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# IgA Deficiency and Psoriasis: Relevance of IgA in the Pathogenesis of Psoriasis

## Key Words

Immunology  
Immunoglobulins  
Psoriasis

## Abstract

A psoriatic patient with absolute deficiency of IgA is reported. The manifestations of psoriasis appeared at the age of 6 months and proved to be resistant to various treatments. The present case report and the data available in the literature on IgA and psoriasis all converge on the hypothesis that IgA is a systemic factor which belongs to the 'off switches' of the psoriatic process.

Inflammation, epidermal proliferation and abnormal differentiation of the epidermis are the well-known abnormalities in the pathogenesis of psoriasis. The immunology of psoriasis has been reviewed recently [1]. One part of the immunology in psoriasis regards the 'noncellular components'. An important observation, highlighting the relevance of non-cellular systemic factors in psoriasis is the fact that serum from patients with stable psoriasis repressed the K bner reaction, whereas serum derived from patients with unstable relapsing psoriasis failed to suppress this phenomenon [2]. This observation may indicate that serum factors in chronic stable psoriasis have an antipsoriatic potential, protecting the patient against progression.

Functionally, serum factors may modulate chemotaxis of monocytes and neutrophils and may interfere with angiogenesis and proliferation of fibroblasts [3-6]. Non-cellular systemic factors which might be involved in the immunopathogenesis of psoriasis are numerous. It has been suggested that IgA and IgA immune complexes are involved in the pathogenesis of psoriasis [7-9].

Recently, we had the opportunity to treat a patient with a complete deficiency of IgA who had psoriasis of the chronic plaque type with an unusual course: initiation of psoriasis was at the age of 6 months, and the course was resistant to various treatments.

## Case Report

A 23-year-old patient presented at our department with psoriasis. Already at the age of 6 months his parents had consulted the regional dermatologist who diagnosed the baby as having psoriasis inversa with typical psoriatic plaques at other localizations. During childhood he had episodes of psoriasis guttata. The skin lesions had persisted although the patient had been treated with topical corticosteroids, tar, dithranol, cyclosporin and phototherapy.

As a child between his second and twelfth year of life, the patient had recurrent episodes of otitis media, rhinitis and bronchitis. At the age of 5 years, he had a submandibular lymphadenitis. At the age of 15 years, he had furunculosis, lymphadenitis and recurrent sinusitis. No abnormalities were observed with respect to T-cell phenotype and T-cell proliferation. However, an absolute deficiency of IgA was shown: less than 1 unit/ml (normal range 50-250 units/ml). Total IgG, IgM and IgE were in the normal range. IgG4 was slightly reduced being 17 units/ml (normal >20 units/ml).

At presentation the patient had classical sharply demarcated erythematous plaques, compatible with the diagnosis of psoriasis inversa, localized in the joint and perianogenital regions, the axillae and umbilicus. On the scalp, elbows and legs he had sharply demarcated erythematous plaques and some punctate papules with a classical symmetrical distribution, compatible with the diagnosis of stable chronic plaque psoriasis. The course of the disease did not indicate any correlation between these recurrent infections and exacerbations of psoriasis.

Microbiological investigation of scrapings taken from the joints revealed *Staphylococcus aureus*. No fungi could be demonstrated by direct investigation and cultures. Histological investigation of a

biopsy taken from the left elbow revealed the classical picture of an active lesion with pronounced acanthosis, parakeratosis, a mixed inflammatory infiltrate and prominent micropustules of Kogoj and microabscesses of Munro.

The patient was admitted to the inpatient department for treatment with dithranol paste. However, due to irritation, already at low concentrations, the treatment had to be changed to a short-contact schedule with dithranol creams, tar preparations, corticosteroids, hydrocolloid dressing and UVB phototherapy. Due to insufficient improvement, additional treatment with acitretin (35–50 mg/day) was given. Following a 10-week admission, the intensified combined treatments had resulted in a considerable improvement, although total clearing was not reached.

## Discussion

The accumulation of T lymphocytes and polymorphonuclear leukocytes and the abnormal functioning of these cells are thought to be relevant and crucial aspects of the pathogenesis of psoriasis [1, 10]. Both cell types have been reported to possess receptors for the Fc portion of the IgA molecule [11–14]. With respect to functioning of the polymorphonuclear leukocyte, it has been shown that IgA paraproteins and polymeric forms of IgA inhibit the chemotaxis and the bactericidal activity of these cells [15–17]. Schröder et al. [18] reported on the neutrophil chemotaxis inhibiting activity of sera from patients with psoriasis and other neutrophilic dermatoses. The inhibition of chemotaxis proved to correlate with increases in serum IgA. Patients with chronic stationary psoriasis showed high IgA levels and reduced neutrophil chemotactic activity whereas those patients with relapsing psoriasis had normal IgA levels and increased neutrophil chemotactic activity [18]. This observation lends support for the supposition that an increase in serum IgA might be an antipsoriatic mechanism, stabilizing the psoriatic process. However, although neutrophil chemotaxis inhibiting activity of sera from patients with psoriasis has been shown to depend upon IgA polymeric forms *in vitro*, it is not clear whether this is important *in vivo* [19].

Different groups have reported that serum IgA levels are increased in psoriasis [8, 20–24]. Circulating IgA immune complexes have also been reported to be increased in psoriasis [9, 25]. Guilhou et al. [8] reported that the increase in IgA is specific for psoriasis as no patient with eczema had such an increase. By these authors, the increase in IgA-containing immune complexes was observed in 67% of a group of psoriatics whereas a similar increase was found in only 1 out of 25 patients with hyperkeratotic disorders. Fraser et al. [20] reported increased levels of IgA in patients with dermatitis herpetiformis, acne vulgaris, rosacea and discoid lupus erythematosus. A consistent observation by various

groups is the correlation between increased serum levels of IgA and the extent of the skin manifestations and severity of arthritis [18, 25]. However, it has been reported that psoriasis with arthropathy may coexist with a selective IgA deficiency [26]. Two groups have reported on decreased IgA levels in psoriatic patients [27, 28]. The correlation between disease activity and IgA levels [18, 25] suggests that IgA is of significance in the pathogenesis of psoriasis. It is attractive to hypothesize that IgA is an 'antipsoriatic factor'. Indeed, relatively high levels of IgA have been measured in chronic long-standing plaque psoriasis and in patients with extensive lesions, in contrast to the low levels of IgA in relapsing and guttate-type psoriasis [18]. Patients with decreased or undetectable IgA levels and psoriasis [26–28] may have an unfavorable prognosis according to this hypothesis.

The present case report is in line with a protective role of IgA with respect to the relapse of psoriasis. The unusual course – onset at the age of 6 months and the unstable features, characterized by extreme dithranol sensitivity and therapy resistance – in a patient with IgA deficiency may suggest that IgA indeed belongs to the 'off switches' of the psoriatic process. However, it is remotely possible that the episodes of recurrent infections might have been responsible for the onset of psoriasis in early childhood.

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