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NEUROMYELITIS OPTICA OVERLAPS FREQUENTLY WITH SYSTEMIC RHEUMATIC DISEASES IN AFRICAN-AMERICANS: EXPERIENCE AT A LARGE US ACADEMIC MEDICAL CENTER

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Background: Neuromyelitis optica (NMO) and neuromyelitis optica spectrum disorders (NMOSD) are immune-mediated demyelinating disorders of the central nervous system, primarily characterized by optic neuritis and longitudinal extensive transverse myelitis. Antibodies to aquaporin-4 (AQP4-IgG) are highly specific markers of NMOSD. The coexistence of NMOSD with systemic rheumatologic autoimmune disease has been well documented, especially systemic lupus erythematosus (SLE) and Sjogren syndrome (SS) [1]. Several recent studies have demonstrated that NMOSD may disproportionately affect black patients [2,3].

Objectives: Our objective was to characterize African-Americans diagnosed with NMO at our academic medical center, and identify any associated systemic rheumatologic autoimmune diseases.

Methods: We conducted a retrospective chart review of adult African-American patients diagnosed with NMO at a single academic medical center between 2011 and 2017. The charts reviewed were identified using ICD codes for NMO, transverse myelitis and optic neuritis. The diagnosis of NMO was then confirmed as per the 2015 International Consensus Diagnostic Criteria. The diagnosis of SLE and SS were also confirmed based on existing diagnostic criteria.

Results: Of the 60 charts reviewed, 25 patients (41.7%) met criteria for NMO, and of those, 21 patients (84%) were African-American. Of the 21 African-American NMO patients, 89.5% (n=19) were AQP4-IgG positive, 95.2% were women, and 38.1% had an associated systemic rheumatologic autoimmune disease (SLE 62.5% and primary SS 37.5%). In 60% of the NMO patients with SLE, the diagnosis of NMO preceded the SLE diagnosis. Conversely, in 100% of the NMO patients with SS, the diagnosis of NMO either followed, or was made simultaneous to, the SS diagnosis. Notably, 80% of the SLE patients had primarily hematologic manifestations. ANA was positive in 78.6% of the African-American patients in whom ANA testing was performed (n=14), with anti-SSA (64.3%, n=14) and anti-SSB (50%, n=11) the next most prevalent autoantibodies. Mean age of NMO onset was 46.7 years (SD 13.6). Transverse

myelitis was the most common presenting manifestation (71.4%), followed by optic neuritis (61.9%).

Conclusion: Our population had a higher mean age of onset (46.7 vs 33 years) compared with other cohorts of seropositive NMO African-Americans [2]. Additionally, African-Americans with NMO at our institution had a higher female:male distribution than has been documented[3]. Not surprisingly, SLE was the most common associated systemic rheumatic disease in our cohort, however the frequency of SLE and ANA positivity in our NMO cohort was higher than has been reported (23.8% and 78.6%, respectively) [4]. Additional larger studies are needed to further explore how NMOSD in African-Americans may differ from other populations.

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EFFICACY AND SAFETY OF RITUXIMAB FOR INDUCTION OF REMISSION AND MAINTENANCE OF IGG4-RELATED DISEASE: EXPERIENCE FROM AN ITALIAN NATIONAL REFERRAL CENTRE

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Background: IgG4-related disease(IgG4-RD) is a systemic fibro-inflammatory disease. Treatment is based on corticosteroids(CS) but up to 50% of patients may relapse. Rituximab(RTX) has emerged as an effective treatment option[1].

Objectives: To assess the free relapse rate(FRR) in patients receiving or not maintenance RTX therapy. Secondary outcomes: time to relapse, incidence of adverse events and differences in FRR between Group 2a and 2b patients.

Patient #	Age (years)	Sex	N° of Organs involved	IgG4-RD RI	PGA	ESR (mm/1h)	CRP (mg/L)	IgG4 (mg/L)	Concomitant DMARD therapies	Previous DMARD therapies	Outcomes after RTX induction	Relapses (time to relapse)	N° of RTX infusions	RTX Adverse Events	Follow-up (months)
Group 1															
1	29	F	3(Pm,Ns, Ph)	12	90	38	25	1001	No	CTX	PR	No	1	No	32
2	47	M	2(Ly,Sg)	6	50	8	1,2	4420	No	MTX	DR	No	1	No	30
3	70	F	1(Ao)	6	70	26	3,7	850	MTX	No	DR	Yes(7)	2	No	26
4	49	M	2(Bd,Pa)	6	60	39	1,9	3360	No	No	DR	Yes(7)	2	No	26
5	52	M	4(Pm,Si,Or, Pa)	15	90	7	21	1700	No	MTX	DR	Yes(12)	2	No	18
6	52	M	3(Ly,Sg,Lu)	12	70	8	1,9	7904	MTX	No	CR	Yes(11)	2	No	18
7	80	M	3(Pa,Ly,Bd)	9	60	22	6	3620	No	MTX	DR	Yes(9)	2	No	18
Group 2															
1	53	M	5(Ly,Si,Lg, Pa,Sg)	12	80	7	0,1	5713	No	AZA	CR	0	3	No	18
2	49	F	4(Ly,Lg,Or, Ph)	15	90	22	0,3	13800	No	No	CR	0	4	No	20
3	16	F	3(Si,Or,Ly)	9	60	6	0,2	630	No	No	DR	0	7	No	48
4	28	F	4(Hy,Pm, Or,Ns)	15	90	65	5,7	2990	No	CTX	CR	0	3	Yes (Asthma)	18
5	73	F	3(Or,Ly,Pa)	12	70	17	1,2	21800	MTX	AZA	CR	0	5	Yes(Rash)	30
6	51	F	3(Lg,Or,Lu)	12	70	16	6,9	2530	No	No	DR	0	4	No	18
7	76	M	1(Bd)	6	70	31	36	470	No	No	DR	0	3	No	21