Valproate-induced Reversible Hemichoreoathetosis in a Patient With Rasmussen Encephalitis

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A 24-year-old woman with Rasmussen encephalitis diagnosed at age 6 underwent investigation of involuntary movements. She developed continuous left-sided hemichoreoathetosis at age 11, after addition of valproic acid (VPA) to carbamazepine and phenobarbital therapy. Admission workup was unremarkable, except for toxic VPA levels (129 mg/L). Cessation of movements was observed the day after VPA was discontinued (Video).

In this case, pre-existing encephalic damage (Figure) may have shifted the excitation-inhibition balance in basal ganglia-thalamocortical circuits towards motor cortex activation, which was then enhanced by toxic VPA levels (via glutamate acid decarboxylase inhibition, leading to low GABA levels), therefore generating contralateral hemichorea\(^1\,2\).

WNL-2022-200548_vid1 -- http://links.lww.com/WNL/B887

References:


Video: Valproate-induced reversible hemichoreoathetosis

Woman with history of Rasmussen encephalitis developed left-sided hemichoreoathetosis provoked by valproic acid. The patient is seen lying on her back, sitting and walking. Note impressive cessation of choreiform movements after valproic acid withdrawal, even after thirteen years of continuous chorea.

Figure: Brain MRI findings at age 24

Axial and coronal FLAIR MRI sequences (respectively, A and B) show right-sided frontal lobe atrophy, subcortical hyperintensities, atrophy of the head of caudate and lentiform nuclei as well as ex-vacuo ipsilateral anterior horn dilatation (arrows). No restricted diffusion or gadolinium enhancement was observed.
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