REVIEW

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Clinical Approach to Erythroderma

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ABSTRACT

Erythroderma is a state in which greater than 90% of the total body surface area is erythematous and desquamated. It is a dermatological emergency which can be seen in every age group. The disease first presents with erythematous plaques that have a tendency to merge. Pruritus is frequently present. The rash spreads to the entire body within two to six days and widespread desquamation ensues. Etiological factors of erythroderma can be grouped into four categories which are pre-existing dermatological diseases, drug reactions, malignancy related and idiopathic. The most important part of erythroderma management is the supportive measures. Fluid and electrolyte balance should be maintained. Local wound care should be done with antiseptic wet dressings and emollients. Mid-potency topical corticosteroid creams should be used. The disease prognosis is dependent upon the etiology.

Keywords: Diagnosis, Erythroderma, Etiology, Treatment

Introduction

Erythroderma is a state in which greater than 90% of the total body surface area is erythematous and desquamated. It is a dermatological emergency which can be seen in every age group. The most common ages to present with erythroderma are 45 years and older. Erythroderma can be observed in children due to atopic dermatitis and hereditary diseases as well. It is two to four times more common in males compared to females. The incidence can vary according to the geographic region [1,2].

Clinical Presentation

The disease first presents with erythematous plaques that have a tendency to merge. Pruritus is frequently present. The rash spreads to the entire body within two to six days and widespread desquamation ensues. The skin is bright red, endurated, desquamating and warm. Wide desquamated plaques are seen in the chronic form of the disease whereas small plaques are present in acute forms. Heat and fluid loss due to erythroderma may lead to hypothermia and heart failure in severe cases. Chronic cases may lead to alopecia, longitudinal ridging of the nail plate and onychochysis. Lymphadenopathies, hepatomegaly and splenomegaly may also be observed [2,3].

Etiology

Etiological factors of erythroderma can be grouped into four categories which are pre-existing dermatological diseases, drug reactions, malignancy related and idiopathic [2]. A study from Singapore, regarding the etiological factors of erythroderma has concluded the following in 225 patients. Pre-existing dermatological diseases are the most common cause of erythroderma (68.9%) with eczema and psoriasis being the leading dermatoses. Idiopathic erythroderma is the second most common category (14.2%). Drug related erythroderma is the third most common group (10.7%). Malignancies are the least common cause of erythroderma (4%) with 2.2% consisting of cutaneous malignancies [4]. A study from Turkey revealed that the most common etiological factor for erythroderma

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was psoriasis vulgaris (59.6%) followed by drug eruption (17%), mycosisi fungoides (12.8%), atopic dermatitis (4.3%), bullous pemphigoid (2.1%), pytriasis rubra pilaris (2.1%) and polymorphic light eruption (2.1%) [5].

The common dermatoses that may present with or lead to erythroderma are psoriasis, airborne contact dermatitis, seborrheic dermatitis, atopic dermatitis, staphylococcal scalded skin syndrome, phytophotodermatitis, photosensitive dermatitis, pityriasis rubra pilaris, Pemphigus foliaceus, stasis dermatitis and ichthyosiform erythroderma. Less common pre-existing dermatological causes of erythroderma are candidiasis, dermatophytosis, mastocytosis, lichen planus, Reiter's syndrome, toxic epidermal necrolysis, diffuse/erythrodermic mastocytosis, sarcoidosis, pemphigoid, lupus erythematosus and crusted (Norwegian) scabies [2].

Common drugs to cause erythroderma are acetaminophen, minocycline, actinomycin-D, nitrofurantoin, allopurinol, omeprazole, arsenic, para-amino salicylic acid, barbiturates, penicillin, captopril, phenothiazine, chloroquine diphosphate, phenytoin, chlorpromazine, quinidine, cemetidine, rifampicin, dapsone, streptomycin, gold, sulfadiazine, hydantoin sodium, sulfonyl urea, interferon, tetracycline, isoniazid/isonicotinic hydrazide, thalidomide, isotretinoin, tolbutamide, lithium, vancomycin and mercurials [2].

The etiological factors can be grouped according to age as well. The causes of erythroderma in neonates and infants are grouped as congenital and non-congenital. The congenital causes are nonsyndromic congenital ichthyosis, syndromic congenital ichthyosis, Omenn syndrome, graft-versus-host disease, congenital cutaneous candidosis, psoriasis, diffuse cutaneous mastocytosis and staphylococcal scalded skin syndrome. The non-congenital causes are psoriasis, eczemas (atopic dermatitis, seborrheic dermatitis, staphylococcal scalded skin syndrome), drugs (vancomycin and ceftriaxone), metabolic disorders (holocarboxylase synthetase and biotinidase deficiency, essential fatty acid deficiency). The causes of erythroderma in the school ages are infections (staphylococcal scalded skin syndrome, crusted scabies), drugs (antiepileptics, amoxicillin, sulfonamides, antitubercular drugs), atopic dermatitis and psoriasis. The causes of erythroderma in adults are preexisting dermatoses (psoriasis, contact dermatitis, airborne contact dermatitis, chronic actinic dermatitis atopic dermatitis), drugs (antiepileptics, antimicrobials, analgesics), cutaneous T-cell lymphomas (Sézary syndrome, mycosis fungoides), internal malignancies, Multisystem disorders (dermatomyositis, subacute cutaneous lupus erythematosus) and idiopathic [1].

Diagnosis

Following the diagnosis of erythroderma, each patient should be assesed with the following [3,6]:

- Weight
- Body temperature
- Heart rate
- Respiratory rate
- Complete blood count
- Sedimentation rate/C-reactive protein
- Liver and kidney function tests
- Urinalysis
- Chest X-ray
- Electrocardiography
- Biopsy
- Viral serology

More specific test are total immunoglobulin E level if atopic dermatitis is considered; patch test if allergy is considered; skin scrapings if scabies or dermatophyte infections are considered; lymph node biopsy, peripheric staining and bone marrow aspiration if lymphoma is considered; and malignancy screening for elderly patients [3,6].

Management

The most important part of erythroderma management is the supportive measures. Fluid and electrolyte balance should be maintained. Hydration is of utmost importance. The patient should be kept at a warm and humid environment in order to prevent hypothermia. Protein rich nutritients are recommended. Local wound care should be done with antiseptic wet dressings and emollients. Mid-potencty topical corticosteroid creams should be used in treatment instead of high-potency creams because systemic side effects should be observed due to increased surface area of vulnerable skin. Seditising antihistaminic drugs are recommended for pruritic symptoms. Systemic antibiotherapy should be initiated if infection is suspected. Leg elevation and anti-diureticdrugs are recommended for peripheral edema [3,7].

The underlying diseases causing erythroderma should also be treated. Cyclosporine, retinoids, metothrexate, infliximab or phototherapy are recommended for the underlying psoriasis; systemic steroid and antimicrobials for atopic dermatitis; systemic steroid, retinoid and metothrexate for pytriasis rubra pilaris; intravenous immunoglobulin for toxic epidermal necrolysis; extracorporeal photophoresis, phototherapy, alkalizing chemotherapy or radiotherapy for lymphomas; and ivermectin and topical antiscabicidal drugs for scabies. Systemic steroid treatment can be initiated in idiopathic cases after the exclusion of psoriasis or staphylococcal scalded skin syndrome. The culprit drugs should be stopped in drug-induced cases as well [3,7].

In a study comparing the therapeutic options for idiopathic erythroderma, the authors concluded that the most effective treatment modality was cyclosporin (50-100 mg/day) [6].

Prognosis

The disease prognosis is dependant upon the etiology. Rapid treatment response is observed in drug-induced cases, lymphoma, leukemia, contact allergy and staphylococcal scalded skin syndrome. The response is slower if there is an underlying dermatosis such as psoriasis or atopic dermatitis. The disease may be mortal in elderly patients due to infectioni dehydration, electrolyte imbalances, heat intolerances and high-output heart failure. Postlesional hyperpigmentation or hypopigmentation may be observed in dark skinned individuals. Nail dystrophies, nevi evolvement, keloids, alopecia, generalized vitiligo and pyogenic granuloma have been reported in patients surviving erythroderma [3,7].

Ethics

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: D.Ö., T.K.U., Concept: D.Ö., T.K.U., Design: D.Ö., T.K.U., Literature Search: D.Ö., T.K.U., Writing: D.Ö., T.K.U.

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