

UNILATERAL RIGHT HEMIPLEGIA DUE TO UPPER MOTOR NEURON DYSFUNCTION: A CASE REPORT OF MILLS' SYNDROME

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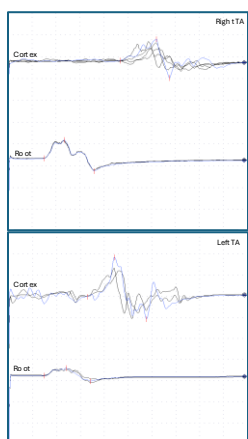


Background

We report the case of a patient with a ten-year history of progressive gait disturbances due to right lower limb weakness. After five years, the patient developed right upper limb weakness, prompting further investigations. A cervical MRI showed mild C4-C6 stenosis without spinal cord signal alterations.

Methods

Neurological examination exhibited right steppage gait with diffuse right-side muscle weakness and increased reflex on the right side. Electrophysiological studies showed normal needle EMG and nerve conduction study but exhibited markedly increased central motor conduction time from right orbicularis oris muscles, upper (brachial bicep, first dorsal interosseus) and lower (anterior tibialis and abductor hallucis) limb muscles. Somatosensory-evoked-potential showed no significance interside abnormalities. Spinal MRI identified a right-sided intramedullary signal alteration at C2, suggestive of axonal degeneration. Brain PET-CT revealed left hypometabolism in the sensory-motor area.



Muscle	Site	Right			Left		
		Lat (ms)	Amp (mV)	CCT (ms)	Lat (ms)	Amp (mV)	CCT (ms)
Orb Oris	Cortex	14.5	1.2	10.2	9.1	3.1	4.8
	Root	4.3	4		4.3	4.9	
FDI	Cortex	30.3	0.5	16	22.5	7.6	7.5
	Root	14.3	1.46		15	1.27	
TA	Cortex	46.8	0.89	31.7	32.9	2.9	18.1
	Root	15.1	0.71		14.8	0.62	
Abd hal	Cortex	54.1	0.3	24.3	49.8	1.32	20.6
	F wave	56.9	-		56.1	-	

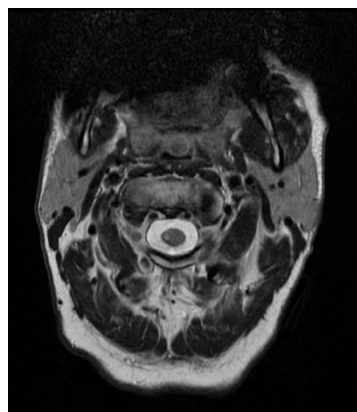


Figure 1 and 2. Motor Evoked Potentials (MEPs) study obtained recording from Tibialis Anterior (TA) right (figure up) and left (figure down).

Table 1. Motor Evoked Potentials (MEPs) study demonstrated a markedly increased central motor conduction time from right orbicularis oris muscles, first dorsal interosseus, anterior tibialis and abductor hallucis.

Figure 3. a T2-weighted axial MRI sequence of the brain showed a right-sided intramedullary signal alteration at C2, suggestive of axonal degeneration.

Results

All these findings supported the diagnosis of Mills' syndrome: an extremely rare neurodegenerative slowly progressive disorder characterized by unilateral upper motor neuron involvement.

Conclusions

This case highlights the diagnostic challenges of Mills' syndrome. The combination of clinical, electrophysiological, and imaging findings was crucial for a correct diagnosis. Long-term follow-up is essential to monitor disease progression and differentiate it from other motor neuron disorders.



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