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# Adult-onset Alexander disease: could facial myokymia be a symptom?

## Doença de Alexander de início no adulto: mioquimia facial pode ser sintoma?

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A 24-year-old man presented with progressive cerebellar syndrome, which after six months evolved with dysphagia, dysphonia, corticospinal tract involvement and facial myokymia. Electromyography showed myokymic discharges (Figure). Brain and spinal cord magnetic resonance imaging (MRI) were suggestive of Alexander disease (AD) (Figure). Genetic analysis in *GFAP* gene confirmed AD (Asp360Asn).

Adult-onset AD diagnoses can be strongly suggested by MRI findings[1,2,3,4]. Electromyography can usually revealed mild chronic neurogenic abnormalities or myoclonus rhythm in some patients[1]. Myokymia has not been reported associated to AD[1,4]. However, facial myokymia although typically a benign condition, may reflect the brainstem evolvement of the AD. Therefore, our case includes myokymia in

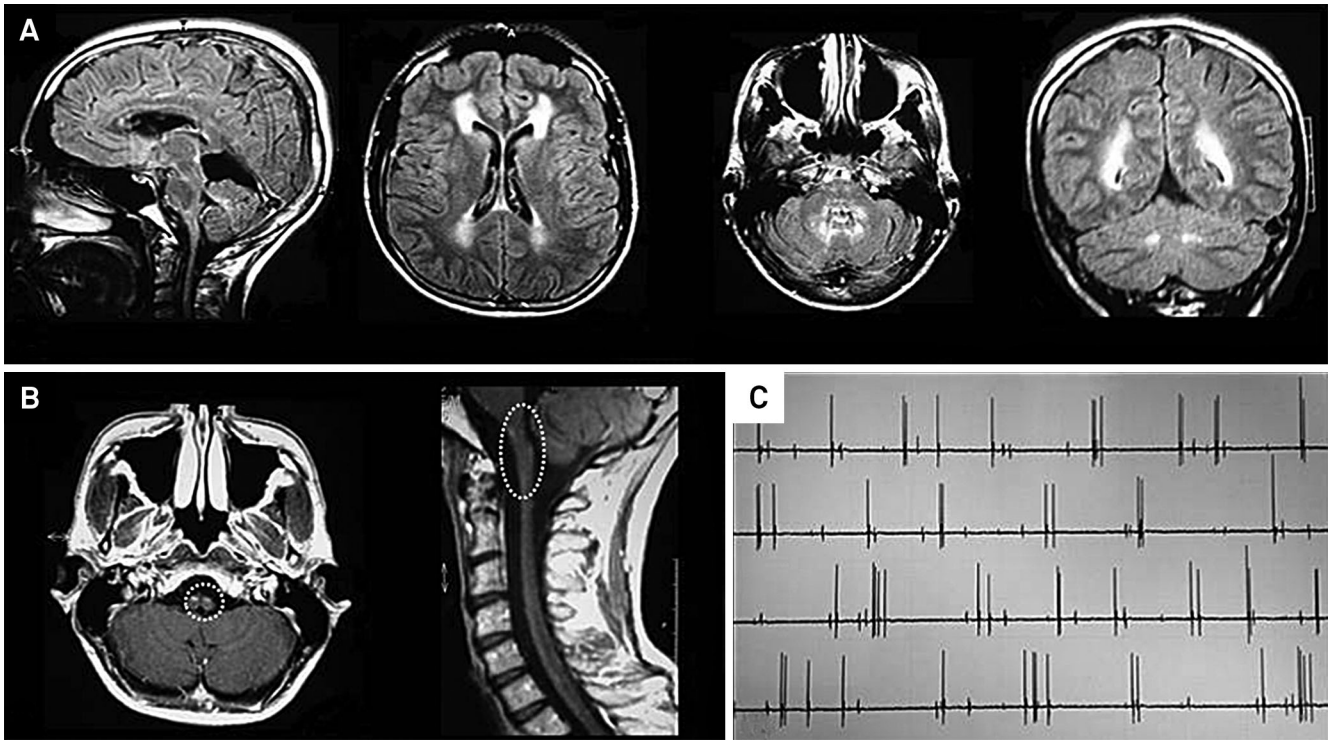


**Figure.** Brain and spinal cord magnetic resonance imaging studies showed: (A) infratentorial and upper cervical spinal cord atrophy (especially in medulla oblongata) with symmetrical areas of high signal on FLAIR images in the periventricular white matter of the brain hemispheres, brainstem and cerebellum; and (B) T1-weighted images following gadolinium contrast showing mild enhancement in medulla oblongata and upper cervical spinal cord (circles); and (C) Needle electromyography showed myokymic discharge characterized by groups of single or few spikes firing repetitively in a burst, recurring at intervals that are semirhythmic, in left orbicularis oris muscle (200uV/0.2seg).

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the clinical and electrophysiological characteristics of the adult-onset AD, as well as, AD in the differential diagnosis of facial myokymia.

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