Recurrent Ataxia and Dystonia with Anti-Neurochondrin Autoantibodies

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We report the case of a 7-year-old boy who developed severe recurrent episodes of ataxia. Following the persistence of intrathecal pleocytosis and oligoclonal bands, autoimmune workup was performed, and anti-neurochondrin[1] antibodies in the cerebro-spinal fluid were discovered. Other investigations returned normal, no oncologic accompaniments were found.

Clinical examination identified cerebellar ataxia, cervical dystonia, choreic movements of the upper limbs, action tremor, opsoclonus, dysarthria, and akathisis (►**video 1**). Cognitive

Table 1 Physiotherapy and cognitive assessment: significant motor improvement under corticosteroids (0–8 months), with subsequent stabilization under mycophenolic acid (8–18 months); lesser recovery in cognitive functions, including executive, attentional, and visuomotor functions

	Tests	Corticosteroid	Mycophenolate acid	
		T0	T1 (6 mo)	T2 (18 mo)
Physiotherapy assessment	MFM D1: standing and transfers	61.53%	76.92%	87.18%
	MFM D2: axial and proximal motor skills	77.77%	94.44%	97.22%
	MFM D3: distal motor skills	80.95%	85.71%	90.48%
	MFM total: global motor skills	71.87%	85.42%	91.67%
	6 MWT: walking test	378 m (accompanied)	408 m (alone)	536 m (alone)
Cognitive assessment	Long-term visual memory (NEPSY)	/	15/32 (<P5)	24/32 (P37)
	Attentional functions Alert (KITAP) Visual attention (NEPSY) Divided attention (KITAP) Distractibility (KITAP) Auditory vigilance (NEPSY)	/	667 (P1) 6 (<P5) 1,180 (P4) 28 (P8) 7 (P16)	710 (<P1) 8 (<P5) 946 (P14) 35 (<P1) 7 (P16)
	Executive function Flexibility (KITAP) Trail Making Test	/	11 (P4) 267''	12 (P1) 187''
	Visuo-motor precision (NEPSY)	/	2 (<P1)	2 (<P1)

Abbreviations: 6 MWT, 6 minutes walking test; KITAP, German test battery of attention performance; MFM, Mesure de la Fonction Motrice (measurement scale of motor function); NEPSY, neuropsychological assessment.

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affective cerebellar syndrome, including neuropsychological impairment associated with significant emotional lability, was also found (►**Table 1**).

Video 1

Walking test showing ataxic gait with deviation, instability especially during the turn around, increase space lift and excessive swinging arms; during the second part, the patient experiences difficulties in fine grasping and intention tremor. Note the presence of cervical dystonia (left laterocollis). Online content including video sequences viewable at: https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0040-1722675.

Initial corticosteroid therapy (intravenous methylprednisolone 30 mg/kg for 3 days) resulted in significant improvement. However, following the third relapse, a long-term corticotherapy with monthly intravenous methylprednisolone 500 mg/m^2 was initiated. After 8 months, to avoid side effects, this treatment was replaced by mycophenolic acid. No relapse was observed since then. The child currently keeps slightly ataxic gait as well as significant cognitive impairment.

Autoimmune etiologies of the movement disorders are increasingly recognized, even in children.[2,3] Unexplained repeated episodes of ataxia of subacute onset may require a workup with comprehensive neural IgG screening, especially since immune therapies seem more effective than what is observed in adults.[4]

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Conflict of Interest
None declared.

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