Proprioceptive Stimuli as a New Type of Trigger for Epilepsy in Stiff Person Syndrome

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A healthy 74-year-old woman was diagnosed with Stiff-Person (SP) syndrome (encephalopathy and stiff leg) with amphiphysin autoantibodies and T1N1M0 breast cancer (HER2+,PR-,ER-). She received immunotherapy (2 high-dose steroid cycles and 7 plasmapheresis sessions), followed by right mastectomy, axillary chain lymphadenectomy and adjuvant chemotherapy (letrozole, trastuzumab, cyclophosphamide), with poor clinical response (ECOG 4). After discharge, she presented to the emergency room with seizures induced by knee flexion of the stiff leg (video 1). EEG showed epileptic seizures initiating in the left temporal lobe (image 1). Reduced presynaptic GABAergic inhibition could be the underlying mechanism of the core signs of amphiphysin-SP, which might explain the presence of epilepsy in these patients(1). However, no reflex proprioceptive seizures had been previously reported in SP.

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References

Figure 1. Electroencephalogram Shows Epileptiform Discharges Initiating in Left Temporal Region

[For comp: no legend]

Video 1.
Passive knee flexion of the left leg produced clonic movements that started in the contralateral limb, and quickly evolved to bilateral tonic-clonic activity. No other triggers were found. The patient remained aware during the whole seizure. After 3mg of intravenous diazepam, epileptic activity ceased.
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