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

Düşük Doz Siklosporin A ile İyileşen Ağır Bir 'Yayın İfantil Püstüler Psöriazis' Vakası

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


Anahtar Kelimeler: Psoriasis; tedavi bilimi


Psoriasis, a papulosquamous 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.1 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.2 A severe ‘generalized infantile pustular psoriasis’ case healed with low dose cyclosporine A is presented.
because of generalized erythroderma 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 rush with a diagnosis of scarlet fever. The rush 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 erythroderma 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-
DISCUSSION

GPP is a rare and severe psoriasis form and may be life threatening in children. 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. On this basis, primary systemic treatments for severe psoriasis include phototherapy, retinoids, methotrexate and CsA. Of them, retinoids are the most commonly used oral drugs. 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. 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. 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. 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 erythrodernmic pustulosis. It was excluded with skin biopsy and clinical picture.

GPP is generally preceded to psoriasis vulgaris and more severe in adults. 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. 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.

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