Demyelinating polyneuropathy in a case of anti-LGI1 encephalitis: is there a link?

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BACKGROUND

Limbic encephalitis (LE) associated with antibodies against leucine-rich glioma inactivated 1 (anti-LGI1), previously known as LE with voltage-gated potassium channel (VGKC) antibodies, usually occurs in middle aged males, manifesting, similarly to other types of LE, with cognitive impairment, psychiatric symptoms and seizures. However, some specific features can lead to the diagnoses, such as sleep behavior disorders, hyponatriemia and facio-brachial dystonic seizures (FBDS). Usually the disease is not tumor-related and responds to immunotherapy.

CASE REPORT

A 78 year old man was observed for the acute onset, twenty days before, of sudden jerky movements, lasting a few seconds and affecting the left or the right side of his body with several episodes per day. Specifically, they involved the face causing grimacing and interfering with his speech, and the upper and lower limb causing frequent falls. Moreover the patient complained of insomnia and visual hallucinations. His medical history included glaucoma and type 2 diabetes. Neurological examination was normal except from the involuntary movements. Brain computed tomography (CT) scan and gadolinium magnetic resonance imaging were negative. Prolonged electroencephalogram (EEG) monitoring recorded several episodes without any EEG correlate. Electromyography (EMG) showed a mainly demyelinating sensory-motor polyneuropathy not having the characteristics of a diabetic neuropathy (Tab. 1). Routine blood tests, reumathological and coagulation assessment were normal except for hypopiastrinemia. Serum was tested for anti-GM1, -NMDA, -VGKC and onconeural antibodies showing the presence of anti-LGI1 antibodies. The patient was therefore treated with intravenous (IV) methilprednisolone 1 g daily for 5 days followed by oral prednisone 75 mg daily. FBDS almost immediately decreased in frequency and disappeared at day 3 of the IV therapy. Neuropsycological assessment revealed a normal global pattern. Total-body CT scan excluded the presence of tumors. Six weeks after the end of the IV immunotherapy, the patient was still free from FBDS and hallucinations. The EMG performed 2 months after the onset showed an improvement of conduction velocities, distal motor latencies and F waves (Tab. 1).

	ENG at onset				ENG after 2 months			
Motor nerves	CV (m/s)	Amp (mV)	DML (ms)	F (ms)	CV (m/s)	Amp (mV)	DML (ms)	F (ms)
Deep Peroneal [R]	35.3	3	4.53 (9 cm)	a	43.5	2.2	4.05 (9 cm)	60.2 (3/10)
Tibial [R/L]	33.3/37.1	0.97/3.5	8.7/7.2 (12 cm)	a/67	39.4/40.3	0.92/3.3	5.8/4.87 (12 cm)	a/64
Sensory nerves	CV (m/s)	Amp (μV)			CV (m/s)	Amp (μV)		
Sural [R/L]	35.6/34	3.8/1.96			44.2/42.6	3.2/1.9		

Table 1. Patient's ENG parameters before and after treatment

CONCLUSIONS

Since anti-LGI1 encephalitis is a recently identified syndrome, the amount of data regarding its clinical features is small. According to the available literature it is characterized by memory loss, confusion, FBDS, sleep disorders and hyponatriemia. To date polyneuropathy is not considered part of the disease. However, the finding in our patient of a mainly demyelinating sensory-motor polyneuropathy, in the absence of other possible causes, could suggest the involvement, not only of the central nervous system, but also of peripheral nerves in anti-LGI1 encephalitis.

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