


Generalized Pustular Psoriasis in a 4 Year-old Boy

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ABSTRACT

Background: Generalized pustular psoriasis (GPP) is a variant of psoriasis characterized by widespread erythematous skin studded with sterile pustules. In general, PPG is rare in children. Diagnosis is made from the clinical and histopathological examination. The rarity of GPP in children has resulted in a need for standardized guidelines. **Case**: A 4 year-old boy with a generalized pustular eruption and confluent scaly plaques on erythematous skin. Mild pruritus and general malaise were present. Histopathological examination confirmed diagnosis of GPP. Clinical improvement was achieved with a combination of topical corticosteroids, anti-histamine, and systemic corticosteroids for two weeks. **Conclusion**: A combination of topical corticosteroids, anti-histamine, and systemic corticosteroids can still achieve clinical improvement in children with GPP. Nevertheless, its adverse effects should always be considered, and strict monitoring is required.

Key word: Generalized pustular psoriasis, pediatric psoriasis, corticosteroid

ABSTRAK

Latar Belakang: Psoriasis pustulosa generalisata (PPG) merupakan salah satu varian klinis psoriasis yang ditandai oleh kulit eritema disertai erupsi pustul steril yang tersebar luas. Pada umumnya, PPG jarang pada anak-anak. Diagnosis berdasarkan pemeriksaan klinis dan histopatologis. Jarangnya kasus PPG pada anak-anak menyebabkan kurangnya pedoman tata laksana terstandarisasi. **Kasus:** Seorang anak laki-laki berusia 4 tahun dengan kelainan kulit berupa erupsi pustulosa generalisata beserta kumpulan plak berskuama pada dasar kulit eritema. Keluhan disertai gatal ringan serta rasa lemah dan lesu. Pemeriksaan histopatologis memperkuat diagnosis PPG. Perbaikan klinis tercapai dengan tata laksana kombinasi *corticosteroid* topikal, anti-histamin, dan *corticosteroid* sistemik selama 2 minggu. **Simpulan:** Kombinasi *corticosteroid* topikal, anti-histamin, dan *corticosteroid* sistemik mampu menghasilkan perbaikan klinis pada anak-anak dengan PPG. Meskipun demikian, efek sampingnya harus selalu diperhitungkan dan membutuhkan pengawasan ketat. **Rievanda Ayu Natasya, Diana Wijayati. Psoriasis Pustulosa Generalisata pada Anak Laki-laki 4 Tahun.**

Kata kunci: Psoriasis pustulosa generalisata, psoriasis pediatri, *corticosteroid*

Introduction

Leopold Von Zumbusch first reported generalized pustular psoriasis (GPP) in 1910.1 It is a rare and severe variant of psoriasis characterized by the acute onset of widely distributed pustular eruptions on inflamed erythematous skin. GPP affects only 3% global population and occurs more commonly in adults and rarely in children.[2] Various factors can trigger the disease, including infections, stress, drugs, vaccination, and trauma.[3] Early diagnosis and appropriate treatment are necessary to prevent complications and improve children's quality of life. A combination of topical and systemic therapy may be required.

Case

A 4 year-old boy was admitted to the Department of Dermatology with a 4-day history of generalized pustular eruption with confluent scaly plaques on the erythematous background. Lesions had initially developed on the scalp and face, involving most of the body surface within three days. He complained of mild pruritus and general malaise but no fever or other associated symptoms. There was no history of infection preceding the eruption. The patient had a history of food allergy to cow milk—no history of drug consumption before the eruption. A family history of any dermatoses, including psoriasis, was denied.

The patient looked moderately ill. Vital signs were within standard limits. Physical examination revealed erythematous plaques arranged in an annular pattern spreading all over the body, with multiple coalescing pustules covering the surface, forming a lake of pus. Most ruptured pustules around the trunk had resulted in superficial scaling and crusting with an erythematous basis. Several thick yellowish scales on the scalp were also found. There were no nail changes or mucosal involvements.

Complete blood count was only found increased leukocyte count (14,200 cells/mm3). No abnormality was seen in the liver function test. Gram examination revealed sterile pustules. Histopathological examination showed parakeratosis and acanthotic epidermis with elongation of rete ridges. Exocytosis of neutrophils and the presence of pustules in the stratum corneum forming' spongiform pustules of Kogoj' were identified. The subcorneal pustules had a thin

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roof of stratum corneum. The capillary vessels in the papillary dermis were dilated, giving the dermal papillae an edematous appearance. Perivascular inflammatory infiltrates also noted.

Based on the clinical and histopathological findings, generalized pustular psoriasis was diagnosed. The score of psoriasis area and severity index (PASI) was 35.6% (severe). The body surface area (BSA) involved was 90% (powerful). Children's dermatology life quality index (CDLQI) was 17 (very large effect). The patient was hospitalized for five days and treated with 2.5 mg dexamethasone IV injection per 12 hours, 25 mg ranitidine IV injection per 12 hours, 5 mg cetirizine syrup twice daily, and desoximethasone 0.25% cream. Physiologic saline gauze compression was applied to remove the crusts. The systemic corticosteroid was tapered off in the following week. After two weeks of treatment, some skin lesions had resolved, leaving a PASI score of 25, which indicates a 30% improvement.

Discussion

Psoriasis is a chronic immune-mediated inflammatory disease characterized by keratinocyte hyperproliferation. Pustular psoriasis is one of the clinical variants of psoriasis, classified into generalized pustular psoriasis (GPP) and localized pustular psoriasis. It is characterized by the presence of sterile pustules appearing on erythema.[2] GPP is a rare and potentially life-threatening form of psoriasis, representing approximately 3% of all clinical cases of psoriasis.[3] GPP is observed most frequently in middle-aged adults and rarely occurs in children.[4]

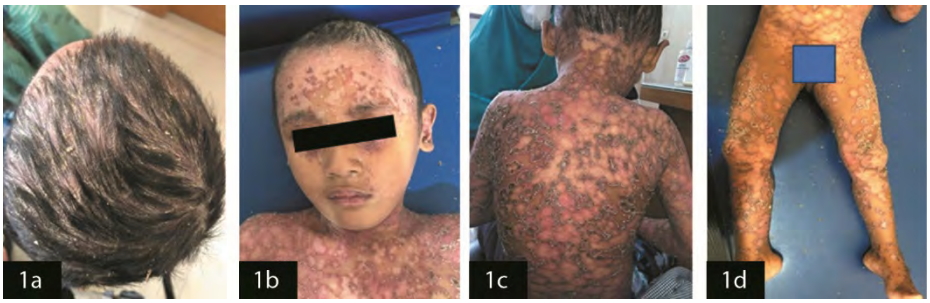

Figure 1. **1a.** Scaly plaques on the scalp. **1b-1d.** Multiple coalescing pustules on erythematous base, the majority had already dried off resulting in superficial crusts and scales.

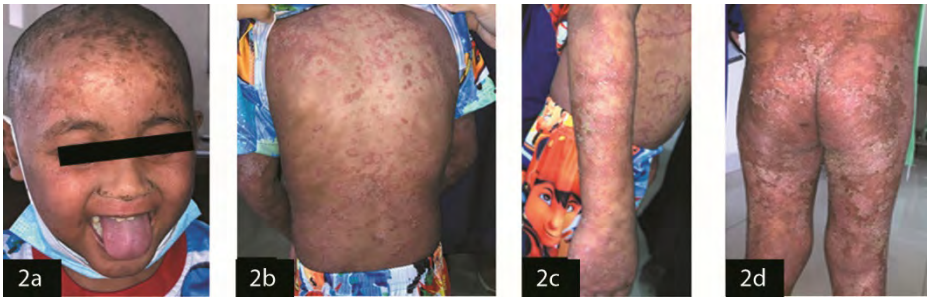

Figure 2.2a-2d. Decreased pustules, erythema, and scaly plaques after two weeks of treatment.

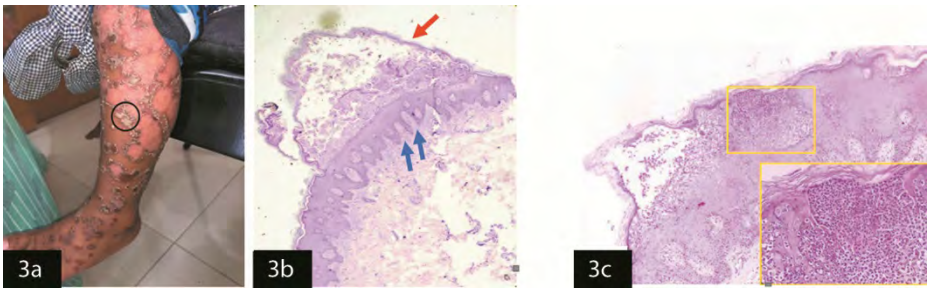

Figure 3. 3a. Punch-biopsy was conducted upon the lake of pus from lower left leg region (black circle). **3b.** Hyperplasia psoriasiform (blue arrow); subcorneal pustules (red arrow). **3c.** A large collection of neutrophils beneath stratum corneum, also called 'spongiform pustules of Kogoj'.

GPP presents a rapid onset of widespread erythematous inflamed skin studded with 2-3 mm sterile pustules that may expand and coalesce into irregular 'lakes of pus' before drying off and desquamation, resulting in profuse thin scales.[1,5] The pustules are disseminated over the trunk and extremities, including nail beds, palms, and soles. Patients may appear systemically ill, with high-grade fever, chills, malaise, and anorexia.1 In our case, the systemic symptoms were not extremely prominent; however, the skin lesion was by GPP.

The exact pathogenesis of GPP has not been fully elaborated. GPP patients have an increased frequency of the HLA-B27 allele compared to the general population. Many cases appear to be idiopathic; however, various triggers, including emotional stress, streptococcal infections, trauma, vaccination, and drugs such as penicillin and withdrawal from steroids, have been proposed to provoke GPP.3 A combination of genetic susceptibility and exposure to certain inciting factors lead to an upregulation of specific cytokines (e.g., IL-1, IL-17, IL-36, TNF-a, IFN-g) and accumulation of neutrophils in the epidermis.[6]

Differential diagnosis of GPP includes subcorneal pustular dermatosis (SPD). SPD is also a rare, chronic, recurrent pustular eruption characterized by subcorneal accumulation of neutrophils. A histopathological examination must be performed to distinguish them. Histological changes in SPD and GPP include subcorneal pustules filled with neutrophils and occasional eosinophils. However, the hallmark features of GPP include classic psoriasiform changes seen in the epidermis, such as parakeratosis and elongation of rete ridges.[7] In GPP, neutrophils that migrate from dilated vessels in the papillary dermis into epidermis aggregate beneath the stratum corneum to form the 'spongiform pustules of Kogoj'.[8] The histopathological result in our case suited the characteristic of GPP.

Complications of childhood GPP include hepatic, and renal failure, cholestasis, and severe infections, which occasionally lead to fatal outcomes.[4] Patients, may experience frequent flares, causing a reduction in quality of life. Possible psychological burdens of


this condition from a young age should be considered seriously.[3] The children's dermatology life quality index (CDLQI) score, in this case, showed a significant effect on the patient.

The rarity of GPP in children has led to a need for standardized guidelines. Thus, a general approach should be performed. The choice of the most appropriate treatment should consider the patient's age, the clinical severity of the disease, the impact on quality of life, the psychological burden of the condition, the presence of comorbidities, and the patient's previous treatments and preferences.[9]

Management involves topical therapy and trigger avoidance to reduce the number of flares, preserve skin surfaces, and provide physical relief from the disease. Systemic treatment is reserved for severe and unstable psoriatic illness, which may progress to significant complications. Topical therapy modalities include emollients, keratolytics, corticosteroids, vitamin D3 analogs, dithranol, tazarotene, and calcineurin inhibitors.[10] Acitretin, cyclosporine, etanercept, or methotrexate can be used safely as first-line therapeutic options in children with GPP. Second-line therapeutic options include adalimumab, infliximab, and UVB phototherapy.[11]

Topical corticosteroids have proved helpful in the pediatric population; however, they should be applied with caution due to their adverse effects. Infants, toddlers, and young children (age 0-6 years) are more susceptible to HPA (hypothalamic-pituitary-adrenal) suppression because their body surface area is more significant than their body volume.[12]

Regarding financial reasons, the patient could not afford the main systemic therapeutic options, such as cyclosporine or methotrexate. Hence, systemic corticosteroid was incorporated as a part of the treatment. Systemic corticosteroid use is generally avoided when treating GPP since the medication withdrawal will lead to a risk of flare. However, due to their anti-inflammatory, immunosuppressive, and vasoconstriction effects, a corticosteroid may be used in the acute stage and under circumstances where other therapeutic modalities are unavailable.[13] Furthermore, the psoriasis flare rate during systemic corticosteroid courses is very low and predominantly associated with mild flaring.[14]

In our case, 30% clinical improvement was achieved within two weeks of treatment with a topical corticosteroid, anti-histamine, and systemic corticosteroid, which was slowly tapered off in the following week. We are keeping this patient under follow-up to observe any spontaneous relapse or adverse effects from the therapy.

Conclusion

Generalized pustular psoriasis (GPP) in the pediatric population is uncommon. The choice of treatment for GPP in children should be determined individually depending on the severity of the disease, the patient's age and preferences, and the availability of therapeutic options. A combination of topical corticosteroids, anti-histamine, and systemic corticosteroids can still achieve clinical improvement in children with GPP. Nevertheless, its adverse effects on the pediatric population should always be considered, and strict monitoring is required.

Conflict of Interest

There is no conflict of interest in this study.

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