



Case Report

Persistence of pustular psoriasis and psoriatic arthritis: A male patient's two-year experience with methylprednisolone and doxycycline therapy

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Abstract

Generalized pustular psoriasis (GPP) is a rare and severe inflammatory dermatosis characterized by sudden eruptions of sterile pustules and possible systemic involvement. Its variable presentation and lack of standardized diagnostic criteria often complicate timely recognition. We report a 32-year-old male with a three-month history of widespread erythematous, scaly, and pruritic lesions, progressing from the lower extremities to nearly the entire body. He had received long-term methylprednisolone and doxycycline for arthritis. Examination revealed extensive erythematous macules with thick scales affecting 64% body surface area, with elevated inflammatory markers (PCT 50.8, leukocytes 12.85, CRP 19.93). Microbiology was negative, and skin biopsy confirmed pustular psoriasis. Severity scores showed GPPASI 7.5, JDA-GPPSI 9, GPPGA 8, and DLQI 22. The patient improved after four weeks of intravenous fluids, methotrexate, and topical therapy. This case highlights the heterogeneous presentation of GPP and the role of standardized assessment tools in evaluating treatment response.

Keywords: Adult psoriasis, Generalized pustular psoriasis, Methotrexate.

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1. Introduction

Psoriasis is a persistent dermatological disorder induced by the immune system.¹⁻⁵ It impacts up to 30% of patients with psoriasis, resulting in joint inflammation (psoriatic arthritis). This systemic condition is associated with multiple health complications, including cardiovascular disease and metabolic disorders. Genetic and environmental factors contribute to its development. The prevalence varies worldwide, with an estimated 1.87% of the population in the United States affected. Plaque psoriasis is the most prevalent variety; however, additional variants include guttate, inverse, and pustular psoriasis. Generalized pustular psoriasis (GPP) is an uncommon, severe dermatological condition marked by the abrupt onset of extensive, visibly prominent primary sterile pustules on nonacral skin. Seven to nine its prevalence ranges from 2% to 3.5% globally, whereas in Indonesia, it is 2.5% of the population. It may or may not correlate with

systemic inflammation. Approximately 65% of patients with generalized pustular psoriasis (GPP) have a prior diagnosis of plaque psoriasis. Three to six The European Rare and Severe Psoriasis Expert Network (ERASPEN) indicates that the clinical progression of GPP may have at least one relapse or persist for over three months. Untreated GPP fares can be lethal due to possible systemic consequences such as bacterial infections, prerenal insufficiency, and cardiovascular failure. Corticosteroids can induce these effects upon administration or cessation.^{6,7}

The pathogenic mechanisms of GPP fares remain poorly elucidated. Recent advancements in our comprehension of the biological and genetic mechanisms underlying autoinflammation and autoimmunity have facilitated the identification of significant genetic variants associated with the occurrence and pathophysiology of GPP. Two to four they exhibit genetic divergence from psoriasis vulgaris and

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are associated with alterations in three genes constituting the skin immune system: IL36RN, CARD14, and AP1S3. This case report will examine the treatment trend of adult generalized pustular psoriasis at Soetomo Academic General Hospital.

2. Case Presentation

A 32-year-old male from Lamongan came to Dermatology OPC Soetomo Academic General Hospital on June 25th 2024, with reddish, scaly skin that felt itchy for the past 3 months. The patient complained of reddish, scaly skin that felt itchy for the past 3 months. The complaint started with the appearance of scaly skin on both legs and then appeared on the body and hands. The patient admitted that since last week, complaints of reddish skin and thick, itchy scales have become increasingly severe, almost all over the body and accompanied by complaints of fever. Complaints begin with redness, then contain small white fluids then merge and dry out into scales. The patient also complained of joint pain. For the last 2 years, the patient has been regularly seeing an internal medicine doctor and received the same medication, namely Methylprednisolone 4mg and Doxycycline 100mg, for 2 years without stopping. Then, 3 weeks ago, the patient went to the Dermatologist Professor and stopped all the drugs, and was asked to be admitted to the RSU for a biopsy and further treatment. The same skin complaint first appeared 11 years ago when the patient was in college with a high study load and stress situation. The patient admitted that the same complaint appeared and was not as severe as in the last 3 months and always improved when given cloderma ointment and oral medication, which the patient did not understand. The name was given by the dermatologist. The patient denies any history of allergies. History of arthritis for the last two years, while asthma, hypertension, diabetes, allergic, and drug allergic were denied. A family history of similar complaints in a sibling was noted and history of allergic were denied. The patient is married, works as a programmer and food trader, bathes twice a day with bar soap, and has difficulty walking due to leg pain; he is assisted by his family. He already consumed methylprednisolone 4 mg for 2 years and doxycycline 100 mg for 2 years.

macules with indistinct boundaries covered by thick, yellowish-white scales appearing at the facial region, no geographic tongue (**Figure 1**).

From the anterior and posterior trunk and abdomen, multiple erythematous macules appear, some with firm boundaries and some with indistinct boundaries, accompanied by thick white scales and erythematous macules with indistinct boundaries covered with thick white scales (**Figure 2**).



Figure 2: Trunk and abdominal dermatological findings

Multiple erythematous macules with indistinct boundaries covered by a thick white scale, multiple pustules appeared, there is lake of pus was seen in bilateral upper extremities (**Figure 3**).



Figure 3: Upper extremities dermatological findings



Figure 1: Head and neck dermatological findings

From physical examination found normal vital signs, with dermatological findings such as erythematous macules appearing with indistinct boundaries accompanied by thick white scales at the scalp region, multiple erythematous



Figure 4: Lower extremities dermatological findings

At the bilateral lower extremities region found, multiple erythematous macules with indistinct boundaries covered by thick white scale appear with no pussy and erosion and look

edematous. (**Figure 4**). Erythematous macules appear with unclear boundaries, some covered by thin white scales also found in the genital region.

Other tools findings showed results GPPASI: 7.5, JDA-GPPSI = 9, GPPGA = 8, Dermatology Life Quality Index = 22, and 64% body surface area (BSA) affected. Laboratory examination found an increase in PCT 50.8, Leucocyte 12.85, and CRP 19.93. Clinical microbiology from blood sample findings didn't show the growth of anaerobic bacteria. Gram and KOH examination found nothing. Skin biopsy showed characteristics of pustular psoriasis. The treatment plan involving IVFD NaCl 0.9% 1000cc/24 hours, methotrexate plans to enter 15mg (2tab-2tab-2tab), paracetamol 3x500mg if fever or pain, cetirizine 1x10mg, levertrans 2x/day for thick scales, vaseline album 2x/day on thin scale, and sodium fusidate cream 2x/day were given. After 4 weeks (July 29th 2024), multiple erythematous macules with indistinct boundaries covered by a thick white scale have already reduced compared to the initial treatment. Other findings also showed improvement for leucocyte result (9.04), Dermatology Life Quality Index = 7, GPPASI = 3.3, JDA-GPPSI = 4, BSA 36%, and GPPGA = 4.

3. Discussion

The most severe form of psoriasis, known as generalized pustular psoriasis (GPP), is typified by the development of intermittent extensive cutaneous erythema, which is covered in pustules, as well as systemic symptoms and extracutaneous manifestations like cholestasis, arthritis, uveitis, acute respiratory distress syndrome, and cardiovascular shock.¹⁻³ The occurrence of several fused sterile pustules, which may be localized or widespread, is a characteristic of GPP. A sterile pustule eruption with a diameter of 2 to 3 mm and a fever that lasts for several days are the first signs of generalized pustular psoriasis.⁷⁻⁹ A large crimson rash will first form on the patient's skin, followed a few hours later by a sterile pustule that spreads and partially fuses. Although GPP predilection can occur anywhere on the skin, it is more prevalent in the upper and lower extremities and torso.^{8,9} Our findings are in line with the patient complaints of reddish skin and thick, itchy scales almost all over the body and accompanied by complaints of fever.

Both newly healthy skin and pre-existing plaque psoriasis lesions may develop lesions.¹⁰ Periods of exacerbation may result in pustular exanthema of the oral mucosa, persistent geographic tongue, and signs of ulceration and desquamation on the lips. Von Zumbusch classified generalized pustular psoriasis into two subtypes: a full pustular eruption and a spread that includes systemic symptoms like fever and arthralgia.^{11,12} The pre-pustular phase, the flare phase, and the post-flash chronic phase—which may carry over into the subsequent flare phase—are the three stages of GPP. Throughout life, this recurrent pattern may persist in episodic form. Certain medications and the onset of illnesses can cause or exacerbate the illness.⁷⁻⁹

GPP can appear at any age, from childhood to the elderly with a median age around 50 years, which is different than our finding at 32 years old. Based on skin biopsy, macroscopic and histopathologic features of GPP can appear as acanthotic changes with neutrophil infiltration causing pustule formation, in line with our finding.^{13,14}

The systemic therapies presently utilized for individuals with GPP comprise acitretin, cyclosporine, methotrexate, or biologics. Methotrexate possesses anti-inflammatory, antiproliferative, and immunosuppressive characteristics. The medication inhibits dihydrofolate reductase (DHFR) and hence activates folic acid. This leads to the inhibition of the activity of thymidylate synthase which is a necessary component for the synthesis of purines and pyrimidines, and thus for the synthesis of DNA. The disruption in that route occurs during the S-phase of the cell cycle, resulting in growth inhibition and apoptosis. Thirteen to fourteen by obstructing DNA synthesis, MTX curtails epithelial hyperplasia, enhances the death of activated T cells, and impedes the chemotaxis of neutrophils. The precise mechanism of action of MTX in psoriasis vulgaris remains ambiguous, apart from its antiproliferative and immunomodulatory effects. Fourteen the patient exhibited improvement with methotrexate at an effective dosage of 15 mg (range 5 – 15 mg/week). Weekly dosages may reach 50 mg; however, certain studies suggest a maximum of 25 mg.

4. Conclusion

GPP is distinct from plaque psoriasis and involves sterile pustules related to specific genetic mutations. The assessment tools used (GPPASI, JDA-GPPSI, etc.) provide insights into the severity and extent of the disease, crucial for monitoring treatment efficacy.

5. Ethics and Consent

Written informed consent for publication of clinical details and images was obtained from the patient. Ethical clearance was not required for this case report.

6. Author Contribution Statement

All authors contributed to the conception and design of the study; acquisition, analysis, and interpretation of data; drafting and critical revision of the manuscript; and approval of the final submitted version in accordance with ICMJE authorship criteria.

7. Conflict of Interest

The authors declare that there is no conflict of interest.

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