Erythrodermic psoriasis secondary to systemic corticosteroids

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ABSTRACT

Erythroderma is an uncommon, potentially life-threatening condition, which has many causes, including uncontrolled psoriasis. We present a 58-year-old woman with psoriasis who, after an abrupt discontinuation of systemic corticosteroids, developed erythrodermic psoriasis with extensive body surface involvement requiring hospital admission. Erythrodermic psoriasis requires a high index of suspicion to diagnose it in a timely fashion and treat it to prevent complications.

KEYWORDS Erythroderma; erythrodermic psoriasis; psoriasis; systemic corticosteroids; topicals

Erythroderma is characterized by generalized erythema of the skin and frequently involves >80% to 90% of body surface area (BSA).^{1–3} It is a dramatic presentation that can arise from psoriasis, atopic dermatitis, cutaneous T-cell lymphoma, or drug reactions, but is idiopathic in about 25% of cases.^{1,2} Acute erythroderma can lead to complications such as water-electrolyte imbalance, hypothermia, cardiac failure, and secondary infections.^{1,4} The differential diagnosis includes other potentially life-threatening dermatologic conditions: drug reactions, erythema multiforme, Stevens-Johnson syndrome (SJS), and toxic epidermal necrolysis (TEN).⁵

CASE DESCRIPTION

A 58-year-old woman presented with a 2-month history of rash initially involving her left lower extremity, which became more diffuse. At an outside clinic, the patient told the provider that she had a history of psoriasis, and oral prednisone was prescribed and continued for 5 months. During this time, the dosage was up-titrated but symptoms persisted. The prednisone was stopped due to hematemesis and concern for gastritis. Over the following 2 weeks, the rash involved >90% of her total BSA. The rash was erythematous and exfoliative with significant sloughing of skin, accompanied by pruritus, burning pain, occasional clear discharge, and xerophthalmia. Notably, there was no mucosal membrane involvement. A punch biopsy was obtained,

revealing erythrodermic psoriasis (EP). The patient was treated with intravenous fluids, moisturizing eye drops, triamcinolone 0.1% ointment for the body, and hydrocortisone 2.5% ointment for the face, resulting in significant improvement of the rash.

DISCUSSION

The pathophysiology of erythroderma is not well understood.³ EP is believed to result from dysregulation of T cells, inflammatory cytokines and chemokines, and angiogenic growth factors, similar to the process of psoriasis.³ Psoriasis is a common cause of erythroderma along with dermatitis, drug reactions, and cutaneous T-cell lymphoma.^{2,3,6} Unstable psoriasis with new infection, hypocalcemia, and excessive use or withdrawal of systemic corticosteroids are likely causes of EP.^{2,4,6} Systemic corticosteroid cessation is classically associated with psoriasis flares, highlighting the importance of avoiding systemic corticosteroids in the management of psoriasis.^{4,6}

Initial symptoms of erythroderma include widespread erythema, fever, pruritus, pain, edema, malaise, and fatigue.^{2,3,6} It is important to rule out other conditions with high morbidity and mortality such as SJS, TEN, erythema multiforme, systemic drug eruptions, infection, and sepsis. Careful documentation of new medication timelines, pre-existing medical conditions, and comorbidities is imperative. A detailed physical exam is necessary, with focus on the appearance of the rash, BSA involvement,

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Table 1. Distinguishing features of erythrodermic conditions on the differential diagnosis

Erythrodermic condition	Unique features	Diagnostic workup	Other
Erythrodermic psoriasis	<ul style="list-style-type: none"> Nail bed involvement Mucous membranes spared 	<ul style="list-style-type: none"> Biopsy showing parakeratosis with loss of granular layer 	<ul style="list-style-type: none"> Skin tends to lose patchiness of psoriasis during flare
Autoimmune (BP, PV)	<ul style="list-style-type: none"> Flaccid or tense blisters, depending on level of split 	<ul style="list-style-type: none"> DIF will be positive in autoimmune conditions 	<ul style="list-style-type: none"> Can see eosinophilia in BP
Drug reactions	<ul style="list-style-type: none"> Morbilloform eruption Fever, arthralgias 	<ul style="list-style-type: none"> May see liver or kidney abnormalities in certain drug reactions 	<ul style="list-style-type: none"> Drug history is imperative
Pityriasis rubra pilaris	<ul style="list-style-type: none"> Islands of spared skin Hyperkeratosis and waxy orange appearance of palms and soles² 	<ul style="list-style-type: none"> Biopsy with alternating orthokeratosis and parakeratosis ("checkerboard pattern"), acanthosis, and hyperkeratosis of epidermis, follicular plugging⁷ 	<ul style="list-style-type: none"> Does not improve with systemic corticosteroids
Cutaneous T-cell lymphoma	<ul style="list-style-type: none"> Rash begins in "bathing suit" area Can see lymphadenopathy 	<ul style="list-style-type: none"> Flow cytometry shows increased CD4/CD8 ratio, aberrant T-cell receptors CD26, CD7, CD27⁸ 	<ul style="list-style-type: none"> Epidermotropism on biopsy⁸
SJS/TEN	<ul style="list-style-type: none"> Flaccid blisters (Nikolsky sign) Mucous membrane involvement⁹ 	<ul style="list-style-type: none"> Biopsy showing epidermal necrosis, subepidermal vesicle formation⁹ 	<ul style="list-style-type: none"> Drug history is imperative

BP indicates bullous pemphigoid; DIF, direct immunofluorescence; PV, pemphigus vulgaris; SJS, Stevens-Johnson syndrome; TEN, toxic epidermal necrolysis.

and involvement of mucous membranes, nails, and hair. SJS, TEN, and erythema multiforme will have impressive mucosal findings, and the absence of such should exclude these from the differential. Drug reactions are usually accompanied by laboratory abnormalities, such as eosinophilia or liver function abnormalities. Infections and sepsis are commonly associated with vital sign and laboratory abnormalities. By comparison, a patient with EP is more likely to have a history of poorly controlled psoriasis and a physical exam showing coalescing erythema with psoriasis plaques in classic locations, involvement of nails, and spared central face and mucous membranes.^{2,3,6} If the cause of erythroderma remains unknown, a dermatology consult must be considered. A biopsy can confirm a suspected etiology but is often nonspecific and is not necessary to diagnose EP.^{2,4} **Table 1** provides a differential diagnosis with distinguishing features of each disease.^{2,7-9}

EP is associated with several complications, with mortality as high as 64%.^{2,6} Fluid and protein losses and electrolyte imbalances are immediate concerns, and widespread cutaneous vasodilation can lead to thermoregulation, third spacing of fluids, and high-output cardiac failure.^{2,4,6} There is increased risk of superinfections and sepsis from cutaneous microbes, particularly *Staphylococcus aureus*.^{3,4}

Effective treatment of EP depends on supportive care and treatment of the underlying cause. All patients require fluid resuscitation, temperature control, and monitoring of electrolyte imbalances and hypoalbuminemia.^{2,4,6} Sedating antihistamines are helpful for both severe pruritus and pain.² For treatment of underlying psoriasis, first-line therapy includes topical steroids and topical nonsteroidals. Light therapy can be helpful in the outpatient setting, but phototherapy in an EP flare should be avoided as it is associated with recurrence.⁴ In severe cases, addition of an immunosuppressant such as cyclosporine, methotrexate, mycophenolate mofetil, or azathioprine may help but requires proper laboratory monitoring. Acitretin is another

option except in patients with liver failure or alcoholism. Biologic therapy can be considered if conventional therapy fails. Systemic corticosteroids should be avoided at all costs, as this is associated with psoriasis flares and recurrences.^{2,4,6,7}

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- Navrotski BRF, Nihi FM, Camilleri MJ, et al. Wet wrap dressings as a rescue therapy option for erythrodermic psoriasis. *An Bras Dermatol*. 2018;93:598–600. doi:10.1590/abd1806-4841.20186414.
- Mistry N, Gupta A, Alavi A, et al. A review of the diagnosis and management of erythroderma (generalized red skin). *Adv Skin Wound Care*. 2015;28:228–236. doi:10.1097/01.ASW.0000463573.40637.73.
- Singh RK, Lee KM, Ucmak D, et al. Erythrodermic psoriasis: pathophysiology and current treatment perspectives. *Psoriasis (Auckland, NZ)*. 2016;6:93–104. doi:10.2147/PTT.S101232.
- Rosenbach M, Hsu S, Korman NJ, et al. Treatment of erythrodermic psoriasis: from the medical board of the National Psoriasis Foundation. *J Am Acad Dermatol*. 2010;62:655–662. doi:10.1016/j.jaad.2009.05.048.
- Chen SX, Hinds BR, Goodman AM, et al. Erythrodermic psoriasis in a man with monoclonal B-cell lymphocytosis. *Cureus*. 2017;9:e1936. doi:10.7759/cureus.1936.
- Boyd AS, Menter A. Erythrodermic psoriasis. Precipitating factors, course, and prognosis in 50 patients. *J Am Acad Dermatol*. 1989;21:985–991. doi:10.1016/S0190-9622(89)70287-5.
- Klein A, Landthaler M, Karrer S. Pityriasis rubra pilaris: a review of diagnosis and treatment. *Am J Clin Dermatol*. 2010;11:157–170. doi:10.2165/11530070-000000000-00000.
- Nagler AR, Samimi S, Schaffer A, et al. Peripheral blood findings in erythrodermic patients: importance for the differential diagnosis of Sézary syndrome. *J Am Acad Dermatol*. 2012;66:503–508. doi:10.1016/j.jaad.2011.06.014.
- Creamer D, Walsh SA, Dziewulski P, et al. U.K. guidelines for the management of Stevens-Johnson syndrome/toxic epidermal necrolysis in adults 2016. *Br J Dermatol*. 2016;174:1194–1227. doi:10.1111/bjd.14530.