

## Psoriasis: An Unusual Autoimmune Manifestation in a Boy with Common Variable Immunodeficiency

Sir,

Common variable immunodeficiency (CVID) is the most common clinically significant primary immune deficiency and predisposes an individual to recurrent infections, inflammatory and autoimmune complications. We report psoriasis as an associated autoimmune disorder in an adolescent boy with CVID and also emphasize the role of immunoglobulin replacement therapy in its management.

A 13-year-old boy was symptomatic since the age of 2 years. He had had recurrent episodes of pneumonia and otitis media. He presented to us at the age of 10 years with an episode of community-acquired pneumonia. After ruling out human immunodeficiency virus (HIV) infection, he was evaluated for an underlying primary immunodeficiency. Results of his investigations are given in Table 1. His overall clinical and immunologic

profile was suggestive of CVID. CD81, CD19, B cell activating factor receptor (BAFF-R) protein expression on B cells, and inducible co-stimulator (ICOS) expression on T cells were normal. No mutation was detected in the transmembrane activator and calcium-modulator and cyclophilin ligand interactor (TACI). He was initiated on cotrimoxazole prophylaxis and parents were counselled for immunoglobulin replacement therapy.

At the age of 13 years, he presented with erythematous, painless, and nonitchy skin lesions, noticed for last 6 months. On examination, there were well-defined erythematous plaques with fine silvery scales on the surface distributed all over the body [Figures 1a and b]. The morphology of lesions was consistent with psoriasis vulgaris. Histopathology showed hyperkeratosis, parakeratosis, and acanthotic epidermis with focal loss of

granular cell layer and a pustule in the parakeratotic layer along with focal spongiosis. His psoriasis area severity index (PASI) was 31. Topical coal tar, emollients, and oral acitretin were initiated. He was also treated with 15 gm of intravenous immunoglobulin (IVIG) replacement therapy which was repeated every 4 weeks. There was significant improvement in psoriasis and majority of the lesions disappeared in the next 2 months [Figure 1c and d]. At present, he is being given immunoglobulin replacement therapy every 4 weeks; lesions of psoriasis have healed completely and had no new infections in the last 6 months.

Autoimmunity in CVID has been reported in up to 20–30% of patients, with hemolytic anemia

and thrombocytopenia being the most common manifestations.<sup>[1]</sup> There is no well-known mechanism for the development of autoimmunity in CVID; however, the role of genetic factors and various cellular mechanisms have been proposed.<sup>[2]</sup> Vitiligo, lichen planus, alopecia, and psoriasis are the most common autoimmune skin diseases in CVID.<sup>[1,2]</sup> Psoriasis is an autoimmune skin disorder characterized by keratinocytes proliferation and abnormal differentiation. Among the various subtypes, plaque psoriasis (psoriasis vulgaris) is most frequent.<sup>[3]</sup> Psoriasis is not a common autoimmune manifestation in CVID and has been reported to be seen in less than 1% of patients.<sup>[2,4]</sup> This association has not been reported previously from India. In general, therapy for psoriasis depends on its type, extent of disease, and site of involvement and involves both topical as well as systemic therapy.<sup>[4]</sup> Recently, biological agents and high dose immunoglobulins have also been demonstrated to be helpful in refractory psoriasis cases.<sup>[3,5]</sup> The mechanism by which IVIG improves autoimmunity in CVID has been found to be due to the modulation of function of regulatory T cells, B cells, dendritic cells, and NK cells.<sup>[6,7]</sup> Index child was initiated on IVIG replacement therapy and has shown marked improvement in psoriasis. We emphasize that IVIG replacement therapy has a significant role in the management of psoriasis in CVID patients.

*Financial support and sponsorship*

Nil.

*Conflicts of interest*

There are no conflicts of interest.

Ankur Kumar Jindal, Amit Rawat,  
Avinash Sharma, Sunil Dogra<sup>1</sup>,  
Deepti Suri, Surjit Singh



Figure 1: Well-defined erythematous plaques with fine silvery scales over the trunk consistent with psoriasis vulgaris (a and b); significant improvement after 2 months showing clearance with residual hyperpigmentation (c and d)

Table 1: Details of laboratory investigations with normal reference range

Investigation	Result	Normal reference range
Hemoglobin (gm/L)	96	120-140
White blood cell counts ( $\times 10^9$ cells/L)	7.6	4-11
Differential counts	$N_{60}L_{28}M_6E_6$	
Platelet counts ( $\times 10^9$ /L)	190	150-400
IgG (mg/dl)	<206	540-1610
IgM (mg/dl)	<25	50-190
IgA (mg/dl)	40	80-280
CD 19 <sup>+</sup> B cells	17.6%	5-19%
CD3 <sup>+</sup> T cells	74.9%	50-80%
Class switched B cells (CD19 <sup>+</sup> IgD <sup>-</sup> IgM <sup>-</sup> )	0.7%	8-31%
Naïve B cells (CD19 <sup>+</sup> CD27 <sup>-</sup> IgD <sup>+</sup> )	98.8%	42-82%
Class-switched memory B cells (CD19 <sup>+</sup> CD27 <sup>+</sup> IgD <sup>-</sup> IgM <sup>-</sup> )	0.1%	8-31%
Transitional B cells (CD21 <sup>int</sup> IgM <sup>+++</sup> CD38 <sup>++</sup> )	2.6%	0.6-3%
CD21 <sup>low</sup> cells (IgM <sup>+</sup> CD38 <sup>low</sup> CD21 <sup>low</sup> )	0.4%	0.9-7%
Anti-diphtheria antibody titres	0.003 IU/ml (very low)	
Anti-pneumococcal antibody titres	<3.3 mg/L	

Department of Pediatrics, Pediatric Allergy Immunology Unit,  
<sup>1</sup>Department of Dermatology, Postgraduate Institute of Medical Education  
 and Research, Chandigarh, India

**Address for correspondence:**

Dr. Amit Rawat,  
 Department of Pediatrics, Pediatric Allergy Immunology Unit,  
 Postgraduate Institute of Medical Education and Research,  
 Chandigarh - 160 012, India.  
 E-mail: rawatamit@yahoo.com


**References**

1. Chapel H, Lucas M, Lee M, Bjorkander J, Webster D, Grimbacher B, *et al.* Common variable immunodeficiency disorders: Division into distinct clinical phenotypes. *Blood* 2008;112:277-86.
2. Xiao X, Miao Q, Chang C, Gershwin ME, Ma X. Common variable immunodeficiency and autoimmunity - An inconvenient truth. *Autoimmun Rev* 2014;13:858-64.
3. Benoit S, Hamm H. Childhood psoriasis. *Clin Dermatol* 2007;25:555-62.
4. Dogra S, Kaur I. Childhood psoriasis. *Indian J Dermatol Venereol Leprol* 2010;76:357-65.
5. Gurmin V, Mediwake R, Fernando M, Whittaker S, Rustin MH, Beynon HL. Psoriasis: Response to high-dose intravenous immunoglobulin in three patients. *Br J Dermatol* 2002;147:554-7.
6. Paquin-Proulx D, Sandberg JK. Persistent Immune Activation in

CVID and the Role of IVIg in Its Suppression. *Front Immunol* 2014;5:637.

7. Kaveri SV, Maddur MS, Hegde P, Lacroix-Desmazes S, Bayry J. Intravenous immunoglobulins in immunodeficiencies: More than mere replacement therapy. *Clin Exp Immunol* 2011;164:2-5.

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

Access this article online	
<b>Website:</b> <a href="http://www.idoj.in">www.idoj.in</a>	<b>Quick Response Code</b> 
<b>DOI:</b> 10.4103/2229-5178.209610	

**How to cite this article:** Jindal AK, Rawat A, Sharma A, Dogra S, Suri D, Singh S. Psoriasis: An unusual autoimmune manifestation in a boy with common variable immunodeficiency. *Indian Dermatol Online J* 2017;8:292-4.

**Received:** August, 2016. **Accepted:** September, 2016.

© 2017 Indian Dermatology Online Journal | Published by Wolters Kluwer - Medknow