

# A Severe 'Generalized Infantile Pustular Psoriasis' Case Healed with Low Dose Cyclosporine A

## Düşük Doz Siklosporin A ile İyileşen Ağır Bir 'Yaygın İnfantil Püstüler Psöriazis' Vakası

Alişan YILDIRAN, MD,<sup>a</sup>  
Levent YILDIZ, MD,<sup>b</sup>  
Recep SANCAK, MD<sup>a</sup>

Departments of  
<sup>a</sup>Pediatric Immunology&Allergy,  
<sup>b</sup>Pathology,  
Ondokuz Mayıs University  
Faculty of Medicine, Samsun

Geliş Tarihi/Received: 17.11.2009  
Kabul Tarihi/Accepted: 28.01.2010

Yazışma Adresi/Correspondence:  
Alişan YILDIRAN, MD  
Ondokuz Mayıs University  
Faculty of Medicine,  
Department of Pediatric Immunology,  
Samsun,  
TÜRKİYE/TURKEY  
yildiran@omu.edu.tr

**ABSTRACT** Generalized pustular psoriasis is a rare disease which may be life threatening in children. Cyclosporine A is an immune suppressive agent that inhibits activated T cells via calcineurin inhibition. But the experience of its use in children is limited. Here, a 3-years old boy who diagnosed as pustular psoriasis by a skin biopsy but did not responded to acitretin treatment, then consulted to our clinic because of generalized erythrodermia and presumed sepsis due to loose of skin integrity was presented. In the severe forms of psoriasis that is classified as a Th1 disease, cyclosporine A can be the first choice drug.

**Key Words:** Psoriasis; therapeutics

**ÖZET** Yaygın püstüler psöriazis, çocuklarda oldukça nadir görülen, ağır formu hayatı tehdit edebilen bir hastalıktır. Bir immün süpresif ajan olan siklosporin A ise, hücre içi mediatör kalsinörini inhibe ederek aktive T hücrelerinin etkisini azaltmaktadır. Fakat siklosporin A'nın çocuklardaki kullanımı hakkındaki bilgilerimiz oldukça azdır. Bu yazıda, deri biyopsisi ile püstüler psöriazis teşhisi konulan ancak, asitretine yanıt alınamayan, yaygın eritrodermi ve deri bütünlüğünün kaybı sebebi ile sepsis tablosunda iken kliniğimize danışılan, düşük doz siklosporin A ile iyileşen üç yaşında erkek hasta sunuldu. Bir Th1 hücre hastalığı olduğu düşünülen psöriazisin ağır formlarında, immünsüpresif etkili siklosporin A ilk seçenek olabilir.

**Anahtar Kelimeler:** Psöriazis; tedavi bilimi

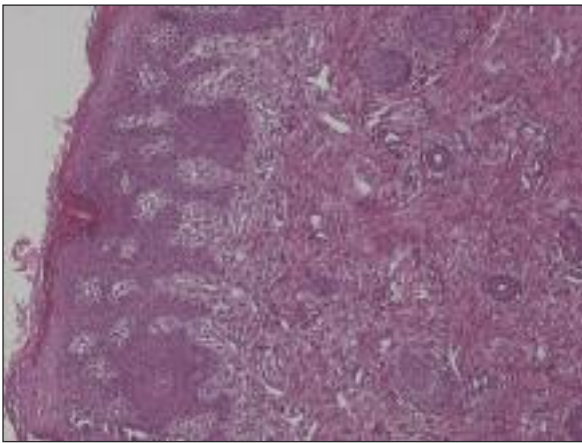
**Türkiye Klinikleri J Pediatr 2011;20(1):77-80**

Psoriasis, a papullosquamous skin disease, was previously thought of as a disorder primarily of keratinocytes but is now classified as a Th1 disease which is consistent with the relative under-representation of Th2 diseases such as atopic dermatitis. Generalized pustular psoriasis (GPP) (von Zumbusch psoriasis) is an acute form in which small, monomorphic sterile pustules develop in painful inflamed skin and may be life threatening in children.<sup>1</sup> Cyclosporine A (CsA) is an immune suppressive agent that inhibits activated T cells via calcineurin inhibition. But the experience of its use in children is limited.<sup>2</sup> A severe 'generalized infantile pustular psoriasis' case healed with low dose cyclosporine A is presented.

### CASE REPORT

A 3-years old boy who diagnosed as pustular psoriasis by a skin biopsy but did not responded to acitretin treatment, and then consulted to our clinic

because of generalized erythrodermia and presumed sepsis due to loose of skin integrity. Six months ago, a general pediatrician had prescribed him intramuscular cephalosporin for the complaints of purulent ear flux and truncal macular rash with a diagnosis of scarlet fever. The rash increased and transformed to vesiculopustular form, the patient had been hospitalized and a skin biopsy was performed for differential diagnosis of drug eruption, pustular psoriasis and seborrheic dermatitis. Since a skin biopsy specimen showed changes compatible with pustular psoriasis (Figure 1), namely acanthosis with elongation of rete ridges, parakeratosis and spongiosis, acitretin and emollient therapy had been started. A pelvic ultrasonography for his leg pain showed perthes disease. Systemic steroid (prednisolone) was added to acitretin for the last month. Because of no response after three months of this therapy and sepsis findings, the patient was consulted to our clinic. On physical examination, his length is at third percentile, weight is at tenth percentile. Alopecia, generalized erythrodermia and desquamation, nail dystrophies, mycoses on curved areas and anxiety were the striking findings (Figure 2). Laboratory examination revealed that, hemoglobin, lymphocyte, eosinophil, thrombocyte counts, protein, albumin, hepatic enzymes, serum immunoglobulins, isohemagglutinins and lymphocyte subgroups were all normal. But, neutrophil count and acute pha-



**FIGURE 1:** Skin biopsy showed acanthosis with elongation of rete ridges, parakeratosis and spongiosis (HEx10).



**FIGURE 2:** Alopecia, generalized erythrodermia and desquamation, nail dystrophies, mycoses on curved areas and anxiety were the striking findings.



**FIGURE 3:** After one month of CsA therapy.



**FIGURE 4:** On the sixth month of CsA therapy.

se response increased. All drugs except emollient were discontinued gradually and CsA was prescribed with a dose of 3 mg/kg/d orally. After one month of CsA therapy, his skin erythrodermia healed and his hair appeared but, there were desquamations (Figure 3). On the sixth month of therapy his condition was excellent (Figure 4). No adverse effect was seen.

## DISCUSSION

GPP is a rare and severe psoriasis form and may be life threatening in children.<sup>1,2</sup> Psoriasis was previously thought of as a disorder primarily of keratinocytes but is now classified as a Th1 disease which is consistent with the relative under-representation of Th2 diseases such as atopic dermatitis.<sup>1</sup> On this basis, primary systemic treatments for severe psoriasis include phototherapy, retinoids, methotrexate and CsA.<sup>4,5</sup> Of them, retinoids are the most commonly used oral drugs.<sup>1</sup> But this choice failed in our patient. As well as several case reports, Pereira et al described a pediatric case series with pustular psoriasis responded and well tolerated to CsA.<sup>4</sup> CsA has been studied extensively in pediatric atopic dermatitis. Its most serious side effects are nephrotoxicity and hypertension can be controlled easily with discontinuation of the drug.<sup>4</sup> Thus, no side effect was observed for six months in our patient. Dose of CsA is empiric and changes about 1 to 10 mg/kg/day. Kilic et al administered a low dose (1 mg/kg/d) to their patients.<sup>3</sup> We think that a dose of 3 mg/kg/d is also a low dose because our patient's clinic more severe (Figure 2) than the other patients in the literature that we can reached.

The main differential diagnosis in this patient was acute generalized erythrodermic pustulosis.<sup>6</sup> It was excluded with skin biopsy and clinical picture.

GPP is generally preceded to psoriasis vulgaris and more severe in adults.<sup>7</sup> Our patient's clinical picture was severe but no preceding psoriatic lesion.

Some triggering factors have been implicated such as medication, infections, solar irradiation and lastly, streptococcal infection in a child.<sup>7</sup> Thus, there was a possible scarlet fever and medication history in the presented case.

Although, there is a need to larger clinical case series to say that CsA is the first choice drug, we think that low dose CsA treatment is an effective and safe therapy in pediatric GPP.

## REFERENCES

1. Griffiths CE, Barker JN. Pathogenesis and clinical features of psoriasis. *Lancet* 2007;370(9583):263-71.
2. Benoit S, Hamm H. Childhood psoriasis. *Clin Dermatol* 2007;25(6):555-62.
3. Kiliç SS, Hacimustafaoğlu M, Celebi S, Karadeniz A, Ildirim I. Low dose cyclosporin A treatment in generalized pustular psoriasis. *Pediatr Dermatol* 2001;18(3):246-8.
4. Pereira TM, Vieira AP, Fernandes JC, Sousa-Basto A. Cyclosporin A treatment in severe childhood psoriasis. *J Eur Acad Dermatol Venereol* 2006;20(6):651-6.
5. Özçelik S, Akyol M, Özel F, Kızgın S, Marufihah M. [Two cases with pustular psoriasis and effectiveness of methotrexate]. *Turkiye Klinikleri J Dermatol* 2000;10(2):134-8.
6. Kopp T, Karlhofer F, Szépfalusi Z, Schneeberger A, Stingl G, Tanew A. Successful use of acitretin in conjunction with narrowband ultraviolet B phototherapy in a child with severe pustular psoriasis, von Zumbusch type. *Br J Dermatol* 2004;151(4):912-6.
7. Cassandra M, Conte E, Cortez B. Childhood pustular psoriasis elicited by the streptococcal antigen: a case report and review of the literature. *Pediatr Dermatol* 2003;20(6):506-10.