

Cervical Spine Deformity as a Pathophysiological Cause of Hirayama Disease

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Abstract

Background: Hirayama disease (HD) is a cervical segmental myelopathy characterized by juvenile onset of muscular atrophy in unilateral upper extremity. In HD, cervical spine deformities may be frequent comorbidities; however, details are unknown. The aim of this study was to quantitatively assess cervical spine deformities in patients with HD.

Methods: We enrolled 16 patients with HD and 55 healthy subjects younger than 30 years who underwent cervical spine magnetic resonance imaging (MRI) between January 2009 and December 2015. We evaluated cervical spine sagittal alignment with the neck in neutral position using the following methods: C2–7 Cobb angle, centroid measurement of cervical lordosis (CCL), absolute rotation angle (ARA), C7 slope (C7S), and Ishihara curvature index (ICI) according to previously established reports.

Results: C2–7 Cobb angle, CCL, ARA, and C7S in HD were significantly less than in controls (mean C2–7 Cobb: -4.28° vs 5.15° , $p < 0.01$; mean CCL: -5.15° vs 7.25° , $p < 0.01$; mean ARA: -3.38° vs 9.21° , $p < 0.01$; C7S: 5.97° vs 15.0° , $p < 0.01$). Additionally, ICI in HD was smaller than in controls (mean ICI: -2.00 vs 8.48 , $p < 0.01$). There was no statistically significant difference in stage of disease between progression and stable.

Conclusions: This study demonstrates cervical spine deformities in patients with HD using multiple quantitative measurements of cervical spine alignment. These findings also indicate that straight neck or even kyphosis is frequently seen in patients with HD compared with controls. Cervical spine deformities might permanently remain and contribute the pathophysiology of the HD.

Key words: juvenile muscular atrophy, Hirayama disease, magnetic resonance imaging (MRI), cervical spine deformity