Severe Keratoderma Blenorrhagicum Simulating Psoriasis in Reactive Arthritis: A Case Report

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Abstract

Introduction: Severe keratoderma blenorrhagicum (KB) is a rare cutaneous manifestation of reactive arthritis (ReA) which can be indistinguishable from psoriasis, making the diagnosis challenging. This is a case of reactive arthritis in a 33-year-old female presenting with disabling, painful oligoarthritis which was accompanied by generalized pustular and scaly rashes simulating psoriasis.

Case: A 33-year-old female, Filipino, single with no known co-morbidities presented with disabling, painful oligoarthritis which was accompanied by generalized pustular and scaly rashes of two weeks duration. Her symptoms were preceded a few days earlier with a transient episode of conjunctivitis. She also reported having recently received treatment for "urinary tract infection". There were generalized hyperkeratotic papules with areas of desquamation overlying erythematous skin involving the scalp, hairline, trunk, and extremities including palms and soles, with onycholysis on all digits. The right wrist and both ankles were warm, swollen and tender, with dactylitis involving most toes. Dermatology consult concurred with the diagnosis of keratoderma blenorrhagicum associated with reactive arthritis, over psoriasis or psoriatic arthritis, and she was started on prednisone 60 mg/day; methotrexate (MTX) 20 mg/week and folic acid were added a week later. With dramatic resolution of both skin and joint involvement, prednisone was tapered to 10 mg/day over the next three weeks and MTX was maintained at 15 mg/week, with no rebound nor recurrence of symptoms.

Conclusion: Severe KB is a rare cutaneous manifestation of ReA which can be indistinguishable from psoriasis. The acute onset of symptoms, recent history of eye inflammation and genitourinary tract infection strongly favored ReA over psoriasis. A further hallmark of KB is the presence of sterile pustules on the palms and soles. Histologically, KB has more numerous pustules and massive hyperkeratosis compared to psoriasis. Moreover, the dramatic response to systemic steroids, without rebound nor recurrence upon steroid taper or discontinuation favors KB over psoriasis.

Keywords: reactive arthritis, severe keratoderma blenorrhagicum, case report

Introduction

Reactive arthritis (ReA) is a disease with a diverse clinical presentation belonging to a group of seronegative spondyloarthropathies.[1] It can be often difficult to diagnose because of its diverse clinical presentations. It occurs one to four weeks following an infection of the digestive or genitourinary tract, less frequently from a respiratory infection.[2, 3] ReA is classically characterized by a triad of arthritis commonly involving ankle and knee joints, urethritis in men and cervicitis in women, and ocular involvement usually conjunctivitis or uveitis, although a constellation of other symptoms and signs may also be present.[2, 4] Mucocutaneous lesions are seen in up to 50% of patients.[2] They can occur one day to six months after arthritis with average interval of one month, though cutaneous and the articular manifestations may appear simultaneously.[5, 6] A characteristic cutaneous finding is keratoderma blenorrhagicum (KB), described as hyperkeratotic erythematous skin lesions that clinically resemble pustular psoriasis and is found in 10% of ReA patients.[1] Typically, the palms and soles are almost always involved, however it may occur at the extensor surfaces of the legs, dorsal aspects of the toes, feet, hands, fingers, nails, trunk and the scalp.[2]

Case

A 33-year-old Filipino female, with no known co-morbidities presented with disabling, painful oligoarthritis which was accompanied by generalized pustular and scaly rashes of two weeks duration. Her symptoms were preceded a few days earlier with a transient episode of conjunctivitis, and treatment for a reported "unrinary tract infection". On consultation, she was in severe pain and had difficulty

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ambulating. There were generalized hyperkeratotic papules with areas of desquamation overlying erythematous skin involving the scalp, hairline, trunk, and extremities including palms and soles (Figure 1. A-F), with onycholysis on all digits (Figure 1. G). The right wrist and both ankles were warm, swollen and tender, with dactylitis involving most toes (Figure 1. H).

Erythrocyte sedimentation rate (ESR) was high (133mm/H) and rheumatoid factor (RF) was undetected. She also had anemia (97g/dL), leukocytosis (16.20x109/L), and thrombocytosis (823x109/L), as well as pyuria 3-6/hpf, bacteriuria 3+ and yeast cells with hyphal elements.

Because the patient presented with hyperkeratotic erythematous skin lesions that clinically resembled pustular psoriasis which was indistinguishable from the skin manifestations of ReA, the patient was referred to dermatology service which concurred with the stronger likelihood of keratoderma blenorrhagicum associated with reactive arthritis instead of psoriasis or psoriatic arthritis. Dermatology did not deem a skin biopsy necessary in view of the patient's acute manifestations and clinical history strongly suggestive of ReA. She was started on prednisone 60 mg/day; methotrexate (MTX) 20 mg/week and folic acid were added a week later. With dramatic resolution of both skin and joint involvement (Figure 2), prednisone was tapered to 10 mg/day over the next three weeks and MTX maintained at 15 mg/week, with no rebound nor recurrence of symptom.

Discussion

The diagnosis of ReA in our patient was largely based on the medical history and clinical findings – eliciting the clinical triad of urethritis, arthritis and conjunctivitis. Only 30% of patients develop the classic triad of ReA making the diagnosis very challenging.[1, 4] No laboratory tests are specific for ReA, and imaging findings are not conclusive in an acute episode.[1] In 1981, the American Rheumatism Association adapted the diagnostic criteria of peripheral arthritis for longer than one month, associated with urethritis, cervicitis, or both. Willkens et al[7] showed that this definition has 84.3% sensitivity and 98.2% specificity, further elucidating that the triad of arthritis, conjunctivitis, and urethritis has only 50.6% sensitivity, but with 98.9% specificity. ReA can be also accompanied by a constellation of extra-articular manifestations such as mucosal, cutaneous, ocular, and cardiac, as well as systemic symptoms of fever, malaise, and anorexia.

The classic cutaneous feature is keratoderma blenorrhagicum, which is observed in 10% of patients.[1] The initial lesion is a dull red macule which rapidly becomes papular and pseudovesicular/pseudopustular. Its color changes from yellow to orange-red as the roof thickens to form a hyperkeratotic plaque. The soles of the feet and the palms are almost always involved,[8] but the extensor surfaces of the legs, the dorsal aspects of the toes, feet, hands, fingers, nails, trunk and scalp are common sites.

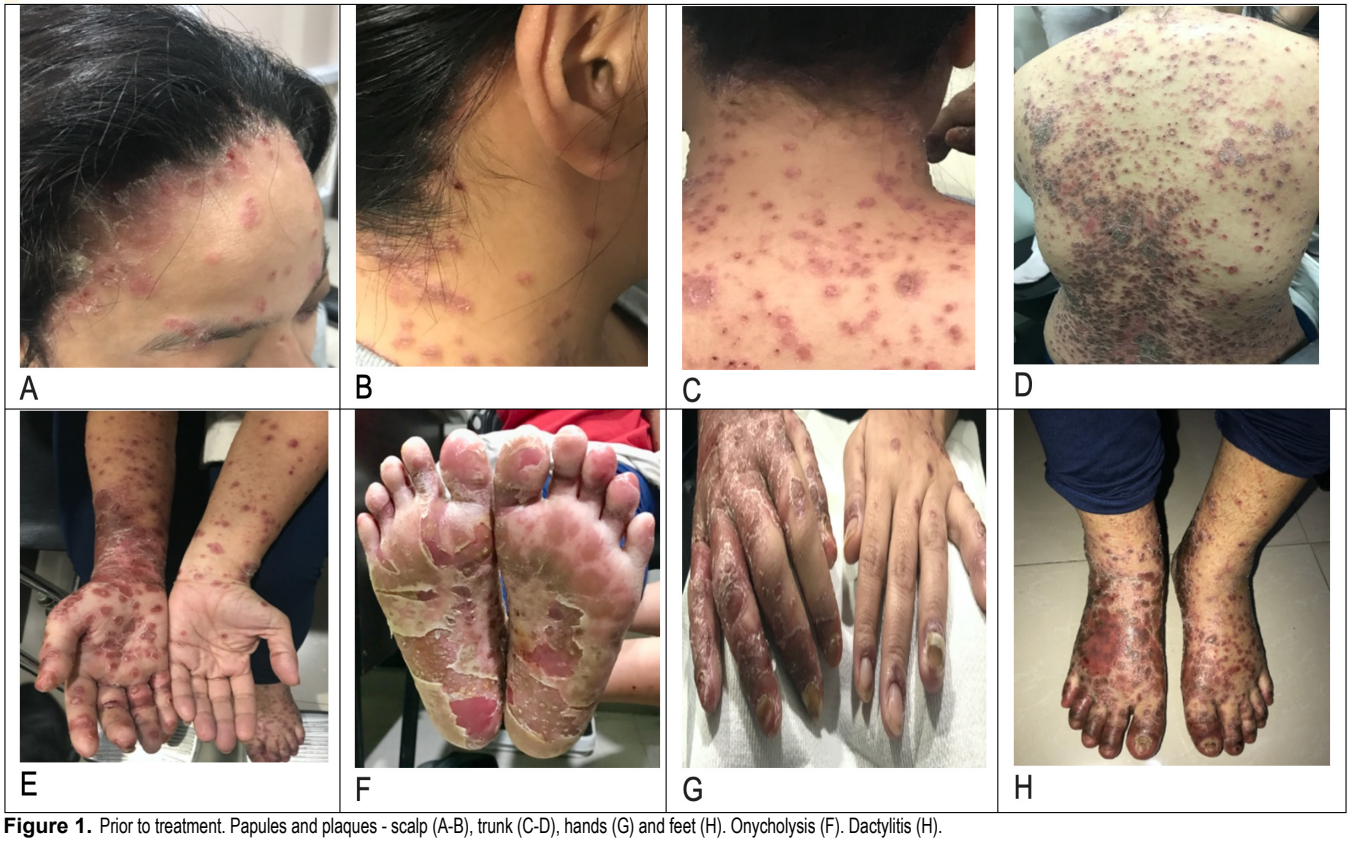

Figure 1. Prior to treatment. Papules and plaques - scalp (A-B), trunk (C-D), hands (G) and feet (H). Onycholysis (F). Dactylitis (H).

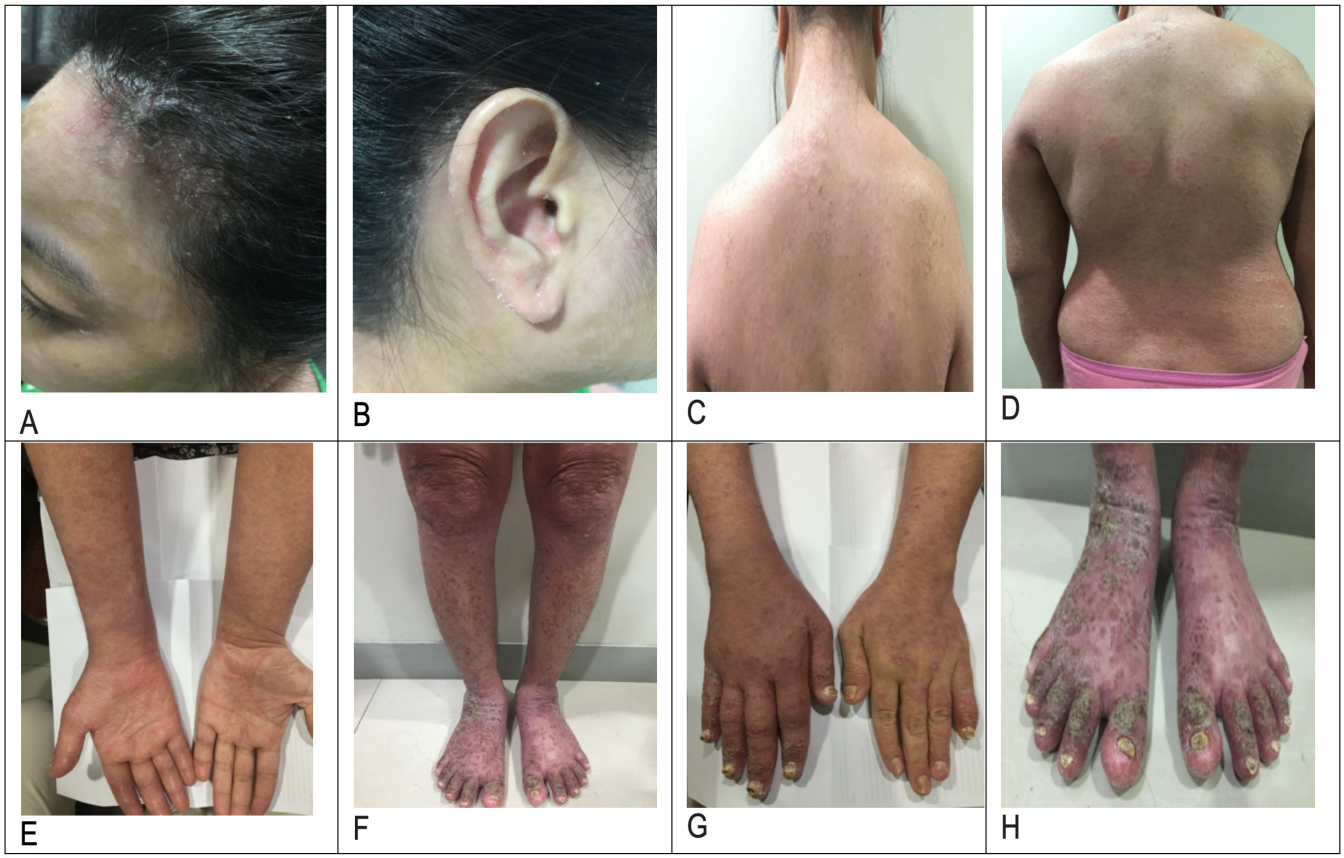

Figure 2. Three weeks after initiation of treatment - showing regression and clearing of skin lesions (A-H).

Occasionally, the skin lesions are very widespread and it may evolve into generalized exfoliative dermatitis (erythroderma). KB, however, are almost indistinguishable from psoriasis, but the hallmark feature in KB is the presence of sterile pustules on the palms and soles.[2,8] Though histologically similar, some authors contend that more numerous pustules and massive hyperkeratosis are observed in KB compared to psoriasis.[5] Another distinguishing feature is the time of onset of arthritis from skin eruption. In KB, the cutaneous and the articular manifestations may appear simultaneously, though generally the arthritis may precede the appearance of the skin eruption from one day to six months.[5] In psoriatic arthritis, psoriasis usually appears years before the onset of arthritic symptoms.

The tips of the fingers and toes may also be involved in KB. The nails are affected in 20%–30% of patients, initially appear as a painless red swelling along the nailfold, finally progressing to thickening of the nail plate.[2] Nails can become dystrophic, thickened and ridged. Nail pitting has also been reported. Subungual hyperkeratosis occurs 6%–12% of patients.

Keratoderma blenorrhagicum (KB) is an acute, self-limited process. It runs a course varying from six weeks to eight months, with a usual duration of four to six months. The arthritis may continue to persist even after the cutaneous lesions have disappeared. This contrasts with psoriasis which may last for a lifetime and is characterized by frequent relapses and remissions.[5]

The goals of treatment for ReA are to decrease pain and inflammation, minimize disability, and prevent relapse or progression to chronic disease where symptoms persist more than six months.[9,10] Skin lesions do not require therapy if mild. Oral steroids are indicated in severe extraarticular manifestations, polyarthritis, and systemic inflammation and the dosage is usually significant, about 20-40 mg/day of prednisone.[11] Our patient presented with a severe KB, and she was prescribed with both prednisone and MTX. MTX is a widely used disease-modifying antirheumatic drug (DMARD) with good effect. The major benefit in the early stages of MTX was noted in the mucocutaneous lesions and the onset of action appears to be rapid with mucocutaneous lesions than arthritis in general.[12] MTX is beneficial in the management of certain patients with ReA especially when refractory to conventional therapy.

Conclusion

This case illustrates the severe cutaneous manifestation of keratoderma blennorhagicum in ReA, which is often indistinguishable from psoriasis. The acute onset of symptoms, a recent history of eye inflammation and gastrointestinal or genitourinary tract infection strongly favor ReA over psoriasis. A further hallmark of KB is the presence of sterile

pustules on the palms and soles. Histologically, KB has more numerous pustules and massive hyperkeratosis compared to psoriasis. Moreover, the dramatic response to systemic steroids, without rebound nor recurrence upon steroid taper or discontinuation favors KB over psoriasis.

References

1. **Generali E, Ceribelli A, Massarotti M, Cantarini L, Selmi C.** Seronegative reactive spondyloarthritis and the skin. Clin Dermatol. 2015;33:531-537.
2. **Stavropoulos P g., Soura E, Kanelleas A, Katsambas A, Antoniou C.** Reactive Arthritis. J Eur Acad Dermatol Venereol. 2015;29:415-424.
3. **Carter JD, Hudson AP.** Reactive arthritis: clinical aspects and medical management. Rheum Dis Clin North Am. 2009;35:21-44.
4. **Wu IB, Schwartz RA.** Reiter's syndrome: the classic triad and more. J Am Acad Dermatol. 2008;59:113-121.
5. **Epstein E.** Differential Diagnosis Of Keratosis Blennorrhagica And Psoriasis Arthropathica. Arch Dermatol Syphilol. 1939;40:547-559.
6. **Krajewska-Włodarczyk M, Owczarczyk-Saczonek A, Placek W.** Cutaneous manifestation of reactive arthritis: Case report. Pol Ann Med. 2015;22:132-135.
7. **Willkens RF, Arnett FC, Bitter T, et al.** Reiter's syndrome. Evaluation of preliminary criteria for definite disease. Arthritis Rheum. 1981;24:844-849.
8. **Szamocki S, Martyn-Hemphill C, Green JSA.** Reactive arthritis: can't see, can't pee, can't climb a tree. Trends Urol Mens Health. 2016;7:17-20.
9. **Kim PS, Klausmeier TL, Orr DP.** Reactive arthritis: a review. J Adolesc Health Off Publ Soc Adolesc Med. 2009;44:309-315.
10. **Colmegna I, Cuchacovich R, Espinoza LR.** HLA-B27-Associated Reactive Arthritis: Pathogenetic and Clinical Considerations. Clin Microbiol Rev. 2004;17:348-369.
11. **Hannu T.** Reactive arthritis. Best Pract Res Clin Rheumatol. 2011;25:347-357.
12. **Lally EV, Ho G.** A review of methotrexate therapy in Reiter syndrome. Semin Arthritis Rheum. 1985;15:139-145.