

SHORT COMMUNICATIONS

Treatment of Refractory Pruritus with Dupilumab in a Patient with Dermatomyositis

Nathan L. Bowers, MD, PhD¹ and William W. Huang MD, MPH¹

¹Department of Dermatology, Wake Forest School of Medicine, Winston-Salem, NC

Dermatomyositis (DM) is an idiopathic inflammatory myopathy characterized by progressive muscle weakness and pathognomonic skin findings. Cutaneous disease in DM is often refractory to treatment and can become the most challenging component to manage effectively. Herein, we present a case of a patient with DM who had recalcitrant cutaneous disease treated with dupilumab with remarkable response.

A 54-year-old woman presented with several month history of fatigue, extremity weakness, and pruritic rash. Her exam was significant for violaceous periorbital erythema and poikilodermatous erythema of the anterior neck, upper back, bilateral arms, and lateral thighs (Fig1). A skin biopsy was obtained demonstrating a vacuolar interface dermatitis. Creatinine kinase and aldolase were within normal range. Given clinical signs of myopathy, a MRI was ordered and demonstrated myositis. Myomarker panel was significant for MDA-5 (P140/CADM-140) antibody positivity. The patient was also diagnosed with interstitial lung disease (ILD) based on chest computed tomography (CT) results. Given her concurrent ILD she was started on mycophenolate mofetil and titrated up to 1500 mg BID as well as prednisone 1 mg/kg. She was transiently on hydroxychloroquine, but discontinued due to

lack of efficacy. Despite optimized medical therapy, she continued to have significant muscle weakness as well as cutaneous involvement. Given the recalcitrant nature of her disease she was started on rituximab 1000 mg with protocol of two infusions separated by two weeks. She had subjective and objective improvement in her muscle weakness; however, she endorsed persistent skin flaring with significant pruritus. To address her recalcitrant cutaneous disease, she was started on dupilumab with a loading dose of 600 mg and two-week maintenance doses of 300 mg. Importantly, at time of initiating dupilumab patient was on stable dose of mycophenolate mofetil 1500 mg BID and stable dose of prednisone 10 mg for several months and her most recent rituximab infusion was two months prior to first dose of dupilumab. At two-week follow-up she endorsed complete resolution of her pruritus and improvement in her skin. At six-week follow-up, she demonstrated no erythema on exam with only post-inflammatory hyperpigmentation (Fig1). She tolerated dupilumab with no adverse side effects.

Dupilumab is an Interleukin (IL)-4-receptor monoclonal antibody inhibiting signaling of IL-4 and IL-13, key drivers of type 2-driven inflammation (Th2). Interestingly, a recent study demonstrated significantly elevated

May 2021 Volume 5 Issue 3



Figure 1. (A) Poikilodermatous erythematous plaques on back and lateral leg prior to treatment with dupilumab. (B) After six weeks of treatment with dupilumab background erythema is minimal to absent with only post-inflammatory hyperpigmentation.

muscle tissue IL-4 levels in patients with DM¹, prior to this Ishii et al, found that Th2 cells predominate in peripheral blood of active DM, and that a decreased intracellular IFN- γ /IL-4 ration in CD4⁺ cells may be a useful as a marker of disease activity.² In addition to inhibiting signaling of IL-4, a key driver of Th2 mediated inflammation, dupilumab also demonstrates the ability to reduce expression of IL-31.³ A recent study highlighted the role of IL-31 in DM, demonstrating increase gene expression of IL-31 and IL-31RA in lesional skin compared to non-lesional skin and healthy controls.⁴ Collectively, this suggest a possible mechanism of action of dupilimab through its direct effect on Th2 mediated inflammation in DM as well as an indirect effect on the expression of IL-31, a proposed driver of pruritus in DM. This case highlights the use of dupilumab as a novel therapy in the treatment of cutaneous manifestations of

DM. Additional studies are needed to better assess efficacy and safety profile.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:

Nathan L. Bowers, MD, PhD
 Department of Dermatology
 Wake Forest School of Medicine.
 Medical Center Boulevard
 Winston-Salem, NC 27157-1071
 Phone: 336-716-7740
 Fax: 336-716-7732
 E-mail: nbower@wakehealth.edu

References:

1. Giris M, Durmus H, Yetimler B, Tasli H, Parman Y, Tuzun E. Elevated IL-4 and IFN-gamma Levels in Muscle Tissue of Patients with Dermatomyositis. *In Vivo*. Jul-Aug 2017;31(4):657-660. doi:10.21873/invivo.11108
2. Ishii W, Matsuda M, Shimojima Y, Itoh S, Sumida T, Ikeda S. Flow cytometric analysis of

May 2021 Volume 5 Issue 3

lymphocyte subpopulations and TH1/TH2 balance in patients with polymyositis and dermatomyositis. *Intern Med.* 2008;47(18):1593-9. doi:10.2169/internalmedicine.47.0967

3. Guttman-Yassky E, Bissonnette R, Ungar B, et al. Dupilumab progressively improves systemic and cutaneous abnormalities in patients with atopic dermatitis. *J Allergy Clin Immunol.* Jan 2019;143(1):155-172. doi:10.1016/j.jaci.2018.08.022
4. Kim HJ, Zeidi M, Bonciani D, et al. Itch in dermatomyositis: the role of increased skin interleukin-31. *Br J Dermatol.* Sep 2018;179(3):669-678. doi:10.1111/bjd.16498