Intravenous Immunoglobulin for the Rescue in Refractory Cutaneous Lupus

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Dear Editor,

A 47-year-old woman who had been a known case of mucocutaneous lupus erythematosus for last 10 years presented with worsening scaly erythematous plaques over back, trunk, face and both hands and feet [Figure 1a and b]. Her skin lesions failed to respond to multiple drugs including hydroxychloroquine, azathioprine, methotrexate, dapsone, cyclophosphamide, mycophenolate mofetil, tacrolimus, and rituximab. She was receiving deflazacort 18mg and Apremilast 30mg for past four weeks without any response and had developed leukopenia (1300/mm^3), low C4, positive anti-dsDNA and anti-Ro antibodies. Intravenous immunoglobulin (IVIG, 2g/kg over five days) was started for refractory skin disease. Skin lesions started to improve along with normalization of total leucocyte count. Intravenous immunoglobulin (1g/kg) was administered monthly for 4 months and a significant improvement in palm and sole lesions were noted [Figure 1c and d]. Her joint symptoms also improved and she was able to do her regular routine works like writing with a pen. We were able to taper deflazacort to 6 mg/day with no relapse of skin lesions. Skin lesions are still in remission after 4 months of completion of her IVIG therapy without any maintenance therapy till date.

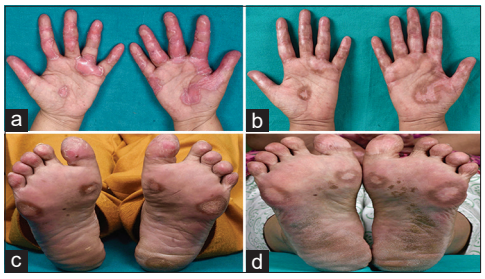

Figure 1: (a and b): Plaques with scaling over both hands and target like lesions over feet. (c and d): Improvement in lesions after intravenous immunoglobulin therapy

Cutaneous manifestations of systemic lupus erythematosus are divided into lupus erythematosus (LE) specific and LE non-specific. LE specific manifestations are divided into three different categories: acute, subacute and chronic cutaneous lupus erythematosus (CLE). Chronic CLE is further classified into discoid lupus erythematosus, LE panniculitis, chilblain LE, and LE tumidus.[1] Treatment options for cutaneous lupus includes life style modifications like smoking cessation, sun protection, and topical therapy, like steroids and calcineurin inhibitors. Moderate disease may also require oral steroids and hydroxychloroquine in addition to topical therapy. In cases of refractory and difficult to treat lesions, other systemic medications have been used such as dapsone, methotrexate, azathioprine, cyclophosphamide, thalidomide, biologicals including rituximab and intravenous immunoglobulin (IVIG).[2,3] Recent systemic review by Fairley *et al*. showed moderate evidence in support of use of any of the synthetic DMARDs and biologicals with conflicting data on IVIG.[4] IVIG therapy in cutaneous lupus has response ranging from partial to almost complete resolution of lesions, however S2k guidelines do not suggest use of IVIG for CLE due to flare of lesions documented in various case reports.[4-6] Optimal dosage, frequency and duration of IVIG treatment has not been yet well defined. It has been used mostly in case reports and series in various schedules with maximum duration of up to 6 months.[5,6] IVIG combined with corticosteroids have shown to reduce recovery time and decreased mortality in patients with StevensJohnson syndrome and toxic epidermal necrolysis.[7,8] This case highlights that IVIG can be used as safe alternative option for patients with refractory cutaneous lupus, in which drugs like DMARDs and biological did not succeed.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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