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Minipolymyoclonus revealing Hirayama disease

Video Animation

Hirayama disease is a rare, sporadic, benign lower motor neuron disorder, affecting predominantly males in their second and third decade.

We hereby present the case of a 17-year-old male, presenting with a history of tremor and weakness in his left hand. Initially he wasn't concerned, as his mother suffered from essential tremor. However, over five months the weakness in the left-hand progressed causing difficulty in fine motor skills.

The physical exam revealed slight atrophy of the left-hand predominating on the ulnar side and a more marked atrophy of the medial left forearm with sparing of the brachioradialis. With fingers outstretched there was arrhythmic, low amplitude, rapid tremulous movement of fingers (Video 1). The left upper limb was weak grade 3 on the medical research council scale (MRC). There was no bulbar or pseudobulbar involvement. Superficial and deep sensory examination was normal. There were no signs of upper motor neuron involvement.

Electromyogram showed severe chronic denervation in C7 and C8 muscles with sparing of the brachioradilais.

Trans-spinal magnetic stimulation did not elicit a motor response in the left upper limb.

Cervical magnetic resonance imaging (MRI) in neutral position did not show any abnormality other than a loss of cervical lordosis.

Because of the high clinical and neurophysiological suspicion of Hirayama disease, cervical flexion MRI was

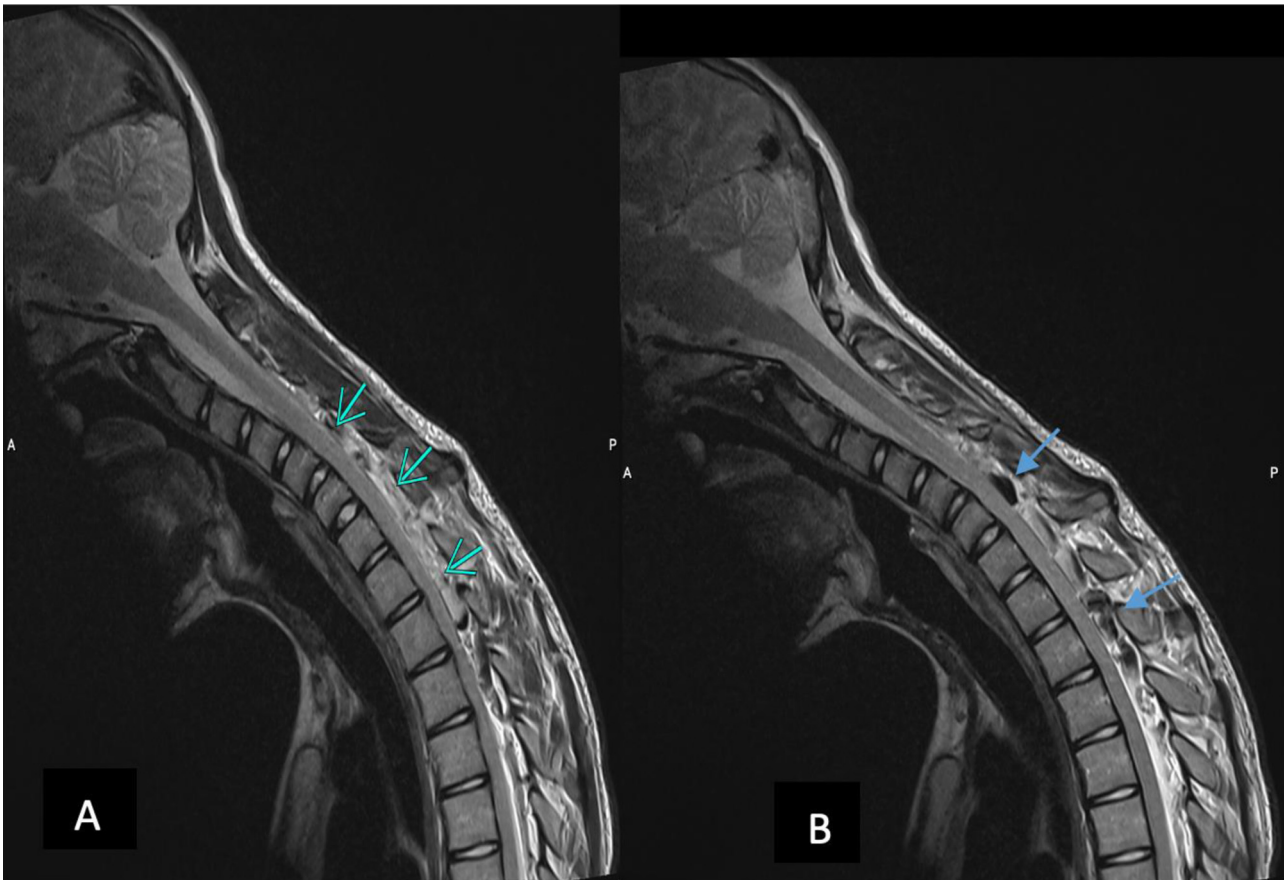

Fig. 1 – Sagittal T2 weighted flexion cervical MRI. A. Sagittal T2 weighted flexion cervical MRI showing enlarged epidural posterior space. B. Sagittal T2 weighted flexion cervical MRI showing turgescent epidural veins.

performed showing epidural space expansion from C4–C5 to mid thoracic level accompanied by turgescent epidural veins (Fig. 1).

Treatment consisted in wearing a cervical collar. At six months there was no further progression of the disease.

1. Discussion

In his 1963 paper, Hirayama described tremorlike movement of fingers, often observed at rest in the thumb, index and little finger. Our patients presented the same involuntary movements consistent with minipolymyoclonus.

In terms of pathophysiology, minipolymyoclonus may be the reflect of anterior horn cells disruption manifesting with frequent fasciculations [1]. Growth spurts occurring in puberty with disproportional vertebral growth outrunning the cord may lead to tightness of the dura. During neck flexion because of relative tightness of dura, enlarged epidural posterior space pushes the cord towards the posterior vertebral margins. In time, this may lead to chronic compression and ischemic changes to the anterior horn cells [2]. Autopsy findings in a patient with Hiarayma disease favor the hypothesis of chronic ischemic changes, revealed by necrosis and mild gliosis at C7 and C8 level [3].

In our patient cervical, MRI did not show signs of chronic ischemia. However, absence of spinal motor evoked potential for the left upper limb was in favor of lower motor neuron lesion in the C8 myotome.

Natural history of Hirayama disease is generally self-limited in time. Progression will stop in less than five years in 62% of patients [4].

Cervical collar has been found useful in slowing the progression of the disease [5].

A recent systematic review and meta-analysis of patient data found that surgical treatment was responsible for improvement of neurological symptoms in 85% of treated patients [6]. Due to scarce reports and rarity of the disease, there are no clear indications for surgery.

Disclosure of interest

The authors declare that they have no competing interest.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.neurol.2024.02.391.

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