Autoantibodies in Myasthenia Gravis

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Acquired myasthenia gravis (MG) is a well-established organ-specific, autoantibody-mediated disease caused by circulating antibodies directed against the proteins at the neuromuscular junction (NMJ). Antibodies to the muscle nicotinic acetylcholine receptor (AChR) are detected in approximately 85% of generalized MG. In the remaining AChR seronegative MG patients, up to 50% harbor antibodies against muscle-specific tyrosine kinase (MuSK), an enzyme critical for NMJ formation and agrin-induced AChR clustering. In some of the double seronegative cases, antibodies against AChR can only be detected by binding of clustered AChRs in a cell-based assay. Recently, distinct antigenic targets have been identified which include LRP4 (Lipoprotein receptor-related protein 4) and agrin. MG is also associated with other antibodies that recognize skeletal muscle proteins, which may be useful in identifying the subtypes of MG, although their role in disease pathogenesis is unclear. There is a variable relationship between the levels of serum anti-AChR, anti-MuSK or other MG-related muscle autoantibodies to disease severity and response to treatment in MG. In this talk I will review the autoantibodies that have been associated with MG and discuss their role in MG pathology, diagnosis and their potential utility as therapeutic biomarkers.