PARANEOPLASTIC MANIFESTATIONS OF THYMOMA: A CASE REPORT

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INTRODUCTION:

Myasthenia gravis (MG) is the most common autoimmune disease associated with thymoma; however other neurological conditions can be found in thymomatous patients, including some that affect at the same time the central nervous system (CNS), the peripheral nervous system (PNS) and the autonomic nervous system (ANS).

We describe the case of a MG patient with thymoma who presented with generalized seizures in the setting of recurrent thymoma with antibodies to Caspr2 (contactin-2 associated protein).

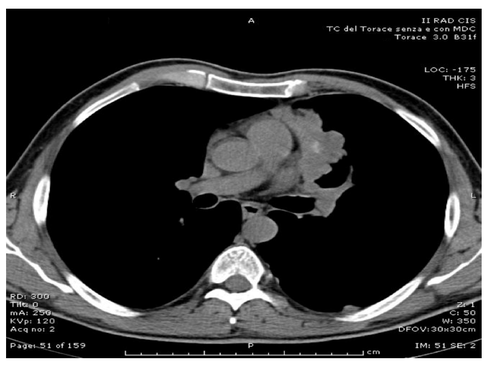

Fig. 1: CT scan with evidence of an infiltrative thymoma

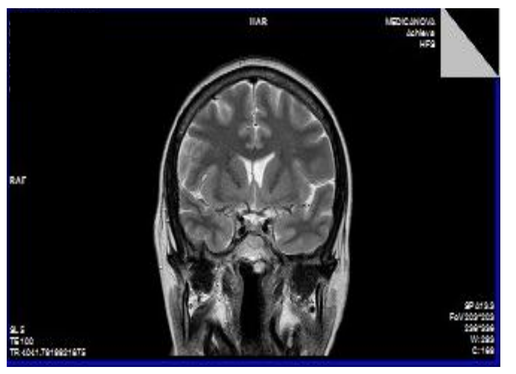

Fig. 2: Brain MRI (T2W_TSE) was normal

CASE REPORT:

- ❖In 2007 a forty years old female with a diagnosis of generalized Myasthenia Gravis (MG) with antibodies to acetylcholine receptors (AChR-Ab) underwent thymectomy for a B2 thymoma, followed by radiotherapy
- ❖In 2009 she came to our attention and started therapy with prednisolone and pyridostigmine bromide.
- ❖In 2010 she had a pleural recurrence of thymoma surgically treated.
- ❖In February 2011 she started suffering from cramps and involuntary muscle twitching that steadily got worse despite the interruption of the cholinesterase inhibitor.
- ❖In April 2013 she developed sensation of tremor around trunk in the inframammary region, eyelid myokimia and limb muscle pain, hyperhidrosis, insomnia and cognitive impairment.
- ❖In May 2013 and in March 2014 she had two episodes of generalized seizures; EEG showed a peculiar pattern, comprising lateralized right fronto-temporal spikes and generalized sharp waves. An antiepileptic therapy (Levetiracetam 500 mg BD) was attempted.
- ❖In February 2014 she underwent radiotherapy for another recurrence of thymoma. Moderate-dose steroid therapy has been maintained to avoid other recurrences of thymoma.
- ❖In November 2015 another generalized seizure occurred; brain MRI was normal except for a small focus of gliosis.

Clinical suspicion of another paraneoplastic syndrome associated with thymoma was confirmed by the serological detection of antibodies to **Caspr2** (fixed cell-based assay). Peripheral and autonomic symptoms greatly improved with steroid treatment; however, episodes of disorientation and dizziness without loss of consciousness still occur.

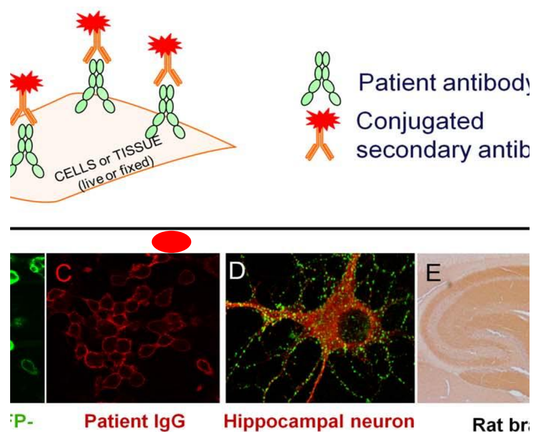

Figure 3: Depiction of the voltage-gated potassium channel (VGKC) complex. Antibodies bind the extracellular domains of LGI1 (in patients with limbic encephalitis, facio- brachial dystonic seizures [FBDS], and Morvan syndrome [MoS]) and CASPR2 (in patients with MoS more frequently than in neuromyotonia [NMT] or LE). (fROM)

CONCLUSION:

Caspr2 antibodies are an infrequent accompaniment to thymoma, however they should be considered as the possible cause of epilepsy in patients with a thymoma particularly active. CNS and PNS manifestations strongly improved with corticosteroid treatment which represents an useful tool in the long-term control of autoimmune reactions triggered by thymic pathology although relapses can occur. interestingly, EEG showed a peculiar pattern that has been already described in patients with autoimmune encephalitis.

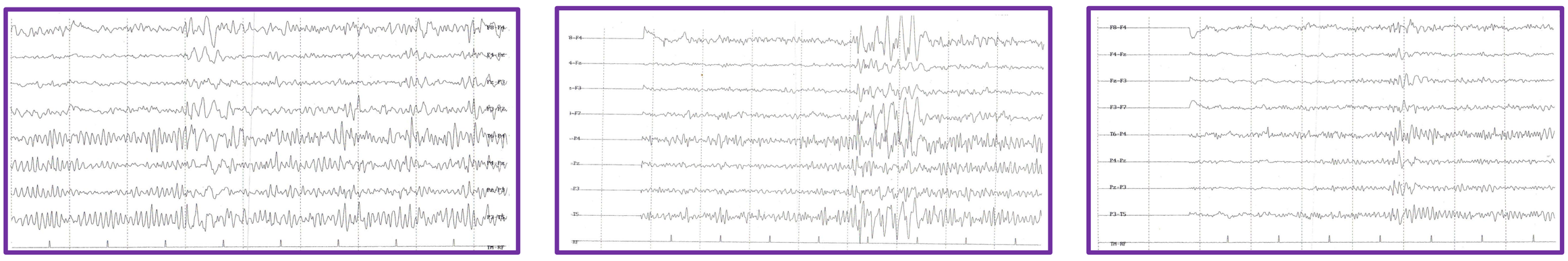

Figure 4: Electroencephalogram showed lateralized right fronto-temporal spikes and interictal generalized sharp waves

References:

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