Case Report of Severe Cutaneous Lupus Erythematosus Flare in a Patient Treated with Eculizumab for the Treatment of Neuromyelitis Optica Spectrum Disorder

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INTRODUCTION

Eculizumab is a long-acting humanized monoclonal antibody targeted against complement C5. By preventing the cleavage of C5 into C5a and C5b, the deployment of the terminal complement system, including the formation of the membrane attack complex (MAC), is inhibited. The FDA approved Eculizumab in 2019 for the treatment of neuromyelitis optica spectrum disorder (NMOSD) in adult patients who are anti-aquaporin-4 (AQP4) antibody positive.

CASE SUMMARY

Here we present a case of a 37 year old female patient diagnosed with NMOSD in 2018 with a serum AQP4 level of 3260.5 U/mL. She had been diagnosed with Cutaneous Lupus Erythematosus (CLE) seven years prior. CLE was well-controlled with hydroxychloroquine sulfate and prednisone.

September 2019 - Patient received her first dose of eculizumab with repeat infusions every two weeks.

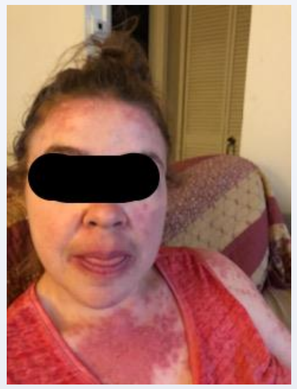
October 2019 - Patient developed multiple painful cold sores, which was not an expected side effect. Despite treatment with an anti-viral agent, valcyclovir, cold sores persisted throughout her treatment. MRI of the brain and cervical spine indicated stability of NMOSD without recurrent episodes of optic neuritis.

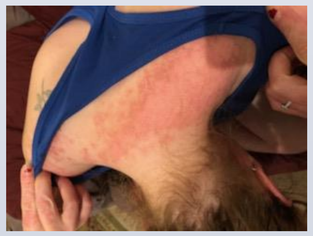
February 2020 - Patient developed new symptom of dry eyes.

April 2020 – Patient presented with a painful rash around her face, neck and chest. She was evaluated by Dermatology and Rheumatology.

June 2020 - Skin biopsy confirmed CLE flare. She was started on high dose steroids and Eculizumab was discontinued.

IMAGES OF DEMATOLOGIC CHANGES





REVIEW OF LITERATURE

SLE can be triggered by a deficiency in the classical complement pathway leading to an inability to clear antigen-antibody complexes, chromatin, or other immune aggregates.

However, this is most commonly due to deficiency in C1q, C2, or C4. Eculizumab does not target these.

The exact molecular mechanism of CLE is not fully understood. Drug Induced CLE is histopathologically identical to idiopathic CLE

Drug induced CLE has been reported in at least 38 different medications, including various antihypertensives, anticonvulsants, antifungals, TNFa inhibitors, immune checkpoint inhibitors, PPIs, and chemotherapeutics

Lowe et al noted that most of these medications can increase photosensitivity. They hypothesized that these medications may nonspecifically induce CLE skin lesions via an isomorphic response

Eculizumab does have a reported increased risk of photosensitivity

CONCLUSIONS

- We believe this to be the first reported case of CLE flare secondary to Eculizumab
- The underlying etiology is unclear, but it is possible that it is related to photosensitivity
- This could have occurred due to an isomorphic response in an individual with pre-existing CLE

REFERENCES

- Bolton, C., Chen, Y., Hawthorne, R., Schepel, I. R. M., Harriss, E., Hofmann, S. C., Ellis, S., Clarke, A., Wace, H., Martin, B., & Smith, J. (2020). Systematic Review: Monoclonal Antibody-Induced Subacute Cutaneous Lupus Erythematosus. Drugs in R and D, 20(4), 319–330. https://doi.org/10.1007/s40268-020-00320-5
- Fetter, T., & Wenzel, J. (2020). Cutaneous lupus erythematosus: The impact of self-amplifying innate and adaptive immune responses and future prospects of targeted therapies. Experimental Dermatology, 29(11), 1123–1132. https://doi.org/10.1111/exd.14146
- Giang, Jenny et al. Complement activation in inflammatory skin diseases. *Frontiers in Immunology*, April 2018 (9), 1 – 17.
- Gilkeson, G. S. MD. Complement-targeted therapies in Lupus. *Current Treatment Options in Rheumatology* (2015) 1:10-18, DOI 10.1007/s40674-014-0009-9
- Lowe, G., Henderson, C. L., Grau, R. H., Hansen, C. B., & Sontheimer, R. D. (2011). A systematic review of drug-induced subacute cutaneous lupus erythematosus. British Journal of Dermatology, 164(3), 465–472. https://doi.org/10.1111/j.1365-2133.2010.10110.x
- Panelius, J. and Meri, S. Complement system in dermatological diseases – fire under the skin. *Review in Medicine*, January 2015, DOI 10.3389/fmed.2015.00003