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Case Report

Erythroderma patient with chronic kidney disease treated with systemic corticosteroids

Ita Anggraini[1], Yuwandita Tamara Putri Dermawan[2]

[1] Medical Faculty, Padjadjaran University, West Java, Indonesia.
[2] Medical Faculty, Lampung University, Lampung, Indonesia.

Abstract Erythroderma is a rare but emergency case because it is a potentially life-threatening condition. It is described as generalized erythema and skin scaling, involving greater than 90% of body surface area. It can be caused by several etiologies, which the most common cause is psoriasis. The pathogenesis depends on the underlying cause. Establishing the definitive diagnosis is still challenging since the histopathology examination is gold standard for erythroderma. But not all healthcare facilities have this examination. Some of them use clinical approach to diagnose. Moreover, erythroderma patients should have comprehensive management, including nutritional assessment, correction of fluid and electrolytes imbalances, treatment to secondary infections, topical therapy, and systemic agents. Targeted therapy can be used after the underlying cause is established, but it is still considered because of cost-effectiveness and the availability. As an alternative, systemic corticosteroids could be given as one of the treatments, although it is still debated for the side effects. We described a case of erythroderma in a 56-year-old male patient with chronic kidney disease. The patient was diagnosed with his medical history, physical examination and laboratory examination. The management of this patient is comprehensive including topical and systemic agents. Patient's clinical improvement after given topical and systemic corticosteroids.

Key words
Corticosteroids; Erythema; Erythroderma.

Introduction

Erythroderma is generalized erythema and scaling of the skin which involves greater than 90% of body surface area. It is a rare condition with incidence rate of approximately 1 per 100,000 adults and more common in male population. It can be caused by several etiologies. Pathogenesis depends on the underlying cause. It is believed that this disease related to the interaction between cytokines and cellular adhesion molecules, such as IL-1, IL-2, IL-8, intercellular adhesion molecule 1 (ICAM-1), TNF-α and Interferon-γ which involved in the pathogenesis. Histopathology examination needed to establish a definitive diagnosis. But in some healthcare facilities, this examination is still limited.[1-5]

Erythroderma patient require immediate attention and comprehensive management because it is potentially life-threatening condition. Targeted therapy should be administered once the underlying cause is established. The use of systemic corticosteroids is still debated because of the side effects.[1,5] We reported a case of an adult patient who was diagnosed with erythroderma and chronic kidney disease that was given systemic corticosteroids.

Case Report

A 56-year-old man came to the emergency room (ER) with erythematous plaques that had

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Address for correspondence
dr. Ita Anggraini
St. Raya Bandung Sumedang Km.21, Hegarmanah, West Java, 45363, Indonesia.
Ph: +6285781373175
Email: itanggraini05@gmail.com

appeared gradually since two weeks prior to admission. Initially, the lesions are small red spots in the skin of his palms and legs then progressed to the body and scalp overall and developed with skin dryness and exfoliation covered in thin scales. Patient was presented with itching and pain with Visual Analogue Scale (VAS) 6-7. The patient also had intermittent fever, nausea, vomiting, and appetite had decreased since two weeks prior to admission. Swelling and pain in his legs were also noted. Patient also had similar complaint two years ago and got systemic corticosteroids. About five weeks ago, he been admitted to the hospital due to diarrhea, urinary tract infection, chronic kidney disease, anemia, and also with redness throughout his body. There was improvement of his skin condition after eight days in hospital, but after one week of being discharged from hospital, he had small red spots again in his palms and legs.

He denied any history of diabetes mellitus, hypertension, atopic disease, drug allergy, or cancer.

On physical examination, he had a blood pressure of 104/59 mmHg, pulse rate of 105 beats per minute, strong, palpable, and regular, a respiratory rate of 20 cycles per minute, temperature of 37.5°C, and oxygen saturation level of 98% in room air. He had obesity based on anthropometric status (BMI=28.65 kg/m^2). The examination of lymph nodes, lungs, heart, abdomen were all within normal limits. His dermatological status was at regio of head, back, abdomen, superior and inferior extremities appeared erythematous patches multiple, discrete, plaque in sizes, irregular forms, defined borders (almost 90% of body surface area), accompanied with well-defined thin-coated white scales. He also had onycholysis in his fingernails and toenails (**Figure 1**).

The laboratory tests showed decreased of hemoglobin (10.7 g/dL), increased in leukocytes (23,890 x10^3/ul) with differential count 0/3/0/86/5/6, decreased of albumin level (3.1 g/dL), kidney dysfunction (urea of 63 mg/dL; creatinine of 6.7 mg/dL), high potassium of 5.3mEq/L, normal liver function test, normal blood sugar. Chest X-ray result was suspect to pneumonia. USG abdomen test showed chronic

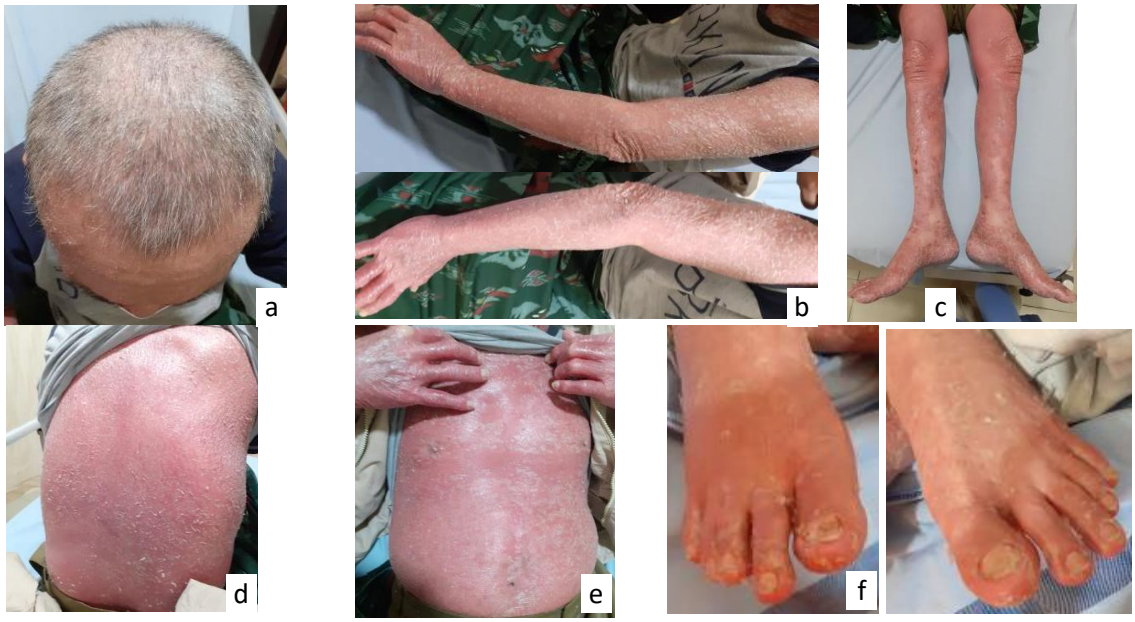

Figure 1 Distribution of lesions. (a) head, (b) superior extremity, (c) inferior extremity, (d) back, (e) abdomen, (f) onycholysis in toenails.

kidney disease bilateral and multiple cyst renal bilateral. Histopathological examination was not performed in this patient, because of limited facilities. Patient also consulted with the internal medicine division for his systemic disease. Patient received systemic corticosteroids as methylprednisolone 2x62.5 mg IV for two days, then tapering off to 2x31.25 mg for two days, and 1x48 mg for fifth day. Additional systemic treatments were antibiotic ampicillin sulbactam 3x1.5 grams IV, normal saline 0.9% 500 mL every 12 hours, omeprazole 2x40 mg IV, cetirizine 1x10 mg orally, polystyrene sulfonate sachet 3x1 orally, albumin capsule 3x1 orally. He also received topical treatment, such as desoxymethasone cream 0.25% and vaseline album. The use of topical and systemic steroids achieved improvement of the skin lesions of this patient.

Patient suggested by the internal medicine division to had hemodialysis due to an increase of urea and creatinine (90 mg/dl; 7.0 mg/dL) and decreased of urine output in a day. But, the patient refused to has hemodialysis.

After five days in the hospital, the patient was discharged. He got home medication including folic acid 2x1 tablet orally, vitamin B12 3x50 mg orally, $CaCo_3$ 3x500 mg orally, sodium bicarbonate 3x1 tablet orally, albumin capsule 3x1 orally, methylprednisolone 2x16 mg orally for seven days, cetirizine 1x10 mg orally, ketoconazole shampoo three times in a week, vaseline album 2x1, bathe with baby soap. He was educated to follow up his conditions at internal medicine polyclinic and dermato-venereology polyclinic in one week after he was discharged from hospital.

Discussion

Erythroderma is a rare condition which has an incidence rate of approximately 1 per 100,000 adults and occurs at an average of 42-61 years. It is more common in males with male to female ratio of 2-4:1. It is similar to previous study in Portugal (2016) and Pakistan (2016) that erythroderma was also age-related with mean age of 54.4 and 48.6, respectively, and predominated by males.[1-4] Manifestations of the disease are erythema, desquamation, pretibial edema, and systemic symptoms like dehydration, fever, malnutrition, fatigue, pruritus, and malaise. It ranges from mild to severe and potentially life-threatening with mortality rate of 4-64%.[1,4,6]

It is still unclear about the mechanism of the disease. Implicated pathogenesis depends on the underlying cause. It is believed that this disease related to the interaction between cytokines and cellular adhesion molecules, such as IL-1, IL-2, IL-8, intercellular adhesion molecule 1 (ICAM-1), TNF-α and Interferon-γ which involved in the pathogenesis. Interactions between these cytokines result in increased epidermal cellular division which increase mitotic rates and shortens transit time of cells though the epidermis, so cause the exfoliation of the cutaneous. The inflammatory cells also play a role in this pathogenesis. The activation of macrophages, keratinocytes, Langerhans cells, Th2 cells and Th17 cells trigger inflammatory reaction, blood vessels dilatation, and widespread desquamation.[3,5] This patient had fever due to loss of temperature regulation, pretibial edema due to significant protein loss which exceeding 9 g/m^2 body surface area per a day, that lead to hypoalbuminemia. Based on the laboratory results, this patient showed anemia, leukocytosis, hypoalbuminemia, electrolyte imbalance (hyperkalemia), with renal dysfunction. Anemia can be caused by chronic disease, malabsorption of the gut, or increased loss of blood from the skin. Leukocytosis can indicate infection or abnormal cells in some hematologic disorders. Erythroderma can lead to

metabolic and physiological complications, such as electrolyte imbalances, high-output cardiac failure, acute respiratory syndromes, and secondary infections. The presence of hepatosplenomegaly (1/3 of cases) and lymphadenopathy (rarely), particularly associated with liver dysfunction. While chronic kidney disease in erythroderma can be caused by immune-mediated renal damage, drug-related renal damage and chronic renal damage.[1,4,7,8]

Erythroderma can be caused by various etiologies. The most common cause is exacerbation of underlying disease, i.e. psoriasis (23%), followed by atopic dermatitis and contact dermatitis (10%). The other causes are drug reactions, systemic diseases (leukemia, Human Immunodeficiency Virus/HIV infection, T-cell lymphoma), and idiopathic. It is similar to a previous study in Portugal, among 103 erythroderma patients induced by the pre-existing dermatoses (65%) and mainly caused by psoriasis (44.7%). Similar results also found in Pakistan (2016), the most common cause of erythroderma is pre-existing dermatoses (65.2%), followed by drug reactions (23.7%), malignancies (11.1%). Nevertheless, research in Albania (2019), showed the most common etiology is still underlying skin disease (78.4%) but followed by idiopathic (16.4%).[1,2,4,8]

As for this case, the 56-year-old man had chief complaint of erythematous plaque for two weeks before admission. This gradual onset can be implied that the cause of erythroderma is systemic or primary cutaneous. From the medical history, patient had repeated similar complaint of skin problems. But, we still differential diagnosed with erythroderma caused by psoriasis and accompanied with systemic disease, i.e. chronic kidney disease. Because based on literature, the most common cause of erythroderma is psoriasis.[1,2] So, the biopsy examination still needed to diagnose the underlying cause of the disease. However, histopathological examination was not performed in this patient because of limited facilities. In previously published series, 9-47% of cases did not have identified cause due to the difficulty in diagnosing the underlying conditions. The other difficulty in diagnosis of erythroderma is non-specific histopathologic alterations. This patient had repeated systemic and topical steroids, so it can alter the histological findings and the nature of the inflammatory cell infiltrates.[4,9,10]

Management of erythroderma consists of nutritional assessments, correction of electrolyte and fluid imbalances, prevention of hypothermia, and treatment of secondary infections. Oral antihistamines could ease severe pruritus. Based on previous clinical trials, systemic agents such as cyclosporin A or methotrexate can be used. New immunomodulators like efalizumab, infliximab, etanercept, or alefacept are also recommended in refractory cases. Other agents like systemic corticosteroids may be necessary in drug reactions or in idiopathic erythroderma, with initial prednisone dose of 1-2 mg/kg/day and maintenance dose of 0.5 mg/kg/day or less. The dose of steroids must be tapering off with careful caution. Furthermore, topical therapy such as open wet dressings and bland emollients or low-potency corticosteroid ointments can be used.[11,12]

That comprehensive management had been applied to this patient. Furthermore, systemic agents with the patient who had chronic kidney disease should be considered with minimal side effects. Previous study showed that methotrexate should be avoided in patients with chronic kidney disease because the primary elimination of this agent is renal excretion. Other alternative systemic agents for patients with chronic kidney disease are etanercept and adalimumab.

Systemic corticosteroids are associated with exacerbations, so tapering should be done carefully. Some researchers stated that systemic corticosteroids can cause recurrences and flare in erythroderma. Meanwhile, systemic corticosteroids were chosen as the therapy in this patient due to this case was acute, there were no other systemic agents available, and the patient had no contraindications to the treatment, so systemic corticosteroids could be considered.[5,13-15] After the treatment, patient showed clinical improvement by diminished extensive erythema and desquamation.

Conclusion

Erythroderma is a rare but life-threatening disease. It can be caused by several etiologies. Establishing the diagnosis and management of this disease is challenging. The use of systemic corticosteroids with careful tapering has shown clinical improvement in erythroderma patients.

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Author's contribution

IA: Identification, diagnosis and management of the case, manuscript writing and critical review, has given final approval of the version to be published.

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References

1. Harper-Kirksey K. Erythroderma. *Life-Threatening Rashes*. 2018;265-77.
2. Hoxha S, Fida M, Malaj R, Vasili E. Erythroderma: A Manifestation of Cutaneous and Systemic Diseases. *EMJ Allergy Immunol*.2020.
3. Cuellar-Barboza A, Ocampo-Candiani J, Herz-Ruelas ME. A Practical Approach to the Diagnosis and Treatment of Adult Erythroderma. *Actas Dermosifiliogr (Engl Ed)*. 2018;**109(9)**:777-90.
4. Mistry N, Gupta A, Alavi A, Sibbald RG. A review of the diagnosis and management of erythroderma (generalized red skin). *Adv Skin Wound Care*. 2015;**28(5)**:228-38.
5. Susanto PM. Erythrodermic pasoriasis adult male patient treated with systemic corticosteroid: a case report. *Jurnal Kesehatan Tambusai*. 2022;**3(1)**:336-41.
6. Teran CG, Teran-Escalera CN, Balderrama C. A severe case of erythrodermic psoriasis associated with advanced nail and joint manifestations: a case report. *J Med Case Rep*. 2010;**4**:179.
7. Hu Y, Yin L, Xu J, Yin Z, Renal function of psoriatic patients: erythrodermic psoriasis has more significant hyperuricemia. *Biomed Res*. 2017;**28(6)**:2515-18.
8. Okoduwa C, Lambert WC, Schwartz RA, *et al*. Erythroderma: review of a potentially life-threatening dermatosis. *Indian J Dermatol*. 2009;**54(1)**:1-6.
9. Miyashiro D, Sanches JA. Erythroderma: a prospective study of 309 patients followed for 12 years in a tertiary center. *Sci Rep*. 2020;**10(1)**:9774.
10. Andrews G. Maximizing the Diagnostic Potential of Skin Biopsies. Kansas State Veterinary Diagnostic Laboratory. 2015.
11. Jean L. Bolognia MD. Erythroderma Treatment. Dermatology 4th edition. Elsevier. 2018.
12. G Madhuri, CH Kallappa, Kumar S. Erythrodermic psoriasis: a case report. *Manipal J Pharm Sci*. 2019;**5(2)**:71-3.
13. Cretu S, Salavastru C, Tiplica GS. Treatment of psoriatic erythroderma using systemic corticosteroids: A timeless option? *Dermatol Ther*.2020;**33(6)**:e14222.
14. Rao S, Bernshteyn M, Sohal R, *et al*. The Management of Erythrodermic Psoriasis Complicated by Cyclosporine. *Case Rep Dermatol Med*. 2020:5215478.
15. Heinrich M, Cook E, Roach J, Juarez RM, Payne D, Atkins R, Stetson C. Erythrodermic psoriasis secondary to systemic corticosteroids. *Proc (Bayl Univ Med Cent)*. 2019;**33(1)**:113-4.