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FACIOBRACHIO-CRURAL DYSTONIC SEIZURES IN LGI1 LIMBIC ENCEPHALITIS: A TREATABLE CAUSE OF FALLS

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Faciobrachial dystonic seizures (FBDS) are closely associated with antibodies (Ab) to the leucine-rich glioma inactivated-1 protein (LGI1) component of the voltage-gated potassium channel (VGKC) complex. They precede or indicate the onset of limbic encephalitis (LE).¹ Immunotherapy can prevent the development of LE and cognitive impairment.^{1,2}

FBDS are very brief, very frequent unilateral dystonic seizure-like events that always affect the arm and commonly affect the ipsilateral face.¹ Involvement of the leg is rare, and little has been reported on so-called facio-brachio-crural dystonic seizures (FBCDS), which are associated with falls as well.¹⁻³ Hence, FBCDS are a rare but treatable differential diagnosis of recurrent falls.

Case report. An 83-year-old woman presented with sudden falls without loss of consciousness. The falls first occurred 4 weeks prior to admission to our ward. She reported knee buckling in the right leg while standing or walking that led to falls to her right side. Previous medical history included arterial hypertension and type 2 diabetes mellitus. Hence, extensive workup, including the implantation of an event recorder, was performed to exclude cardiac and cerebrovascular causes for the falls. Following that, anticonvulsant treatment with levetiracetam (LEV) was started and she was admitted to our ward for long-term video-EEG monitoring. EEG recorded frequent unilateral, alternating in side (more right than left), very brief dystonic seizures up to 5 times an hour involving the face, arm, trunk, and leg (video at Neurology.org/nn). They occurred during the day as well as at night during sleep and led to arousal. Ictal EEG abnormalities were not observed (only ipsilateral muscle artifacts). Interictal EEG showed right frontotemporal slowing with periodic enclosed 1–1.5 second sharp and sharp-slow waves in Fp2, F4, F8, T2, and T4 for up to 20 seconds. Anti-LGI1 Ab were identified in serum (1:2,000) and CSF (1:4) (VGKC-Ab radioimmunoassay kit assay and cell-based assays [Eurotide; Euroimmun, Luebeck, Germany]). The routine CSF examination

(cell counts, glucose, and protein) was unremarkable. Further investigations demonstrated moderate hyponatremia (127 mmol/L). Brain MRI showed high T2/fluid-attenuated inversion recovery (FLAIR) signal intensity in the right amygdala. Formal neuropsychological testing was normal. Pulse methylprednisolone treatment (1,000 mg IV/day) was administered for 5 days followed by oral prednisolone (100 mg/day) tapered to a maintenance dose of 5 mg/day. Ten days after the beginning of the steroid treatment, the FBCDS frequency was significantly reduced. In contrast, LEV (max. 3,000 mg/day), which was started 2 weeks before the steroid treatment, was ineffective.

After discharge from the hospital, no further falls occurred. On follow-up at 3 months, no more FBCDS were recorded in the video-EEG. Interictal EEG showed isolated bitemporal theta waves and infrequent temporal sharp waves (right > left). Overall, the interictal changes declined in frequency. The T2/FLAIR signal intensity in the right amygdala had waned. Memory deficits were not detected on follow-up neuropsychology testing. The LGI1-Ab titer had a decrease in both serum (1:16) and CSF (negative). There was no hyponatremia.

Discussion. Falls are a very common problem in older patients.⁴ The major causes of falls include accidents and environmental hazards with individual risk factors.⁴ In second and third place are gait/balance disorders and dizziness/vertigo.⁴ In up to 21% of cases the etiology of falls remains unknown.⁴ Especially recurrent falls of unknown cause demand attention due to rare differential diagnoses. Video-EEG monitoring can be helpful in these cases and can contribute to diagnosis, as in our case, although ictal and interictal EEG changes were demonstrated in a minority of patients with FBDS.^{1,2} Ictal EEG abnormalities were described during longer clinical events.² In our patient with very brief FBCDS, only interictal EEG changes were observed.

Based on our case report, FBCDS are one of the rare but treatable differential diagnoses of falls without loss of consciousness. The timely detection of FBCDS is crucial, because immunotherapy with corticosteroids can lead to freedom from

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seizures and falls and prevent the development of LE with cognitive impairment.^{1,2} Seizure reduction and normal serum sodium levels correlated in our case with the decrease in LGI1-Ab titer and activity of the LE.¹ Also, the T2/FLAIR signal intensity of the right amygdala and the EEG changes regressed after cortisone therapy corresponding to the fall in LGI1-Ab titer. Anticonvulsants did not show any effect on FBCDS frequency in our patient, as demonstrated for FBDS previously.^{1,2}

In conclusion, FBCDS are a rare but important differential diagnosis for recurrent falls in older patients. Knee buckling preceding falls should prompt further investigations with video-EEG monitoring. Immunosuppressive treatment can reduce falls significantly and prevent the progression of LE with cognitive impairment.

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