in adults. Biological therapy with anti-CD20 (rituximab) is an option in patient that does not respond to conventional therapy.

Methods The aim of this study is to determine the clinical and immunological response in 10 patients with cSLE that received treatment with rituximab in a third level hospital. 10 patients treated with Rituximab between November 2007 and October 2018 was included in a retrospective observational study. The response to treatment at 6 months and one year after the first infusion of Rituximab were assessed. All patients fulfilled four or more of the 1982 revised American College of Rheumatology criteria for the diagnosis of SLE (<16 years).

Results Ten cSLE patients treated with rituximab were included, all of them were female. The age at diagnosis of SLE was a mean of 15,61 years. The mean time duration of disease was 87,55 months (5255 m). 8 patients were caucasians. Rituximab was indicated in 60% of patients for class IV of lupus nephritis (LN), in 10% for class III LN, 10% for Class II LN, 10% for severe cutaneous lupus, and for severe hematological manifestations in 10% of cases (haemolytic anemia). In addition, 60% of the patients had mucocutaneous and articular manifestations. The disease activity of all patients was assessed using SELENA-SLEDAI index pre rituximab infusion, the mean was 17,31 (833). All patients had low level of complement and 90% increased anti-DNA. In 90% of cases the Rituximab was used as a rescue treatment and in a single case as a first line.

90% of patients with renal involvement were previously treated with CF iv and/or mycophenolate. In case of cutaneous involvement the previous treatment was methotrexate, azathioprine (AZA) and Dapsone and in case of hemolytic anemia the treatments was AZA.

The treatment protocol was 1 gram ×2 ×6 m in 8/10 patients, 375 mg/m^2×4 in 1/10 cases and 600 mg monthly for 5 m in the case of HAI. Five patients received more than 1 cycle. After the administration of Rituximab, the SELENA-SLEDAI activity index was 5,1. At 6 months a complete response was obtained in the case of hematological and cutaneous manifestations, in 3 cases of LN (proteinuria <0.5 g/day) and partial response was obtained in 2 cases. Data were not analysed in 2 patients (death and less than 6 months of the first dose of rituximab). Patients with partial response and lack of response achieved complete response at 12 months. 2/9 patients had side effects (Rituximab pneumonitis in 1 case and infections in 2 cases). Mortality was 10% (1/10 patients: infection and lupus activity, SLEDAI pre rituximab=33)

Conclusions In our study, although it consisted of few patients, it was objected that Rituximab therapy in patients with cSLE is effective, reduces lupus activity index, especially in cases of renal, cutaneous and hematologic involvement, that don't respond to conventional therapy. It may be considered in the future as an effective alternative treatment at first line treatment.

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147 CHARACTERIZATION OF CELL-BOUND COMPLEMENT ACTIVATION PRODUCTS ON SLE PBMCS USING MASS CYTOMETRY

Rebecca E Schriefer, Gabriel R Arguelles, Lynne M Mitchell, Stephen T Oh, John P Atkinson, Dennis E Hourcade, Alfred H Kim*. *Washington University School of Medicine*

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Background Complement plays a central role in SLE. Its activation by immune complexes drives type II and III hypersensitivity reactions and classical pathway activation, leading to inflammatory responses in target tissues. Its activation generates an array of bioactive soluble and cell-bound complement activation products (CB-CAPs). Data are lacking detailing the types and quantities of each CB-CAP on SLE immune cells. We speculate that discerning the patterns and impacts of complement signatures on SLE B cells will provide insight into the role of complement activation in the pathogenesis of SLE. To characterize the array of CB-CAPs on SLE immune cells, we created and validated a mass cytometry (MC) panel suitable for detecting over 20 CB-CAPs and complement receptors, then examined PBMCs from paired flare and inactive samples from patients with SLE.

Methods Adults with ACR- or SLICC-classified SLE (n=5) were enrolled and consented for PBMC collection at Washington University School of Medicine. Paired samples were obtained, one during a flare and one after resolution of the flare. Frozen ficoll gradient isolated buffy coat PBMCs were thawed counted, filtered, and treated with cisplatin to confirm viability. Prepared cells were stained with metal-conjugated antibodies to measure surface complement proteins and markers identifying immune cell subset. Stained cells were treated with an Iridium intercalator stain, recounted, washed in Milli-Q water to remove salts, resuspended with a solution of bead standards to facilitate data normalization, and subjected to single cell MC. Sample to sample consistency is optimized by simultaneously examining multiple cell preparations barcoded with palladium metals. Data analysis was performed with Cytobank. Distinct cell sub-populations were identified and organized in a hierarchal fashion.

Results We found the highest frequency of C4d deposition (indicative of classical pathway activation) on B cells compared to T cells and monocytes during SLE flares. Surprising, only B cells had C3d, C5 (common pathway), and Bb (alternative pathway) CAPs with none observed on T cells or monocytes suggesting that incomplete classical pathway activation occurred on these cells. We also found that different B cell subsets differentially activated complement during flares. Transitional B cells universally had C4d and C5 but surprisingly had little C3d, despite the presence of Bb. Mature B cells, particularly the mature anergic subset, and memory B cells had C4d and C3d but little C5. Plasma cells/plasmablasts were devoid of any CB-CAPs. Finally, during disease inactivity, very low levels of all CB-CAPs were observed on all PBMCs.

Conclusions We found a high level of CB-CAP deposition in immune cells obtained from subjects with a SLE flare, which was absent during disease inactivity. The types of CB-CAPs on PBMCs were not uniform, and our data suggest that certain immune cells appear regulate complement activation while others do not. The reasons for this are unclear, but it potentially opens up a previously undescribed heterogeneity in SLE. These pilot data demonstrate the feasibility of the MC complement panel on human samples and the power of this approach in discovering novel mechanisms of complement activation and regulation.

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