No erythroderma	
Clinical diagnosis	Erythroderma	2 <i>True positive</i>	4 <i>False positive</i>	6
	No Erythroderma (<20.00)	2 <i>False negative</i>	8 <i>True negative</i>	10
	Total	4	12	16

Kappa=0.143; 95% confidence interval: -0.338-0.624

$$\text{Sensitivity} = \frac{\text{True Positive}}{\text{True Positive} + \text{False Negative}} \times 100 = \frac{2}{2+2} \times 100 = 50.0\%$$

$$\text{Specificity} = \frac{\text{True Negative}}{\text{True Negative} + \text{False Positive}} \times 100 = \frac{8}{8+4} \times 100 = 66.7\%$$

$$\text{Positive Predictive Value (PPV)} = \frac{\text{True Positive}}{\text{True Positive} + \text{False Positive}} \times 100 = \frac{2}{2+4} \times 100 = 33.3\%$$

$$\text{Negative Predictive Value (NPV)} = \frac{\text{True Negative}}{\text{True Negative} + \text{False Negative}} \times 100 = \frac{8}{8+2} \times 100 = 80.0\%$$

$$\text{Overall Accuracy} = \frac{\text{True Positive} + \text{True Negative}}{\text{All}} \times 100 = \frac{8+2}{16} \times 100 = 62.5\%$$

4. Discussion

In approaching a patient with erythroderma, who usually presents with non-specific clinical features, the history or clinical evidence of characteristic pre-existing inflammatory skin lesions, exposure to medications and chemicals should be sought, in addition to clinical manifestations of a primary dermatosis.

Offending agents should be identified and discontinued, risk factors for HIV should be checked, as well as evaluation for rare causes. Confirmation via histologic examination (multiple skin biopsies; lymph node biopsy if lymphadenopathy exists), laboratory evaluation and imaging should be done if necessary.

The prevalence rate of 0.84 % contrasts with 2.7% which was obtained by Salami et al in Irrua, Edo state, Nigeria [8] where more HIV-positive cases (70%) and antiretroviral medications accounted for 5% of cases of Erythroderma. A study in Senegal gave a prevalence of 0.2% of all medical visits [9]. 13% was observed by Munyao et al [10] in the Kenyatta national hospital, Kenya. This was a 10year study so this could account for the increased prevalence.

The male to female ratio in this study was 1.75:1 which is similar to ratios obtained from previous studies [3,8,11] which is a pointer to male preponderance. The reason remains unclear.

This study had a lot of idiopathic cases, similar to a study in Sao Paulo where there were 16.8% of such cases, though diagnosis for a few patients were later found during follow-up visits [12]. The cases in this study may be as a result of many late presentations and prior modification of rash with various topical medications including corticosteroids and herbal use. [13].

The disease is life-threatening [14] and necessitates hospital admission in some patients with impairment of quality of life.

The location of the initial lesion in the patient who was HIV positive was on the forehead in contrast with a previous study by Salami et al which showed presentations of HIV patients as adverse cutaneous drug reactions. [15]

There was no identifiable underlying malignancy in this patient series however occult malignancies have been identified in older patients. [8]

The test of agreement between clinical diagnosis of erythroderma and histopathological diagnosis was 62.5%,

which is higher than 31% gotten in another study on erythroderma. [16]

Though 62.5% is good, it shows that it very important to do a skin biopsy in all erythroderma patients to aid in the establishment of a definitive diagnosis.

There was no recorded death in this patient series, probably due to early age of presentation, and management at a teaching hospital where specialist care by dermatologists contributed to the outcome.

5. Conclusion

Erythroderma is a challenging diagnosis with aetiology sometimes difficult to establish, so a biopsy is mandatory. There was a male preponderance in this study. Appropriate and timely management is very relevant to preventing increased morbidity and mortality.

Study Limitations

Complete investigations for most of the patients were not done due to financial constraints.

Small sample size as some patients were lost to follow up, probably due to out of pocket payment for healthcare for most individuals.

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