Insomnia as the primary complaint for LGI1 and CASPR2 autoantibodies, neuromyotonia and small fiber neuropathy in a patient with oncocytic Schneiderian papilloma.

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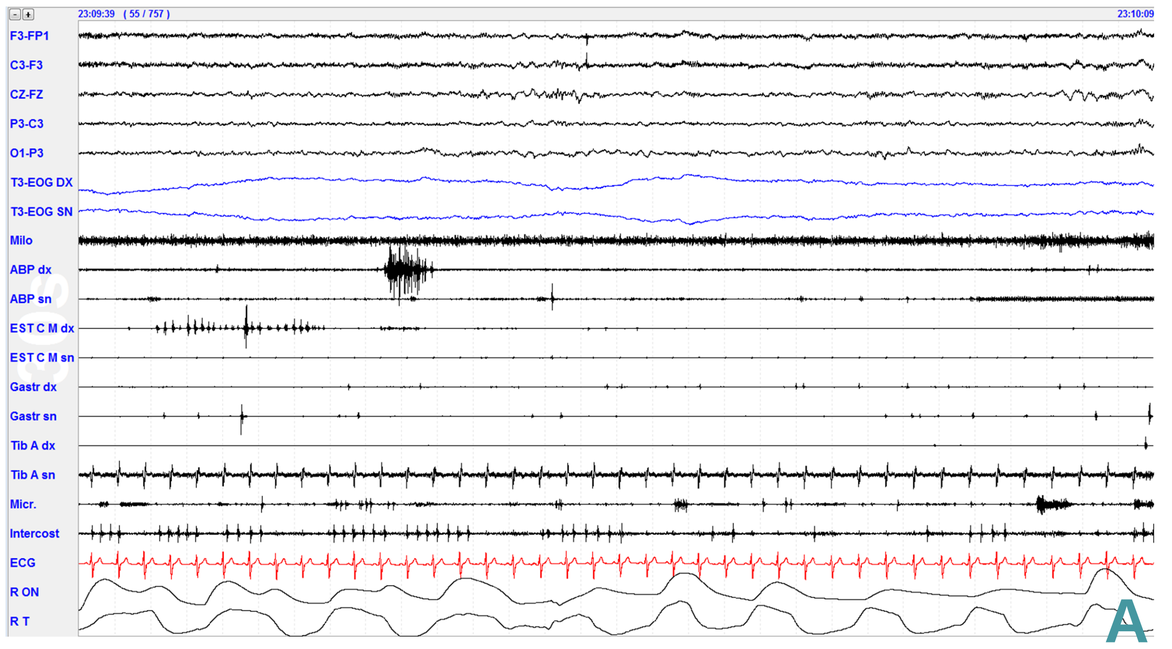
We report a 60 years old male, ex-smoker, treated with ventilation and statins for obstructive sleep apnea and dyslipidemia, who complained for the acute onset of insomnia related to a painful stiffness firstly of the legs and then extending to the arms over a few weeks. He also reported urge to move limbs while resting at evening-night period not modified by movement, distal paresthesiae with hypoesthesia. Pramipexole and pregabalin didn't relief symptoms, that gradually worsened with subjective walking impairment due to pain and heaviness sensations in lower limbs; insomnia partly responded to clonazepam. The patient also reported occasional lower limbs cramps and remarkable weight loss (15 Kg in 2 months).

Materials and method: Our workup included nocturnal video-polysomnography (V-PSG), actigraphy, electromyography (EMG), brain MRI, whole body 18FDG TC/PET, skin biopsy and voltage-gated potassium channel (VGKC) complex autoantibody assay.

Results: Neurological examination revealed diffused fasciculations/myokymia, distal tactile and thermoalgesic hypoestesia; Epworth sleepiness scale scored 5/24 (normal). V-PSG showed an alteration of sleep macrostructure with reduced sleep efficiency (SE 23%), continuous muscular activity (Fig. A), a brief enacted behavior at sleep onset, and absence of REM sleep atonia. EMG showed after-discharges at nerve conduction study (Fig. B) and spontaneous activity with doublet or multiplet single motor unit discharges. Brain MRI was normal except for right maxillary sinus thickening, that showed intense hyperfixation at 18FDG TC/PET. Skin biopsy revealed autonomic and somatic small fiber neuropathy (Fig. C). VGKC complex autoantibody assay resulted positive for LGI1 and CASPR2 autoantibodies. Accordingly, the patient underwent high dose steroids and three sessions of plasma exchange, followed by 5 monthly sessions. Patient's symptoms dramatically improved and he regained weight. Maxillary sinus biopsy revealed the presence of oncocytic Schneiderian papilloma, and its removal was recently performed. Control V-PSG and EMG at 5 months showed normal sleep macrostructure (SE 86%) with physiological REM sleep atonia (Fig. D), and the disappearance of spontaneous EMG activity. Long-time actigraphy monitoring showed a progressive increase in nocturnal sleep with concurrent reduction in daytime sleep (Fig. E) .



Figures legend:

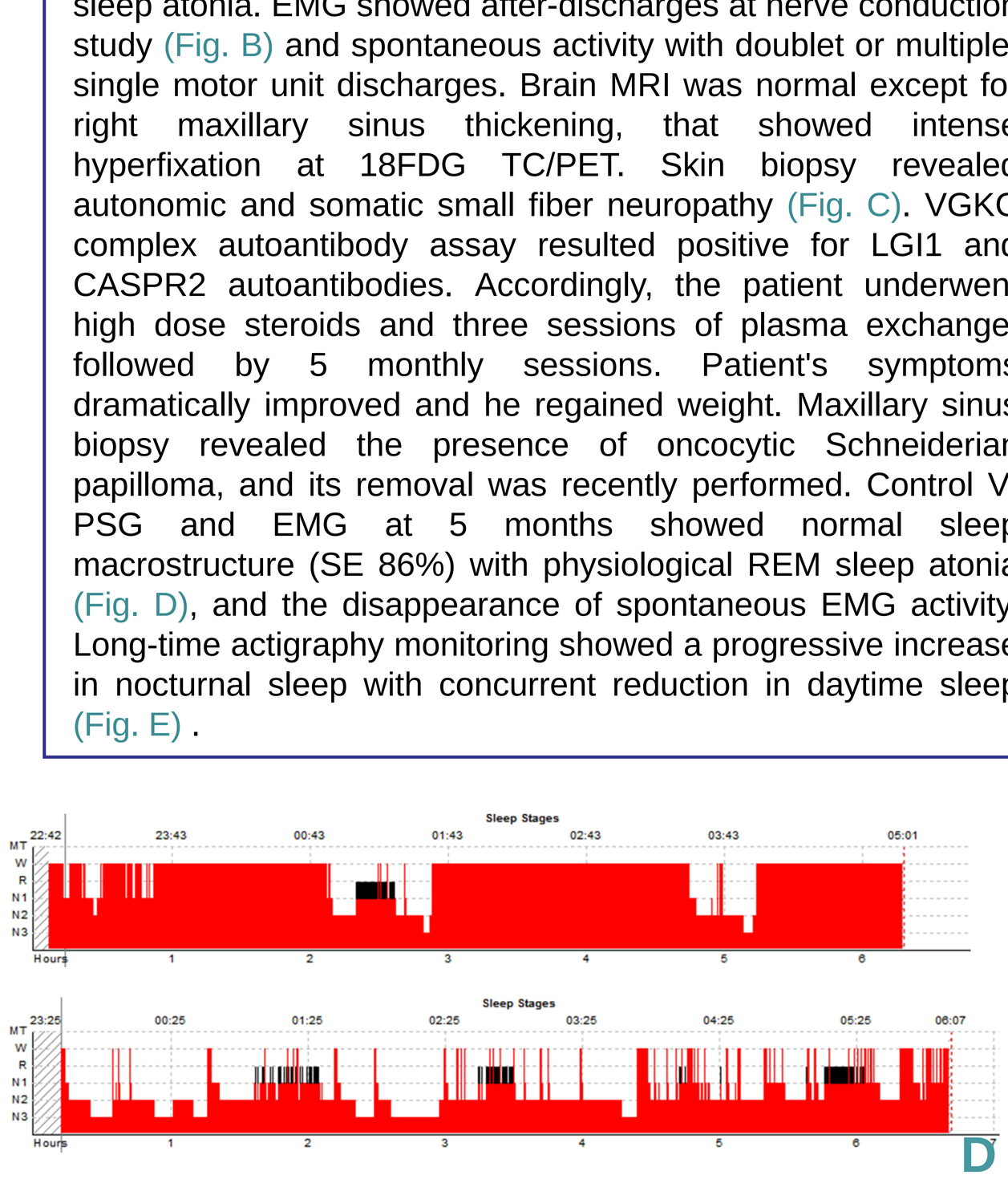
A. *Basal PSG: muscular activity in sleep*

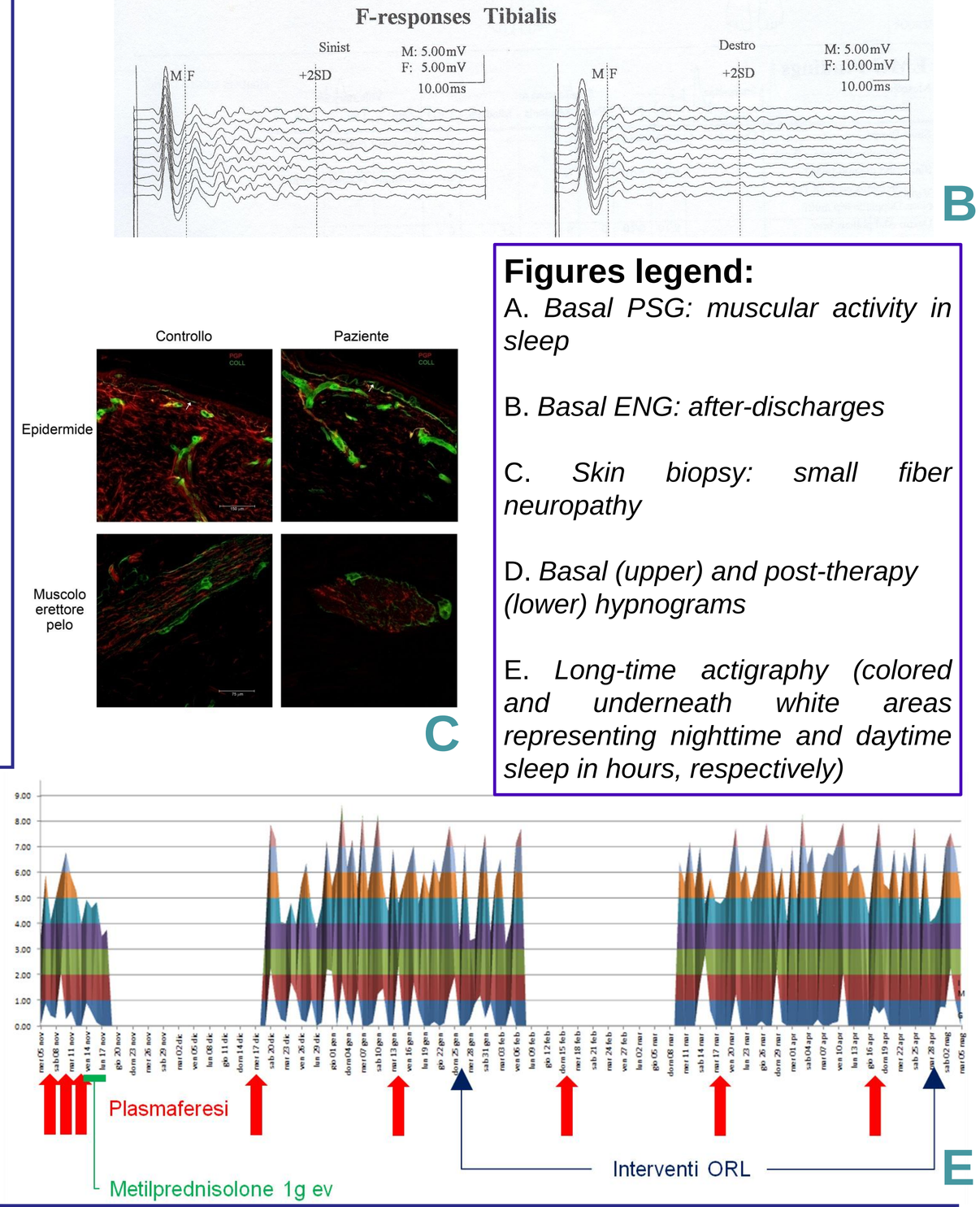
B. *Basal ENG: after-discharges*

C. *Skin biopsy: small fiber neuropathy*

D. *Basal (upper) and post-therapy (lower) hypnograms*

E. *Long-time actigraphy (colored and underneath white areas representing nighttime and daytime sleep in hours, respectively)*





Discussion: Our patient suffered insomnia, neuromyotonia, and small fiber neuropathy associated to LGI1 & CASPR2 autoantibodies and oncocytic Schneiderian papilloma, a benign neoplasm never described in association to VGKC complex autoantibodies. CNS involvement was represented mainly by a quantitative sleep disruption instead of the dramatic features of agrypnia excitata. Patients with VGKC complex autoantibodies may show variable degree of central and peripheral neurological involvement. Prompt diagnosis and treatment may prevent from widespread central involvement.

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10-13 OTTOBRE 2015 – GENOVA

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