Limited NMOSD phenotypes in patients with low anti-AQP4+ titers

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INTRODUCTION: Neuromyelitis Optica (NMO) is an inflammatory immune-mediated disease of the CNS, characterized by severe attacks of optic neuritis (ON), transverse myelitis (TM) and area postrema syndrome (APS). The disease may present with more limited forms, called Neuromyelitis Optica Spectrum Disease (NMOSD). The recent diagnostic criteria from the International Panel (Wingerchuk, 2015) requires one typical NMOSD attack in patients who are positive for aquaporin-4 antibodies (anti-AQP4+). **OBJECTIVE:** To compare the patients with anti-AQP4 titers above and below 1:128x. **METHODS:** Anti-AQP4 antibodies were detected using a live cell-based assay using M23-AQP4 transfected HEK293 cells. Anti-AQP4 titers were calculated using end-up double-dilutions by two blind raters. **RESULTS:** Thirteen patients were positive for anti-AQP4, 10 were female and 3 were male. The median onset age was 51 years (10 – 78 years). Among the 7 patients with anti-AQP4 titers above 1:128x, the median age was 51 years and all were female. Five out of 7 (71%) of the patients had already 2 or more attacks at the time of anti-AQP4 testing and with attacks not limited to one localization. In contrast, 6 patients had anti-AQP4 titers below 1:128, with a median age of 55 years, and half were males. Five out of 6 (83%) had only one attack at the time of anti-AQP4 testing, and 4 patients had only a single attack of myelitis. **CONCLUSIONS:** In this preliminary study, we observed that the patients with low anti-AQP4 titers had some differences on the gender predominance and they may have a more limited clinical phenotype, such as a single myelitis attack.