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Changes in the soleus H-reflex test and correlations between its results and dynamic magnetic resonance imaging abnormalities in patients with Hirayama disease

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HIGHLIGHTS

- Hirayama disease (HD) patients may develop cervical spinal cord injury with disease progression.
- Upper motor neuron lesions are more likely in HD cases with severe neck-flexion abnormalities.
- Cervical cord forward-shifting worsens as disease advances, indicating that HD may not be a self-limiting disorder.

ABSTRACT

Objective: To investigate changes in soleus H-reflex tests in patients with Hirayama disease (HD) and to analyse correlations between these changes and forward-shifting of the cervical cord during neck flexion.
Methods: The amplitude of the soleus H-reflex with and without vibration on the Achilles tendon was recorded bilaterally in 81 HD patients and 34 controls to measure both the vibratory inhibition index (VII) and the H_{max}/M_{max} ratio. The maximum forward-shifting degree of cervical cord during neck flexion was measured using dynamic magnetic resonance imaging in all HD patients.
Results: Significantly higher VII was recorded in 6/81 (7.4%) HD patients, along with abnormal H_{max}/M_{max} ratios in 5 of 6 cases. Compared to illness duration ($r = 0.29–0.36$, $p < 0.05$), the maximum forward-shifting degree of the cervical cord was more strongly correlated with both VII and the H_{max}/M_{max} ratio ($r = 0.51–0.81$, $p < 0.05$).
Conclusions: HD patients may develop cervical spinal cord injury with disease progression, and these lesions may be more likely to occur in cases with relatively severe cervical-flexion structural abnormalities even during early stages.
Significance: More caution should be taken when managing HD patients with severe cervical-flexion abnormalities because of the possible early occurrence of upper motor neuron lesions.

1. Introduction

Hirayama disease (HD) was first reported by the Japanese scholar Keizo Hirayama in 1959 (Hirayama et al., 1959). The aetiology of HD remains controversial. Currently, the main implicated mechanism is chronic cervical spinal cord ischaemia caused by microcirculatory disturbances in the territory of the anterior spinal artery during neck flexion (Hirayama, 2000, 2008). Because anterior horn cells are extremely vulnerable to ischaemia and a previous autopsy case of HD revealed confined necrosis in both the cervical anterior horn and ventral nerve root areas (Hirayama et al., 1987), HD is considered a neurological disease that involves only lower motor neurons (LMN) (e.g., anterior horn cells and ventral nerve roots)

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(Hirayama, 2008). However, both Zhou et al. and Wang et al. found that approximately 2.7–6.3% of HD patients had significant lower limb hyperreflexia, consistent with lesions at the level of upper motor neurons (UMN) (Zhou et al., 2010; Wang et al., 2012). Similar conditions were also reported in our previous studies (Zheng et al., 2016, 2017a). Furthermore, recent case reports have demonstrated that some HD patients may develop apparent long tract signs with disease progression (Sakai et al., 2011; Li and Remmel, 2012). All of these findings suggest that the persistent microcirculatory disturbances that are induced by neck flexion may simultaneously cause abnormalities in corticospinal tracts in the watershed zone of the anterior spinal artery.

The H-reflex was first described by Hoffmann and is evoked by applying electrical stimulation to the low-threshold Ia muscle spindle afferents that activate alpha motoneurons (Fisher, 1992). Because of the monosynaptic nature of the early part of the response, the H-reflex can be safely used as a measure of descending corticospinal tract, which controls segmental motor excitability (Schieppati, 1987). Previous studies demonstrated that the suppression of the H-reflex during vibration is significantly decreased in patients with UMN lesions, whereas the ratio between the maximal soleus H-reflex response and muscle potential (H_{max}/M_{max}) is clearly increased (Taylor et al., 1984; Ongerboer de Visser et al., 1989; Lee et al., 2005).

In the current study, we quantitatively evaluated corticospinal tract function in HD patients using these soleus H-reflex tests (both the H_{max}/M_{max} ratio and the vibratory inhibition index of H-reflex). In addition, the correlations between cortico-spinal tract function and disease duration, clinical characteristics and cervical spinal cord forward-shifting were also analysed.

2. Methods

2.1. Subjects

Eighty-one patients with HD and 34 age- and sex-matched healthy subjects were included in the current study. All patients were recruited at the Huashan Hospital between October 2011 and February 2017. The Human Ethics Committee of Huashan Hospital at Fudan University in China granted ethical committee approval, and each subject provided informed consent. In this study, thirty-four subjects in the patient group were tested using the completely different methods from those used in our previous studies, which had different research objectives, and no main results from prior reports were repeated in this study (Zheng et al., 2017a,b).

The subjects in the control group exhibited normal neurological examination results, including normal muscle strength, sensory function and deep tendon reflexes. Subjects who had undergone cervical spinal surgery and who exhibited current or past cervical radiculopathy, brachial plexus injury, carpel tunnel syndrome, peripheral neuropathy, muscular dystrophy, diabetes, or peripheral vascular disease were excluded from the control group (Zheng et al., 2016, 2017b).

The diagnostic criteria for HD were (Zheng et al., 2016, 2017b) (1) insidious disease onset before 25 years of age, (2) unilateral or asymmetric weakness and amyotrophy of the distal upper extremities without sensory dysfunction or lower limb involvement with denervation that was limited to the unilateral or bilateral upper extremities and was identified using electrophysiological methods in addition to normal sensory nerve function, (3) with or without clinical symptoms, such as "cold paralysis", "fasciculation" and "tremor in the upper extremities", and (4) cervical-flexion magnetic resonance imaging (MRI) showing lower cervical compression that resulted from forward shifting of the posterior dura and a crescent abnormality posterior to the dura.

The exclusion criteria were (Zheng et al., 2016, 2017b) (1) a history of syringomyelia, spinal cord tumour or abnormalities in the cervical vertebrae, (2) focal or multifocal neuropathy, (3) brachial plexus injury, (4) congenital muscular dystrophy, (5) cervical spondylotic amyotrophy, (6) a primary or concomitant disorder of the neuromuscular junction, or (7) concomitant trauma, inflammation or infection.

2.2. Testing methods

All subjects were asked to lay comfortably prone during the recordings. A pillow was placed under the ankle while the knees were slightly flexed at a 120-degree angle, and the ankles were flexed at a 110-degree angle to establish optimal relaxation of the soleus muscle and to avoid the change of the position of the ankle (Kimura, 2001). The standard instructions for the test were provided along with a brief demonstration, and we ensured that all subjects were familiarized with the equipment and the testing procedures before the tests were performed.

Bilateral H-reflexes were recorded from the soleus muscle (Sol H-reflex) for tibial nerve stimulation at the popliteal fossa while the cathode of the surface stimulator was proximal to the anode (Fig. 1A). To electrically stimulate the tibial nerve, we applied 1.0 ms uniphasic pulses through circular electrodes that were 0.3 cm in diameter. The stimuli were delivered every 10 s. The position of the stimulating electrode was systematically adjusted to obtain the H-reflex at the lowest possible threshold in the soleus muscles. Then, the intensity was adjusted to elicit the maximum H-reflex amplitude for each muscle tested. The maximum compound muscle action potential (CMAP) was also recorded from the soleus muscle using surface electrodes. The active recording electrode for the soleus muscle was placed over the medial bulge of the muscle, below the gastrocnemius margin. The reference electrode was placed 3 cm distal to the active electrode, and the ground electrode was placed between the stimulator and the recording electrodes (Fig. 1A).

2.2.1. H_{max}/M_{max} ratio of the H-reflex

The H-reflex was verified as the presence of classical periodic changes in the H-reflex amplitude with increasing stimulation intensity (Kimura, 2001) (Fig. 1). Once the maximum H-reflex was obtained, the same stimulus intensity was used to obtain five consecutive recordings to calculate the average maximum H-reflex amplitude (H_{max}). At the intensity that elicited the maximal M-wave amplitude, five responses were recorded to measure the average maximum M-wave amplitude (M_{max}). These measurements were then used to calculate the H_{max}/M_{max} ratio.

2.2.2. Vibratory inhibition index of the H-reflex

A hand-held vibrator (Shuang Quan SQ-B08, Wenzhou, China; frequency: 50 Hz) was applied to the Achilles tendon of the tested ankle for 300 s, and the same stimulus intensity that was previously used to elicit the maximum H-reflex amplitude was delivered (Fig. 1A) (Lapole et al., 2012). Five consecutive H-reflex amplitudes were then measured during vibration to measure the average H-reflex amplitudes (H_{vib}). The vibratory inhibition index (VII) of the H-reflex was measured using the following formula: $(H_{vib}/H_{max}) \times 100\%$.

The measurements obtained from the HD patients were considered abnormal if both H_{max}/M_{max} ratios and VII were 2 standard deviations (SDs) above the mean values for the controls. All electrophysiological procedures were performed using computer-based EMG equipment (Nihon Kohden MEB-9400, Japan), and all tests were carried out at a skin temperature above 32 °C.