Hirayama disease: Role of diffusion tensor imaging

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CASE REPORT

A 28-year-old male patient presented with progressive weakness of both forearms and hands along with clawing of hands for past eight years. Right side was affected more than the left. There was no history of similar illness in the family. There was no past history of any prolonged illness in childhood. There was no history of sensory, pyramidal or bladder dysfunction. There was atrophy of muscles of both forearms and hands. Muscle power was normal in both upper limbs. Nerve conduction studies showed reduced amplitude in bilateral median and ulnar nerves. Tendon reflexes were normal. Magnetic resonance imaging (MRI) examination was performed in non-flexion and flexed position. Subsequently, diffusion tensor imaging (DTI) was performed in non-flexion, which very clearly showed the cord atrophy at C7–T1 level (Figure 1). This cord atrophy was not appreciated on MRI flexion and non-flexion studies. The diagnosis of Hirayama disease (HD) was thus established on DTI.

Figure 1: Diffusion tensor magnetic resonance image acquired in non-flexion shows atrophy of spinal cord at C7–T1 level (arrows).

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DISCUSSION

Hirayama disease (HD) is a non-progressive, juvenile, spinal muscular atrophy of distal upper limbs and is a kind of cervical myelopathy related to flexion movements of the neck. Imbalance in growth of

vertebrae and dura mater causes a loss of normal dural slack in extension. As a result of this, the tight dural canal causes compression of the spinal cord [1]. It is suggested that HD might be due to microvascular changes following chronic trauma to spinal cord during flexion and extension of the neck [2]. On non-flexion MRI studies, asymmetric cord atrophy especially at the lower cervical level is highly suggestive of HD [3]. Furthermore, the magnetization and diffusion MRI histograms of the cervical cord suggest that the cord damage in HD extends beyond that seen on routine MRI scans [4]. In our opinion, this atrophy of the spinal cord is best demonstrated on diffusion tensor imaging. This makes diffusion tensor imaging a valuable tool in diagnosing Hirayama disease without subjecting the patient to a flexion study of the cervical spine.

CONCLUSION

Diffusion tensor imaging is simple, fast and accurate method for diagnosing Hirayama disease without subjecting the patient to flexion magnetic resonance imaging study.

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Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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