Teaching NeuroImages: Hopkins syndrome

A rare differential diagnosis of neurogenic monomelic amyotrophy

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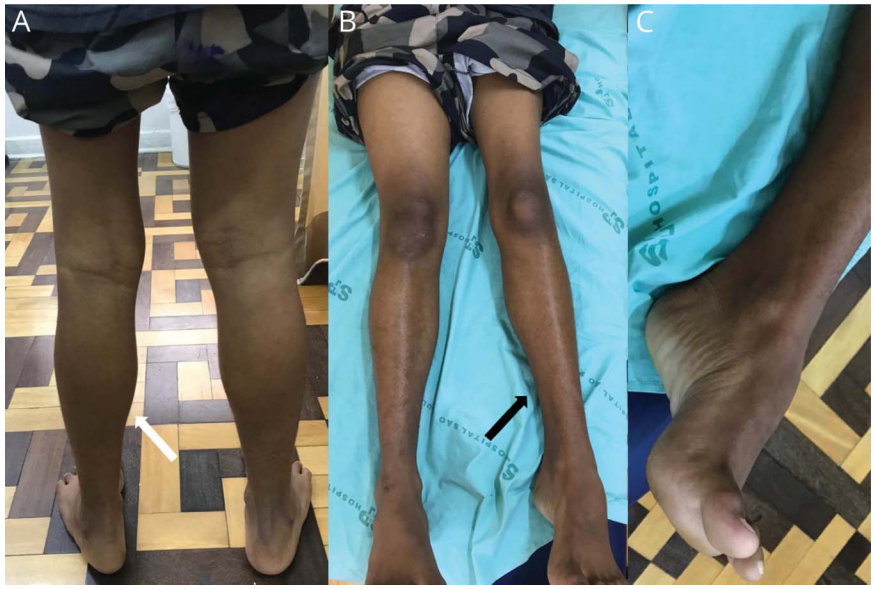
Figure Neurologic examination of the lower limbs in Hopkins syndrome



Marked monomelic amyotrophy involving proximal and distal portions the left lower limb (white and black arrows) (A, B), including pes cavus and toe retraction in the left foot (C).

An 11-year-old Brazilian boy presented with worsening left thigh weakness. At age 2 years, he presented with left lower limb weakness after 4 days of a severe asthma episode. Examination showed amyotrophy, weakness, and areflexia of the left lower limb (figure). Spine MRI and CSF analysis (including serology and PCR for herpesvirus, enterovirus, and arbovirus) were unremarkable. Neurophysiologic studies exhibited ongoing denervation and chronic reinnervation changes involving the left lower limb muscle groups. Our patient was diagnosed with Hopkins syndrome, a rare childhood-onset lower motor neuron disease presenting with monomelic amyotrophy after an acute asthma attack.[1,2]

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