

Segmental areas of denervation in post-polio syndrome

Desnervação segmentar na síndrome pós-pólio

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A one-year-old male disclosed motor developmental delay, appendicular hypotonia, hyporeflexia, and weakness in the left lower limb. He started walking only at three years old. After this time, his symptoms were stable. At 31 years old, the patient started slight asymmetric and slowly progressive paraparesis and myalgia. He was referred with a suspicion of spinal muscular atrophy. Electromyography disclosed bilateral chronic lumbosacral denervation and mild acute denervation involving the right lower limb. Neuroimaging studies (brain and spinal MRI) and genetic testing (MLPA test for quantification of SMN1 and

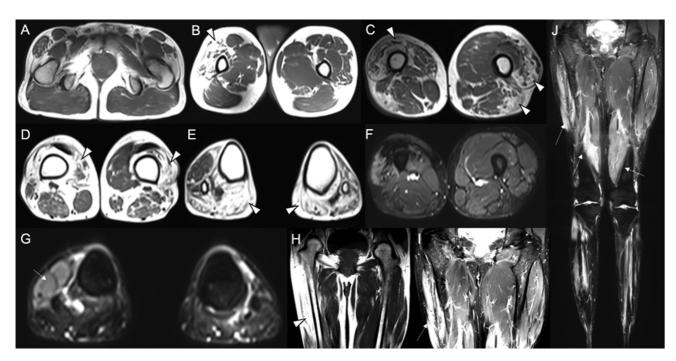


Figure 1 Muscle MRI studies. Axial (A-E) and coronal (H) T1-weighted MRI shows asymmetric amyotrophy and marked fatty replacement involving bilateral vastus lateralis, right vastus intermedius, left biceps femoris, right vastus medialis, left tibialis anterior, and bilateral heads of gastrocnemius. Axial (F,G) and coronal (I,J) STIR MRI shows hyperintensity involving right vastus lateralis and tibialis anterior.

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SMN2 copy numbers and hereditary neuropathy, non-5q, and other motor neuron disease panels) were normal. Muscle MRI disclosed asymmetric neurogenic muscle "islands" alternating areas with and without fatty replacement (**- Figure 1**),¹ suggestive of Post-Poliomyelitis Syndrome (PPS).^{2,3} Muscle MRI is useful to distinguish PPS from other motor neuron diseases.¹⁻³

Authors' Contributions

VLB: conceptualization, data curation, formal analysis, writing – original draft preparation and supervision; HLCM, PHAF, FPS: conceptualization, formal analysis, writing – original draft preparation; IBF: conceptualization, data curation and writing – original draft preparation; PLS: BMLB, MATC, WBVRP: Conceptualization, data curation, formal analysis, writing – review & editing and supervision; PVSS, ASBO: Conceptualization, data curation and writing – review & editing.

Conflict of Interest

There is no conflict of interest to declare.

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