

CLINICAL VIGNETTE

Diagnosis and Management of Erythrodermic Psoriasis

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Introduction

Psoriasis is a well-known, chronic immune mediated inflammatory skin condition that presents as several different clinical subtypes.¹ One of the rare but aggressive forms of this disease is erythrodermic psoriasis (EP). EP affects only 1-2.25% of patients, yet when it occurs, will often involve at least 75% of a patient's body surface area.^{2,3} Identification of this potentially lethal variant involves clinical recognition of diffuse erythema in the context of known or suspected psoriasis. However definitive diagnosis commonly involves skin biopsy. Management includes supportive care, topical corticosteroids, and systemic anti-psoriatic drugs and biologic therapies that have had a documented benefit.^{4,5} This case discusses an EP presentation in a middle-aged man with known psoriasis, focusing on correct clinical diagnosis and subsequent management.

Case Presentation

A 50-year-old male with a history of treated latent Tuberculosis infection, guttate and plaque psoriasis presented to Urgent Care with a diffuse rash covering the majority of his body. Our patient had not been seen by a dermatologist for over one year. Previously, he had been treated with Methotrexate for his psoriasis without improvement, then was started on Etanercept 25mg weekly injection with significant improvement. Etanercept had been discontinued and the patient was maintained on topical clobetasol and triamcinolone. Two months prior to presentation, however, the patient ran out of all topical corticosteroids. Two weeks prior to his presentation he had noted onset of a rash with increasing distribution and intensity. He reported his skin was warm and pruritic and inconsistent with past flares of psoriasis. He also noted mild shortness of breath and decreased urine output but otherwise denied any infectious symptoms or joint pains. He denied any drug use, exposure to new chemicals, and had not started new medications.

Upon presentation to Urgent Care, the patient had a temperature of 36.2C, blood pressure 130/82, heart rate of 96, respiratory rate of 28, and was saturating 98% on room air. His physical exam revealed heart and lungs which were clear to auscultation and a benign abdomen. There was mild tenderness to palpation of his joints scattered diffusely, but no synovitis was noted in any joint. Skin exam showed an annular, erythematous, flaking, coalescing rash diffusely across his scalp, back, chest, abdomen and extremities (Figures 1-4). The rash was warm to touch but non-tender and no purulence was noted. There was no mucosal

involvement of his mouth or genitals. Initial laboratory tests were remarkable for a white blood cell count of 11.4K/cumm, hemoglobin 16.6 g/dL, and a lactic acid level of 2.6mmol/L. Basic chemistries, liver function tests, and chest radiograph were unremarkable.

The patient was admitted for evaluation and treatment of this diffuse rash. A dose of cefazolin 2g IV was given in the Urgent Care given concern for bacterial soft tissue infection. He was resuscitated with three liters of normal saline with normalization of his mild lactic acidosis, leukocytosis, and polycythemia. Based on the patient's clinical presentation and his history of guttate and plaque psoriasis, a presumptive diagnosis of erythrodermic psoriasis was made. SJS/TEN was considered however was less likely in the absence of new medications and lack of mucosal involvement. Cellulitis and staphylococcal scalded skin syndrome was thought unlikely given lack of fever or significant leukocytosis and thus antibiotics were discontinued. Malignancies such as mycosis fungoides was considered but thought less likely as the appearance was atypical and his known history of psoriasis. The patient was initially treated with intravenous fluids with initiation of topical Triamcinolone 0.1% cream and Fluocinolone 0.01% oil. Dermatology and rheumatology specialists agreed with the diagnosis and recommended restarting Etanercept, and his first dose was administered as an inpatient. On the day of discharge, the patient's symptoms were markedly improving. During follow up six months later, the patient reports a 90% improvement in his symptoms with resumed Etanercept injections and minimal need for topical steroids.

Discussion

Psoriasis is an immune driven disease with genetic and environmental risk factors.¹ Its more virulent derivative, EP, is also thought to involve a similar pathogenesis, but is not completely understood. EP presumably involves the Th1, Th2, and Th17 inflammatory pathways with more of a Th2 differentiation dominance.^{2,3} Because of the Th2 dominance, DMARDs have traditionally been a target for therapeutic options. There have also been rarer forms of EP that have a link to environmental or systemic illness. Systemic diseases known to have an association with EP include HIV, leukemia, T-cell Lymphoma, and gout.² Other associations are severe sunburns, alcoholism, withdrawal from common anti-psoriatic medications, contrast exposure, and certain drugs.^{2,3} Medications

reported triggering EP include acitretin, etretinate, infliximab, lithium, antimalarials, and trimethoprim/sulfamethoxazole.²

This patient's favorable outcome was in large part due to quick recognition and subsequent correct targeted medical therapy for erythrodermic psoriasis. The patient was first thought to have a severe cellulitis that did not improve with antibiotic treatment. This is a common misdiagnosis since EP may cause severe skin breakdown causing concomitant cellulitis, erysipelas, or sepsis.² Therefore, stabilizing the patient with fluid resuscitation and broad antibiotic coverage, particularly for *Staphylococcus Aureus*, may be important. We then focus on specific treatment for psoriasis.^{2,3}

Per most recent 2019 American Academy of Dermatology and National Psoriasis Foundation treatment guidelines, mild to moderate forms of psoriasis can be managed with topical medications and phototherapy.⁶ These are less common for EP given the severity of the disease, and preferred initial treatment for acute unstable cases is cyclosporine and infliximab.² Analyzed case series have advocated for the use of cyclosporine, an IL-2 inhibitor, for acute cases of EP.³ In one study of 33 patients, cyclosporine had a 67% complete remission rate after 3 months and an overall response rate of 94%.³ Nevertheless, addition or sole use of biologic agents is safe and likely necessary for moderate to severe forms of psoriasis, such as EP, that do not respond to initial systemic therapy. One agent proven to be beneficial is infliximab, a TNF-alpha inhibitor.^{3,5} A European study examined seven patients with EP who had all previously failed treatment with other therapies such as topicals corticosteroids, methotrexate, acitretin, and cyclosporine.⁵ They were given infliximab administered as three separate intravenous infusions, and showed rapid improvement without serious side effects.⁵

Etanercept, another TNF-alpha inhibitor, had been used to treat our patient with excellent results after he had failed treatment with methotrexate. He had been maintained on topical corticosteroids but had been off all therapy for two weeks prior to admission, which was his presumed trigger for EP. Rapid improvement with fluid resuscitation and topical corticosteroids cemented patient's correct diagnosis of EP, and diagnosis with skin biopsy was unnecessary. Etanercept was started to assist with long term resolution of EP and to help keep his chronic symptoms under control. Other studies have found similar benefits in using Etanercept monotherapy for long term management of EP. One series of 10 patients demonstrated at least 50% or greater psoriasis eradication at 12 weeks, and multicenter retrospective studies of six cases showed benefit, and at least two case reports reported analogous longitudinal improvement. Thus, there is relatively robust evidence that Etanercept is a viable option for long term treatment of EP.²

Conclusion

Erythrodermic psoriasis is an infrequently encountered breed of psoriasis, but it may be potentially life threatening if not diagnosed and treated quickly and appropriately. Biologic agents

continue to show promise as sufficient agents for acute and long-term care of the disease, but topical corticosteroids and supportive care remain important components of treatment. Our patient demonstrates the difficulty and importance in obtaining a timely and accurate diagnosis of EP. With prompt recognition and treatment, patients may experience rapid and prolonged remission of disease while avoiding potentially dangerous or morbid sequelae.

Figures

Images: Figures 1-4: *Extensive skin involvement of trunk, scalp, and extremities consistent with erythrodermic psoriasis.*



Figure 1



Figure 2



Figure 3



Figure 4

6. **Menter A, Strober BE, Kaplan DH, Kivelevitch D, Prater EF, Stoff B, Armstrong AW, Connor C, Cordoro KM, Davis DMR, Elewski BE, Gelfand JM, Gordon KB, Gottlieb AB, Kavanaugh A, Kiselica M, Korman NJ, Kroshinsky D, Lebwohl M, Leonardi CL, Lichten J, Lim HW, Mehta NN, Paller AS, Parra SL, Pathy AL, Rupani RN, Siegel M, Wong EB, Wu JJ, Hariharan V, Elmets CA.** Joint AAD-NPF guidelines of care for the management and treatment of psoriasis with biologics. *J Am Acad Dermatol.* 2019 Apr;80(4):1029-1072. doi: 10.1016/j.jaad.2018.11.057. Epub 2019 Feb 13. PMID: 30772098.

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