

# Infantile acute erythrodermic pustular psoriasis with a good response to acitretin monotherapy

Iran J Dermatol 2019; 22: 120-122

**Dear Editor,**

Pustular psoriasis is an uncommon disease, particularly in childhood, which presents with fever, erythema and pustules. It is classified to generalized (von zumbusch), annular, localized and exanthematic pustular psoriasis. Some disorders can mimic the signs and symptoms of pustular psoriasis and are necessary to be differentiated, particularly acute generalized exanthematous pustulosis (AGEP). Generalized pustular psoriasis is a life-threatening condition, particularly in infancy; therefore, treatment should start as soon as possible <sup>1</sup>. In this paper, we reported a case of infantile generalized erythrodermic pustular psoriasis with a good response to oral acitretin. A previously healthy 8-month-old Afghan girl residing in Iran, as a result of cesarean section at the gestational age of 38 weeks with the birth weight of 2500 gr referred to our dermatology center with generalized scaling and erythroderma. She had normal growth and development. At first, erythematous plaques were developed on diaper area 3 weeks before admission that did not respond to topical antifungal and steroids, and then within 3 days, they extended abruptly

to the lower and upper extremities, trunk, face and scalp. Her past medical and family history was unremarkable.

On physical examination, low grade fever and erythematous patches and plaques with multiple pustules were seen (Figure 1). There were no conjunctivitis, nail changes, mucosal involvement, organomegaly or lymphadenopathy. Skeletal assessment was also normal.

Laboratory studies indicated leukocytosis, elevated C-reactive protein, and erythrocyte sedimentation rate (ESR). Other laboratory data, including blood culture, liver and renal function tests, coagulant parameters, immunoglobulines, electrolytes and interferon-gamma release assay tests were within the normal range. Gram's stain, potassium hydroxide preparation and bacterial cultures from pustules were negative. A skin biopsy from a pustular lesion demonstrated parakeratosis, elongation of rete ridges, thinning of the suprapapillary epidermis, spongiform pustules of Kogoj and microabscesses of Munro consistent with pustular psoriasis. Therefore, based on clinical and histopathological features, the diagnosis of erythrodermic generalized pustular psoriasis (GPP) was established. Oral acitretin (0.75mg/kg/



**Figure 1.** (A) Generalized scaling and erythroderma; (B, C) Multiple pustules, particularly on plantar surfaces and the face.



**Figure 2.** (A, B) Complete clinical improvement after 4 weeks of treatment with acitretin.

day for 4 weeks, then tapered and stopped after 6 months), oral hydroxyzine and emollient were administered. Significant clinical improvement was seen after five days, and skin lesions disappeared completely after 4 weeks (Figure 2). No relapses occurred in the next 6 months after discontinuation of the treatment. An informed consent was also obtained.

Our patient presented with erythrodermic GPP at an unusual age with a dramatic response to oral acitretin. Psoriasis is a common disorder in childhood, but GPP is extremely rare during the first decade of life, particularly in the first year of life<sup>2</sup>. Pathogenesis is not understood completely, but environmental (e.g. stress, infection and drugs) and genetic factors (such as mutation in IL36RN gene and deficiency of IL1 receptor antagonist) play a role<sup>3</sup>. Most common laboratory abnormalities are elevated ESR, leukocytosis, hypocalcemia, hypoalbuminemia, lymphopenia, and abnormal liver enzyme as well as electrolyte abnormalities<sup>3</sup>.

The common locations of psoriasis in childhood

are face, scalp, intertriginous and diaper areas. In the diaper area, it can be misdiagnosed with other disorders such as seborrheic dermatitis and candidiasis<sup>2</sup>, as our patient, whose disease started from the diaper area. Generalized pustular psoriasis can be a life-threatening condition; therefore, early diagnosis, differentiation from acute generalized exanthematous pustulosis, and proper treatment are necessary. Treatment of pustular psoriasis is not evidence-based yet, and suggestions are based on case reports and case series. The first choices of systemic treatment are acitretin, methotrexate, cyclosporine, and etanercept. Age, disease severity, systemic manifestations and comorbidities are among the factors, which should be considered to choose the best treatment<sup>4,5</sup>.

In summary, infantile erythrodermic GPP is rare; thus, a therapeutic challenge arises. We reported an 8-month-old girl with erythrodermic GPP with a good response to oral acitretin. An important side effect of the long-term therapy with acitretin in growing children is skeletal toxicity that should be carefully considered<sup>6</sup>.

Fahimeh Abdollahimajd, MD <sup>1</sup>

Zeinab Bizaval, MD <sup>1,2</sup>

Mohammad Shahidi Dadras, MD <sup>1</sup>

Mohammad Nikvar, MD <sup>3</sup>

1. *Skin Research Center, Shahid Beheshti University of Medical Sciences, Tehran, Iran*
2. *Molecular Dermatology Research Center, Shiraz University of Medical Sciences, Shiraz, Iran*
3. *Pharmacology Department, Islamic Azad University Science and Research Branch, Tehran, Iran*

*Corresponding Author:*

*Fahimeh Abdollahimajd, MD*

*Skin Research Center, Shohada-e-Tajrish*

*Hospital, Shahid Beheshti University of Medical Sciences, Shahr-dari St, 1989934148, Tehran, Iran*

*Email: fabdollahimajd@yahoo.com,*

*fabdollahimajd@sbm.ac.ir*

*Received: 9 November 2018*

*Accepted: 12 May 2019*

**Conflict of Interest:** None declared.

## REFERENCES

1. Chaves YN, Cardoso DN, Jorge PF, et al. Childhood pustular psoriasis: case report. *An Bras Dermatol.* 2010;85:899-902.
2. Dogra S, Kaur I. Childhood psoriasis. *Indian J Dermatol Venereol Leprol.* 2010;76:357-65.
3. Marrakchi S, Guigue P, Renshaw BR, et al. Interleukin-36-receptor antagonist deficiency and generalized pustular psoriasis. *N Engl J Med.* 2011;365:620-8.
4. de Oliveira ST, Maragno L, Arnone M, et al. Generalized pustular psoriasis in childhood. *Pediatr Dermatol.* 2010;27:349-54.
5. Chen P, Li C, Xue R, et al. Efficacy and safety of acitretin monotherapy in children with pustular psoriasis: results from 15 cases and a literature review. *J Dermatolog Treat.* 2018;29:353-63.
6. Subedi S, Yu Q, Chen Z, et al. Management of pediatric psoriasis with acitretin: A review. *Dermatol Ther.* 2018;31:e12571.