Annular Pustular Psoriasis in a 6-year-old child: Case Report

*Patricia Gaile E. Espinosa, MD, Francisco D. Rivera IV, MD, FPDS **

ABSTRACT

Annular pustular psoriasis (APP) is a rare variant of pustular psoriasis occurring only in about 1-5.4% of children with psoriasis. Pustular psoriasis is triggered by endogenous and exogenous factors. Exogenous factors include stress, upper respiratory tract infection and withdrawal of systemic corticosteroids.

This condition usually requires systemic treatment. However, there are still no randomized controlled trials or standardized guidelines proving the advantage of any specific treatment modality over the other.

We report a case of a 6-year-old male who presented with diarrhea followed by appearance of erythematous annular plaques with pustules and nail changes. Punch biopsy showed pustular psoriasis. Patient was treated with topical betamethasone valerate and calcipotriol, and there was noted to have no recurrence of skin lesions for 4 months.

Keywords: *annular pustular psoriasis, children, pustular psoriasis, psoriasis, topical steroids*

**From the Department of Dermatology, Rizal Medical Center*

INTRODUCTION

Psoriasis is a chronic inflammatory skin disease, with a strong genetic basis, characterized by complex alterations in epidermal growth and differentiation and multiple biochemical, immunologic and vascular abnormalities, and a poorly understood relationship to nervous system function. Its root cause remains to be unknown.[1] Annular pustular psoriasis (APP) is a rare variant of pustular psoriasis occurring only in about 1-5.4% of children with psoriasis.[1-2]

Treating pediatric psoriasis can be challenging and requires careful compliance to a specific treatment regimen. There are currently no international standardized guidelines for medical treatment of pediatric psoriasis. To date, treatment is primarily based on published case reports, guidelines for adult psoriasis, expert opinions, and experience with these drugs in other pediatric disorders. The range of psoriasis treatments has expanded during the past several years, and multiple topical agents, phototherapy, and systemic and biological agents are available to date.[2]

This case report involves a 6-year-old child with annular pustular psoriasis and its management.

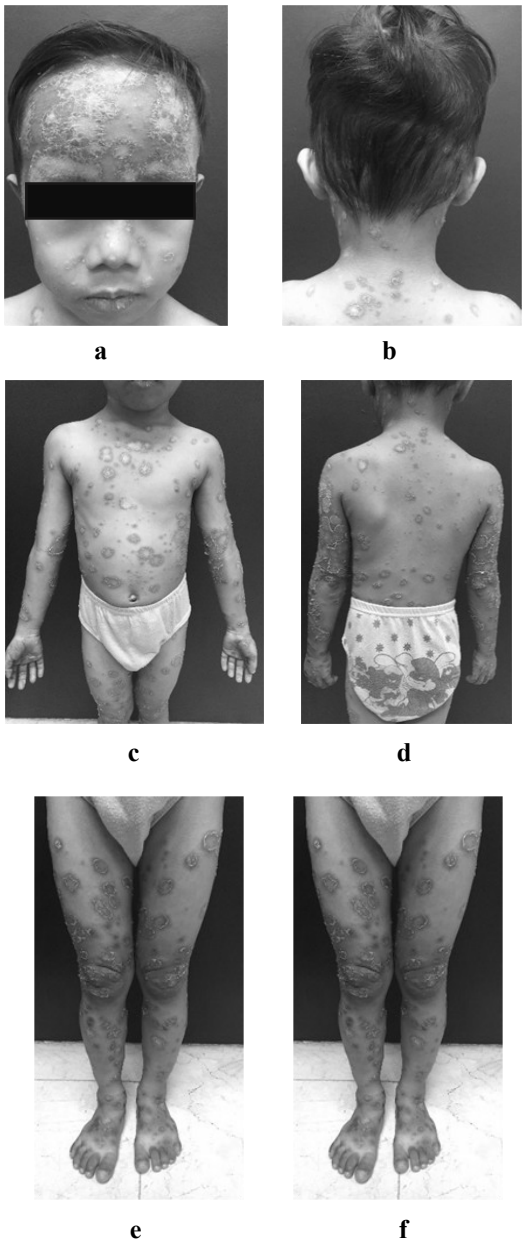
Case Presentation

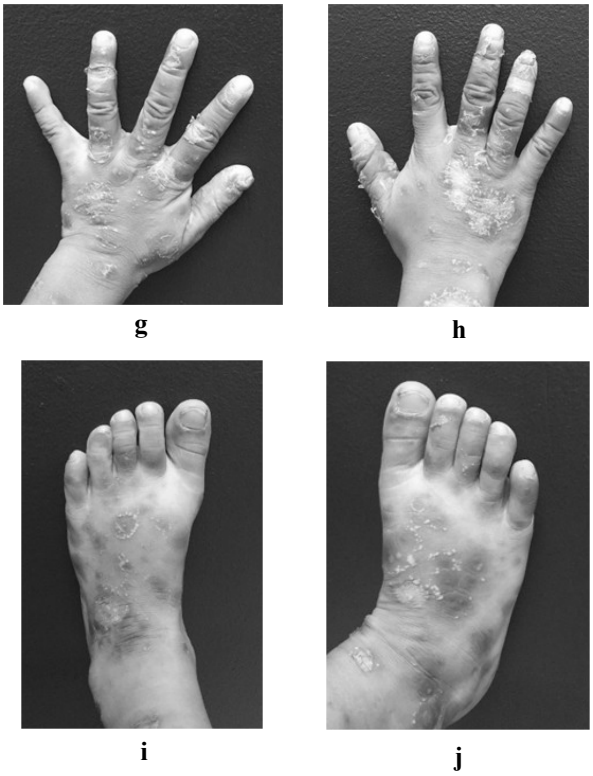
We are presented with a 6-year-old male who initially presented with multiple tender, pinpoint pustules on erythematous base over the right sole. This was preceded by 1 day history of loose, watery stools, accompanied by abdominal pain, decreased activity and poor appetite. There was noted progression of disease over the next 3 weeks, with pustules appearing over the trunk, upper extremities, face and scalp, followed by erythematous annular plaques on the same areas (Figure 1). There was no history of previous skin lesions.

During the course of the disease, patient did not have any cough, colds, fever, weight loss, lympha denopathy, alopecia, abdominal pain, dysphagia, dental caries, or difficulty breathing. There were no similar lesions were noted from any member of the family.

Based on the history and clinical presentation, patient was assessed to have annular pustular psoriasis. Psoriasis Area and Severity Index (PASI), which is used to assess the severity of psoriasis at a given point in time, is measured using the following parameters: erythema, scaling and induration of the lesions, and body surface area involvement.[3] During initial consult, the patient had PASI score of 21. Skin punch biopsy was performed and complete blood count with platelet (CBCP), urinalysis, erythrocyte sedimentation rate (ESR), blood urea nitrogen (BUN), creatinine, erum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), and chest radiography were requested which were all unremarkable. Biopsy findings were consistent with psoriasis. (Fig. 2)

Figure 1

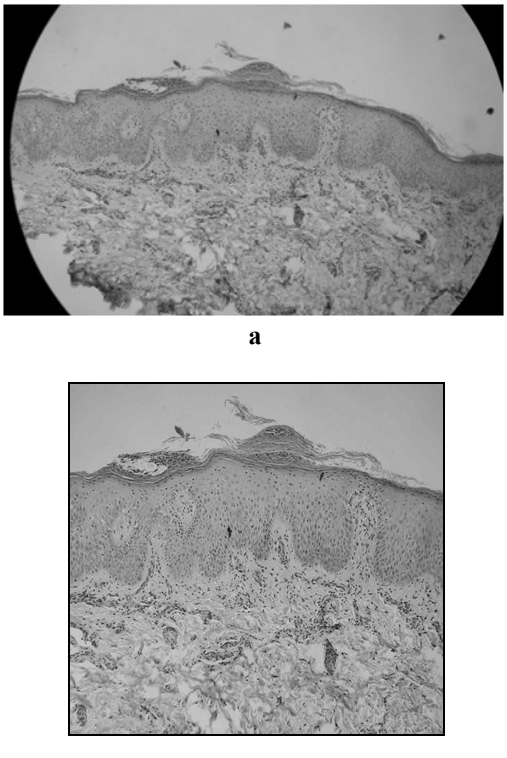




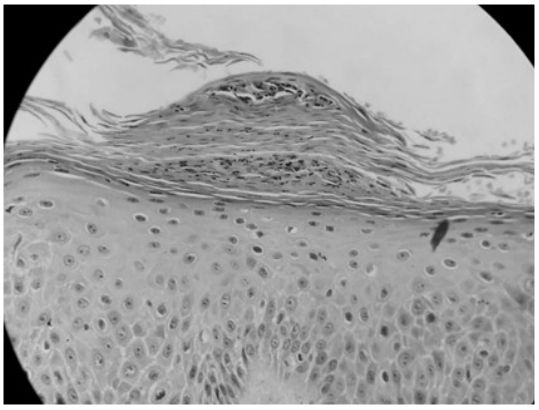
g h i j

Figure 1 shows patient with PASI 21, with multiple annular erythematous scaly plaques over the head (a,b), trunk and all extremities (c,d, e, f). Some digits have onycholysis, nail pitting and oil spots (g, h, i, j).

Figure 2



a b

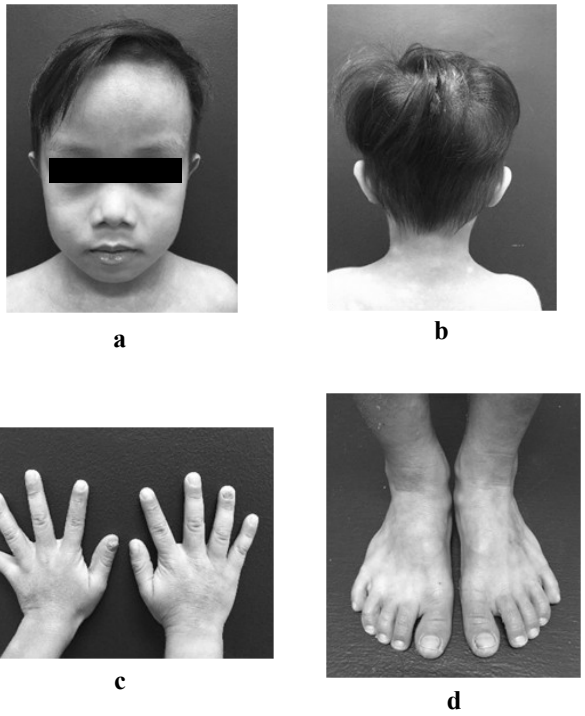


c

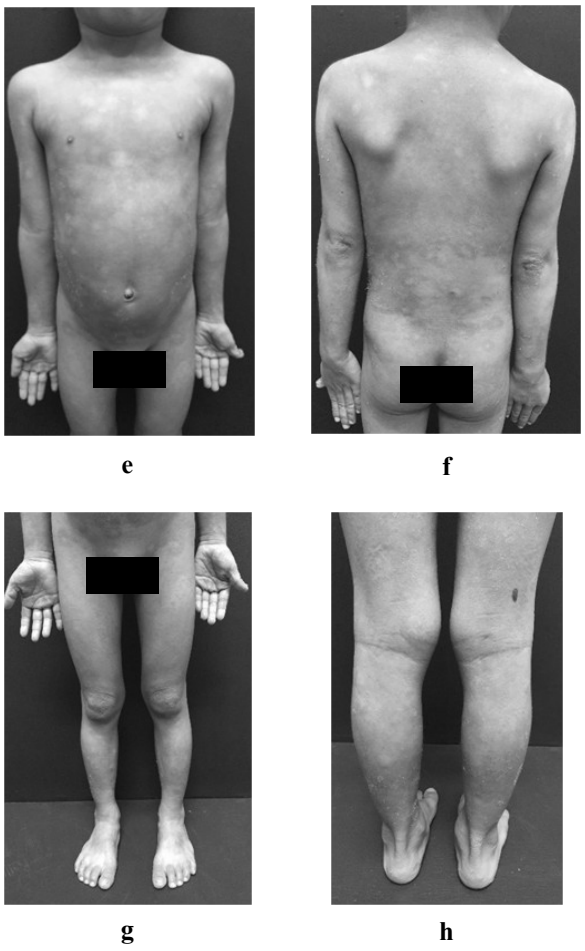
Figure. 2 (a-c) shows histopathologic findings of parakeratosis, hypogranulosis, elongation of rete ridges, neutrophilic aggregates within the stratum corneum, spongiosis in lower third of epidermis, active mitosis near basal layer, dilated blood vessels in the dermis, and perivascular and interstitial lymphocytic infiltrates.

Upon follow-up, he was started on betamethasone valerate lotion applied 2 times a day over the body, calcipotriol gel once a day in the morning over the face and Cetirizine for pruritus. After 9 days of treatment, the patient no longer have pruritus and showed a PASI score of 7.8. There is a 77% improvement of the lesions. There was now marked decrease in erythema and scaling over the whole body. (Fig. 3)

Figure 3



a b c d



e f g h

Figure 3 (a-h) shows patient with a PASI score of 7.8.

Discussion

A. Clinical Presentation and Diagnosis

In this case, the child presented with pruritic, annular erythematous scaly plaques and pustules, which is classic of APP.

Psoriasis has a worldwide occurrence, affecting 0.1 – 3% in various populations. It may appear at any age, however, it is uncommon under the age of 10 years.[1] Gelfand et al. found that the prevalence of psoriasis in childhood in the UK was about 0.55% in children aged 0-9 years and 1.37% in children aged 10-19 years.[4] Pediatric psoriasis was almost absent in an epidemiological study on childhood dermatoses performed in Asia, while pustular psoriasis is seen in only 1.0-5.4% of children with psoriasis.[2] In a Chinese survey of 277 children, only 1.1% presented with pustular psoriasis.[5]

Commonly, triggers of pustular psoriasis are emotional stress, upper respiratory tract infection or withdrawal of oral corticosteroids. However, in this patient, the child exhibited loose bowel stools and abdominal pain.[1-3,6]

In this case, pustular psoriasis can be diagnosed clinically, although biopsy can aid in the diagnosis, showing parakeratosis, loss of granular layer, elongation of rete ridges, neutrophilic aggregates within the epidermis (known as microabscess of Munro), dilated blood vessels in the dermis, and perivascular lymphocytic infiltrates.[1-2]

B. Management

Topical treatment was initially started while patient is awaiting results of laboratory exams.

There is no standard therapy for pustular psoriasis. Disease severity and extent of skin involvement help guide treatment. In children, acitretin, cyclosporine, methotrexate and etanercept are options for first-line therapy. However, no randomized controlled trials exist to prove the efficacy or superiority of a single treatment.[8]

The published first line and second line treatment for childhood generalized pustular psoriasis (GPP) are based on Class III evidences which include non-experimental descriptive studies, such as comparative studies, correlation studies, or case-control studies. First line systemic treatments include acitretin, cyclosporine, methotrexate and etanercept. Second line treatments include adalimumab, infliximab and UVB phototherapy. Topical treatment are started for localized cases and may be used in combination with systemic therapy. [8,9]

In a journal by Buetner et al.,they showed an algorithm for the treatment of childhood psoriasis with greater than 5% body surface area, without psoriatic arthritis. In our case, there was 60% BSA involvement. First line are topical agents, next would be ultraviolet B (UVB) phototherapy treatment. If UVB is not available, systemic treatment is warranted. For oral medications, the following may be considered: Adalimumab, Cyclosporine, Etanercept, Infliximab, Methotrexate, and PUVA.[9]

The use of topical therapy in childhood is the first line of treatment for skin-limited disease, sometimes in combination with oral antibiotics if associated with

infection. With chronicity of illness and in more severe cases, systemic therapy and phototherapy are added to help induce remission.[5]

In this case, we planned on using a topical agent in combination with a systemic treatment. We started the patient on betamethasone valerate lotion, which is a class 3-4 midpotent topical steroid.[10] Topical calcipotriol, a vitamin D analog, was also added to treatment. We planned on giving methotrexate afterwards. The patient responded after twice daily application for 1 week. From a 24 PASI score to a 7.8 PASI score. Improvement was 77% after treatment with both betamethasone valerate and calcipotriol. Since there was marked improvement with topical betamethasone valerate and calcipotriol, the use of oral treatment was no longer needed. There were no noted side effects from the topical used.

Similar to our case, Feicht, et al. also reported in 1982, a case of a 2-year old child who presented with GPP for 3 weeks and was treated with hydrocortisone 1% ointment. 100% clearance was seen. This shows that topical medication can be used and have a favorable outcome in pustular psoriasis. [11]

In our patient, there were no recurrences for 4 months after treatment. Mother religiously applied emollients over the body. Topical treatment with betamethasone valerate and calcipotriol may be another first line, safer treatment option for pustular psoriasis in children, although randomized controlled trials are still needed.

REFERENCES

1. Elder JT, and Gudjonsson JE. Psoriasis. In: Gilchrest B, Goldsmith L, Katz S, Leffell D, Wolff K, and Paller A, editors. Fitzpatrick's dermatology in general medicine. 8th ed. United States: McGraw Hill Companies, Inc.; 2012. p. 197-231.

2. Bronckers IM, Paller AS, van Geel MJ, van de Kerkhof PC, and Seyger MM. Psoriasis in children and adolescents: diagnosis, management and comorbidities. Pediatric Drugs Journal [Internet]. 2015. [January 15, 2018].17:373-384. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4744260/ doi:10.1007/s40272-015-0137-1

3. Gibson J and van Schie P. Clinical trial measuring systems in dermatology. In: Gorouhi F and Maibach H, editors. Evidence based dermatology. 2nd ed. United States: People's Medical Publishing House; 2011. p. 122.

4. Berlin JA, Gelfand JM, Margolis DJ, Neimann AL, Porter SB, and Weinstein R. Prevalence and treatment of psoriasis in the United Kingdom: A population-based study. Journal of the American Medical Association Dermatology [Internet]. 2005. [January 15, 2018].141(12):1537-1541. Available from: https://jamanetwork.com/journals/jamadermatology/fullarticle/401712 doi:10.1001/archderm.141.12.1537

5. Silverberg NB. Pediatric psoriasis: an update. Therapeutics and Clinical Risk Management [Internet]. 2009. [January 15, 2018]. 5:849–856. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2773753/

6. World Health Organization. Global report on psoriasis [Internet]. Geneva: World Health Organization; 2016 [January 15, 2018]. Available from http://apps.who.int/iris/bitstream/10665/204417/1/ 9789241565189_eng.pdf

7. Nikolic M, and Popadic S. Pustular psoriasis in childhood and adolescence: a 20-year single-center experience. Pediatric Dermatology [Internet]. 2014. [February 27, 2018]. 31(5): 1-5. Available from: https://www.ncbi.nlm.nih.gov/pubmed/24602000

8. Bebo BF, Jr, Hsu S, Kalb RE, Korman NJ, Lebwohl MG, Robinson A, et al. Treatment of pustular psoriasis: from the board of the national psoriasis foundation. Journal of the American Academy of Dermatology [Internet]. 2012. [January 15, 2018]. 67(2):279-288. Available from: http://www.jaad.org/article/S0190-9622(11)00304-5/pdfdoi:http://dx.doi.org/10.1016/j.jaad.2011.01.032

9. Beutner KR, Bhushan R, Elmets CA, Feldman SR, Gelfand, JM, Gordon, KB et al. Guidelines of care for the management of psoriasis and psoriatic arthritis. Journal of the American Academy of Dermatology [Internet]. 2009. [January 15, 2018]. 60(4):643-659. Available from: http://www.jaad.org/article/S0190-9622(09)00012-7/fulltext doi:http://dx.doi.org/10.1016/j.jaad.2010.11.055

10. Begolka WS, Berger TG, Bergman JN, Block J, Chamlin SL, Cohen DE, et al. Guidelines of care for the management of atopic dermatitis. Journal of the American Academy of Dermatology [Internet]. 2014. [January 15, 2018].71(1):116-132. Available from: http://www.jaad.org/article/S0190-9622(14)01257-2/fulltext doi: http://dx.doi.org/10.1016/j.jaad.2014.03.023

11. de Jager ME, de Jong EM, van de Kerkhof PC, and Seyger MM. Efficacy and safety of treatments for childhood psoriasis: a systematic literature review. Journal of the American Academy of Dermatology [Internet]. 2010. [January 15, 2018]. 62 (6):1013-1030. Available from: http://www.jaad.org/article/S0190-9622(09)00780-4/fulltext doi: http://dx.doi.org/10.1016/j.jaad.2009.06.048