

Generalized pustular psoriasis with nail psoriasis in children: a case report



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ABSTRACT

Background: Pustular psoriasis in children is one of the clinical variants of psoriasis. It is classified into generalized pustular psoriasis (GPP) and localized pustular psoriasis. The aetiology of psoriasis in children has not been known but is believed to be multifactorial. Nail psoriasis rarely occurs in children who suffer from skin psoriasis, with an incidence lower than that reported in adults. The diagnosis is generally made from the clinical and histological examination. The choice of therapy depends on the severity of the disease. Until now, there have been no specific guidelines for the management of psoriasis in children.

Case report: A-15-years-old Balinese girl presenting with erythema, confluent scaly plaques over the trunk and extremities

with pustules localized on the lower extremity. She had a history of fever before the lesions appear. Right third digital nails examination showed subungual hyperkeratosis and onycholysis. Positive auspitz sign and karsvlek phenomena were found. Biopsy result suitable for psoriasis. The patient got improvement after treated with methotrexate tablet orally and desoximetasone cream topically within four weeks without any side effect.

Conclusion: Combination therapy with methotrexate tablet and desoximetasone cream give an effective result. However, the safety and side effects of methotrexate in children still need further monitoring.

Keywords: Generalized pustular psoriasis, nail psoriasis, pediatric psoriasis, methotrexate

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INTRODUCTION

Leopold Von Zumbusch first reported generalized pustular psoriasis in 1910.¹ It affects only 1-3% of the population globally. Pustular psoriasis is more common in adults and rarely in patients younger than 18 years old.² Generalized pustular psoriasis is often associated with identifiable triggers such as infection, stress, vaccination, and corticosteroid withdrawal. Nail lesion can be found in half the population of psoriasis patients, but rarely in children with prevalence range from 7-13%.^{3,4} The management of PPG and nail psoriasis in children is a challenge. The therapy aims to improve symptoms, prevent complications, and improve the quality of life in children. In mild PPG generally requires topical therapy. In moderate to severe PPG, systemic therapy can be added, as well as supportive therapy to improve the general condition of the patient.⁵

CASE REPORT

A-15-years-old girl presenting with erythema, confluent scaly plaques over the scalp, trunk, and extremities with pustules localized on the lower extremity since 14 days before admission to Sanglah

General Hospital. She also had a history of fever before the lesions appear. The patient also complains about the middle fingernail of the right hand was broken. The patient said that the complaint arose during preparation for the final exam of junior high school. History of the same symptom was denied. History of sore throat, pain during urination, toothache, cold, and cough were denied. History of erythema with thick scales on the elbows and knees were denied. No family member had the same complaints as the patient.

On physical examination found the general condition was moderate with compos mentis awareness. Vital signs within normal limit with visual analogue pain scales score was 1. Body mass index was within the normal range. Dermatological status showed multiple erythematous plaques cover with white until thick yellowish scales on the scalp, face, right to the left ear, anterior to posterior thorax and abdomen, upper and lower extremities. We also found multiple pustules on the left knee region, and some pustules were confluent and became a lake of pus on an erythematous base. Examination of the right third digital nails showed subungual hyperkeratosis and onycholysis (**Figure 1**). Auspitz sign and karsvlek phenomena were positive.

On nail dermoscopy, we found the destruction of the nail plate, onycholysis, and subungual hyperkeratosis. KOH 20% of the nail specimen showed no fungal elements. Gram examination from pustules was sterile. From the laboratory test, we found leukocytosis and a 1-fold increase in liver function test. We also did a histopathological examination and obtained hyperplasia psoriasiform with thinning of the supra-papillary plate, mild spongiosis, and neutrophil infiltrate between

parakeratotic layers (Munro's abscess). Capillary blood vessels dilate around the edematous dermis papillae. The superficial dermis contains perivascular lymphocytes infiltrate with a mild distribution of neutrophil (**Figure 2**). The conclusion from the biopsy results is more suitable for pustular psoriasis.

We diagnosed this patient with generalized pustular psoriasis and nail psoriasis. Psoriasis area and severity index (PASI) score was 20.8 (severe), body surface area (BSA) was 53% (severe), children dermatology life quality index (CDLQI) was 9 (moderate effect on child's life), and nail psoriasis severity index (NAPSI) was 4. The patient was treated with methotrexate tablet 7.5 mg/week intraorally, folic acid tablet 5 mg/week intraorally, desoximetasone 0.25% cream every 12 hours topically, urea 10% cream every 12 hours topically, and planned to consult ENT Department, Dental Department to find out focal infection and management of stress condition. After four weeks of therapy, there was a 56.2% improvement in the PASI score and 50% improvement in the NAPSI score.



Figure 1. Dermatological status. **1a-1k.** Erythematous plaques cover with white until thick yellowish scales. **1l.** Multiple pustules and some were confluent become lake of pus on an erythematous base. **1m.** Subungual hyperkeratosis and onycholysis (as shown in the red circle)

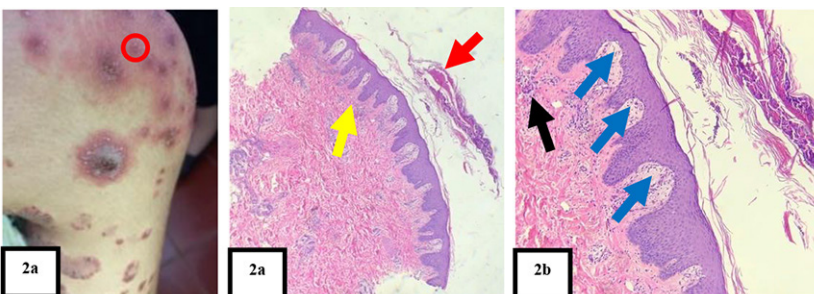


Figure 2. **2a.** The histopathological result from the pustule on the left knee region (red circle). **2b.** 40x magnification: hyperplasia psoriasiform (as shown by the yellow arrow), Munro's abscess (as shown by the red arrow). **2c.** 100x magnification: thinning of the supra-papillary plate and edematous dermis papillae (as shown by the blue arrows), perivascular lymphocytes infiltrate with a mild distribution of neutrophil (as shown by the black arrow)

DISCUSSION

Generalized pustular psoriasis is a rare form of childhood psoriasis, affecting 3% psoriasis patients, but in severe form can be potentially life-threatening.⁶ The clinical manifestations of GPP begin with fever and eruption of sterile pustules with a size of 2-3 mm accompanied by itching, based on erythema of the skin. The pustules can coalesce to form lakes of pus. Eventually, the pustules will dry out, and the skin will peel off.⁷⁻⁹ In our case, the clinical manifestation is in accordance with the literature.

In addition to skin manifestation, psoriasis can also affect the nails. Changes in psoriasis patient's nail can occur in the nail matrix, nail bed, or both. If regarding the nail matrix, pitting, leukonychia, red lunula, Beau's lines can be found, when it comes to nail beds, onycholysis, oil drop sign, splinter haemorrhages, subungual hyperkeratosis are found.⁴ In our case, we only found onycholysis and subungual hyperkeratosis on the third finger of the right hand.

The main differential diagnosis of GPP to be considered is acute generalized exanthematous pustulosis (AGEP). To assist in the diagnosis of GPP, a histopathological examination is performed. Histopathological features that often found in GPP are the psoriasiform epidermis (parakeratosis and elongation of rete ridges), spongiosis, thinning of the supra-papillary epidermis, neutrophil and

lymphocytes infiltration in the dermis, infiltration of neutrophils in the epidermal leading to the development of Munro's abscess.^{5,9} In our case, the histopathological result was in accordance with GPP.

Nail psoriasis has many features in common with onychomycosis. To distinguish from onychomycosis, it is necessary to do a dermoscopic examination, KOH, culture, and histopathological examination. In onychomycosis, the results of dermoscopy will show a longitudinal striae, spiked pattern, linear edge, and distal irregular termination. In contrast, nail psoriasis shows coarse pit, onycholysis, oil drop sign, blood vessel dilation, splinter haemorrhages, streaky capillaries. On KOH, culture, and histopathology examination, fungal elements will be found in onychomycosis but not in nail psoriasis.¹¹⁻¹³ In our case, no culture examination and nail biopsy were performed, from the results of dermoscopy, obtained onycholysis, subungual hyperkeratosis, and destruction of the nail plate. The results of this diagnosis do not provide a feature that can support a differential diagnosis. However, based on KOH 20% examination, there were no fungal elements, so from these results, we conclude it is more suitable for nail psoriasis.

The severity of psoriasis is used to determine therapy and evaluate therapeutic outcomes. Many scores have been used to assess psoriasis' severity, including PASI score, percentage of BSA, and CDLQI. Based on this PASI score, psoriasis is categorized into three groups, mild psoriasis if the PASI score is below 10, moderate psoriasis if the PASI score is 10-20, and severe psoriasis if the PASI score is above 20. Based on BSA, mild psoriasis if the percentage of BSA is less than 10%, the moderate degree if the percentage of BSA is between 10-30%, and the degree of severity if the percentage of BSA is more than 30%.^{14,15} Children dermatology life quality index is a standard measure of the quality of life in children related to health. The total score ranges from 0-30. The higher score obtained, the greater quality of life is disrupted.¹⁵ For nail psoriasis, the severity can be measured by the NAPSI score.¹⁶ In our case, the initial PASI score was 20.8 (severe), BSA was 53% (severe), CDLQI was 9 (moderate effect on a child's life), and NAPSI was 4.

The management of GPP and nail psoriasis in children depends on the severity of the disease. Until now, there have been no specific guidelines for the management of psoriasis in children. Education is the first step that needs to be done, both for children and parents. Topical therapy is still the first-line therapy for mild and localized skin psoriasis, but in the nail, psoriasis is limited

because of its penetration ability.^{5,17-19} Some topical therapy modalities that can be given for GPP are emollients, topical steroids, vitamin D analogues, topical calcineurin inhibitors, topical anthralin, and tazarotene.^{18,20,21} Management of chronic and severe psoriasis can be given systemic therapy. Systemic therapy options that have been reported to be effective in children with GPP include oral retinoids (etretinate, acitretin, isotretinoin), methotrexate, cyclosporin, and biological agents. Those therapies usually also improve nail psoriasis. Based on reported data, clinicians are determined to consider methotrexate, oral retinoids, and cyclosporine as the primary treatment options for PPG cases in children.^{6,18-20,22} On the use of methotrexate, folic acid is also routinely given to increase tolerability, by reducing gastrointestinal side effects, hepatotoxicity, and myelosuppression without disrupting the effectiveness of methotrexate.²³ In our case, based on PASI and BSA score, the patient was classified as severe GPP and got treated with methotrexate tablet 7.5 mg/week intraorally, folic acid tablet 5 mg/week intraorally, desoximetasone 0.25% cream every 12 hours topically, urea 10% cream every 12 hours topically and there was an improvement on skin and nail lesion at the fourth week of therapy without any side effect.

CONCLUSION

Generalized pustular psoriasis with nail psoriasis rarely occurs in children. There have been no specific guidelines for the management of psoriasis in children. Combination therapy with methotrexate tablets and desoximetasone cream gives an effective result. However, the safety and side effects of methotrexate in children still need further monitoring.

CONFLICT OF INTEREST

There is no conflict of interest regarding the publication of this paper

ETHICS IN PUBLICATION

The patient and her parents had signed inform consent regarding the publication of their respective photographs in the journal article.

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