

#1871: Neuromyotonia in thymoma-associated myasthenia gravis: a clinico-serological study

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Type

Abstract

Topic

Neuroimmunology

Category

Poster or Oral

Background and aims

Acquired Neuromyotonia(NMT) is an autoimmune condition frequently associated with anti-contactin-associated-protein-like-2(Caspr2) antibodies. NMT can occur as a paraneoplastic disorder in patients with thymoma, alone or in combination with Myasthenia Gravis (MG). Recently, antibodies against netrin-1-receptors(DCC and UNC5A) have been reported as predictors of thymoma in 6/9 patients with concomitant NMT and MG. We aimed to clinically characterize a large cohort of patients with thymoma-associated MG, and to explore serological correlation of NMT symptoms.

Methods

268 consecutive patients with thymoma-associated MG were retrospectively collected. NMT was defined as muscle twitching/cramps in at least 2 skeletal districts. Patients with NMT(23) were screened for anti-neuronal antibodies by immunohistochemistry on rat brain and cell based assay.

Results

23/268 patients developed NMT symptoms (muscle twitching, 3; cramps, 3, or both, 17). Overall, 33/268 patients with thymoma had a tumor recurrence, which was more frequent in those with(8/23) vs those without NMT(25/245, p=0.003). NMT onset preceded the tumor recurrence in 5/6 patients. In univariate analysis predictors of thymoma recurrence were younger age at thymectomy(odds ratio-OR:0.95, confidence interval-CI:0.93-0.97), Masaoka staging(OR:10.73, CI:2.38-48.36) and NMT(OR:4.69, CI:1.76-12.46). 6 patients with NMT had anti-neuronal antibodies(patient#1: Caspr2; patient#2: AMPAR; patient#3: DCC; patient#4: LGI1; patient#5: Caspr2+LGI1+DCC+UNC5A; patient#6: Caspr2+LGI1+DCC). Thymoma recurrence was found less frequently in negative(3/17) vs positive patients with NMT(4/6, #1, #2, #5 and #6; p=0.045).

Conclusion

The occurrence of NMT symptoms in patients with thymoma-associated MG can predict tumor recurrence, and warrants a closer oncologic follow-up. Anti-neuronal surface autoantibodies may be useful to further stratify the recurrence risk.

Disclosure

This work was funded by the 'Ricerca finalizzata ministeriale 2015-2017' provided by the Italian ministry of health

Affirmations

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