P-14 Severe bilateral Hirayama disease with delayed emergence of intramedullary T2 high signal intensity

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Introduction

Hirayama disease (HD) is a form of cervical myelopathy presenting unilateral amyotrophy in the C7-T1 myotomes, which is related to neck flexion. The progression typically arrests within 3 to 5 years. We report a patient of severe bilateral HD with long-term MRI follow-up, which showed delayed emergence of T2 high signal intensity in anterior horn cells (AHC).

Case Presentation

A 21-year-old man gradually developed bilateral forearm weakness and amyotrophy for 9 months. Clinical examination revealed symmetrical muscular atrophy of the hands and forearms, reduced muscular strength during flexion, extension, and abduction of the fingers, and exaggerated tendon reflexes of biceps and triceps. Needle electromyography showed denervation of the atrophied muscles. Cervical MRI only showed a myelopathy at C6 level. HD, motor neuron diseases, and cervical spondylosis were suspected, and methylcobalamin was prescribed. 2 months later, MRI was conducted again, and revealed anterior disposition of the cervical spine in flexed posture, which confirmed a diagnosis of HD. Despite the treatment, the muscle weakness progressed 1 year later, and MRI showed compression of spine at C5-T1 level by enlarged epidural space, but no intramedullary change on T2-weighted images. 2 year later, symptoms got worsened, and MRI showed T2 high signal intensity in bilateral AHC at C6 level. Finally, he was made to quit his school, and start working, because it became difficult for him to use experimental tools.

Discussion

Although HD is said to be a disease with a good prognosis, there are some severe forms like our patient. The present case shows delayed emergence of T2 high signal intensity in AHC, which implies myelomalacia caused by compression of cervical cord with micro-circulatory changes. Because HD can limit the choice of occupation and ADL, early diagnosis and treatment are important.