Journal of the Neurological Sciences 359 (2015) 213–216

Contents lists available at ScienceDirect

Journal of the Neurological Sciences

journal homepage: www.elsevier.com/locate/jns

Clinical short communication

Do longer necks predispose to Hirayama disease? A comparison with mimics and controls☆

Satish Khadilkar *, Bhagyadhan Patel, Ashish Bhutada, Chetan Chaudhari

Department of Neurology, Grant Medical College and Sir J. J. Group of Hospitals, Mumbai, India

ARTICLE INFO

Article history:
Received 28 August 2015
Received in revised form 1 November 2015
Accepted 3 November 2015
Available online 4 November 2015

ABSTRACT

Background: Dynamic changes in cervical spine during flexion is a proposed mechanism for Hirayama disease [HD], a localized form of anterior horn cell disorder. Apparent shortening of dura as compared to vertebral column leading to dural shift on flexion is considered to be the primary mechanism in this hypothesis. Whether this disproportion is a result of short dura or longer cervical segment is not known and neck length has not been studied in HD. Also, all patients with segmental motor weakness and wasting of upper limbs do not show dural changes; hence comparative evaluation of HD and its mimics is important.

Material and methods: Patients with segmental wasting and weakness limited to upper limbs were subjected to flexion MRI. A special pillow was designed to provide fixed flexion angle of 35°. Patients showing dural changes formed the HD group while rest formed the non-HD group [mimics]. Both groups were analyzed on clinical, electrophysiological and radiological parameters. Whole spine to neck ratio of patients in HD group was compared to the non-HD group and age matched controls.

Results and conclusions: Patients with HD had longer cervical segments as compared to the non-HD group and age matched controls [$p = 0.001$]. The longer cervical segment, in combination with dural changes probably contributes to the pathophysiology of dynamic flexion hypothesis of HD and the onset around the growth spurt. Patients with HD had cold paresis and finger extensors were consistently weaker than flexors. Apart from longer necks, cold paresis and pattern of weakness may help to differentiate HD from its mimics.

1. Introduction

Hirayama disease (HD) affects young males and results in segmental wasting and weakness of one or both upper limbs. HD is reported largely from Asian countries and seems to be uncommon in the western regions [1] Occurrence in adolescence, male preponderance, self-limiting course and geographic distribution are important features of HD. No single explanation to account for all these features is available. However, dynamic changes in cervical cord during flexion have been consistently observed on Magnetic Resonance Imaging (MRI) of patients with HD [2 [3]. This hypothesis revolves around the dural changes during flexion and dynamic compression leading to degeneration of anterior horn cells of cervical cord. Shortening of the dura in relation to the vertebral column and resultant anterior shift during flexion is an important part of this hypothesis [4]. Longer cervical segment can be one of the contributory factors to the dural changes. The present study was undertaken to evaluate this new aspect. A proportion of patients presenting with segmental upper limb wasting do not have the characteristic MRI features of HD; the HD mimics. This study also compared HD and its mimics on clinical and investigative parameters.

2. Methods

This prospective observational study was carried out at the neurology department of a tertiary care university hospital from February 2014 to April 2015. Patients were recruited after written informed consent and ethics committee approval was obtained.

2.1. Inclusion criteria

Patients presenting with unilateral or bilateral pure motor weakness of upper limbs.

2.2. Exclusion criteria

Patients having lower limb involvement, sensory deficits, pyramidal signs and those who did not give consent.

Patients underwent detailed history and examination. Family history was recorded. Cold paresis was considered present when there was

☆ Work done at: Department of Neurology, Grant Medical College and Sir J J Group of Hospitals, Mumbai.

* Corresponding author at: 110, New Wing, First Floor, Bombay Hospital, 12, New Marine Lines, Mumbai 400020, India.
E-mail addresses: khadilkarsatish@gmail.com (S. Khadilkar), bhagyadhan30@gmail.com (B. Patel), drashishbhutada@gmail.com (A. Bhutada), chtn.chaudhari@gmail.com (C. Chaudhari).

http://dx.doi.org/10.1016/j.jns.2015.11.005
0022-510X/

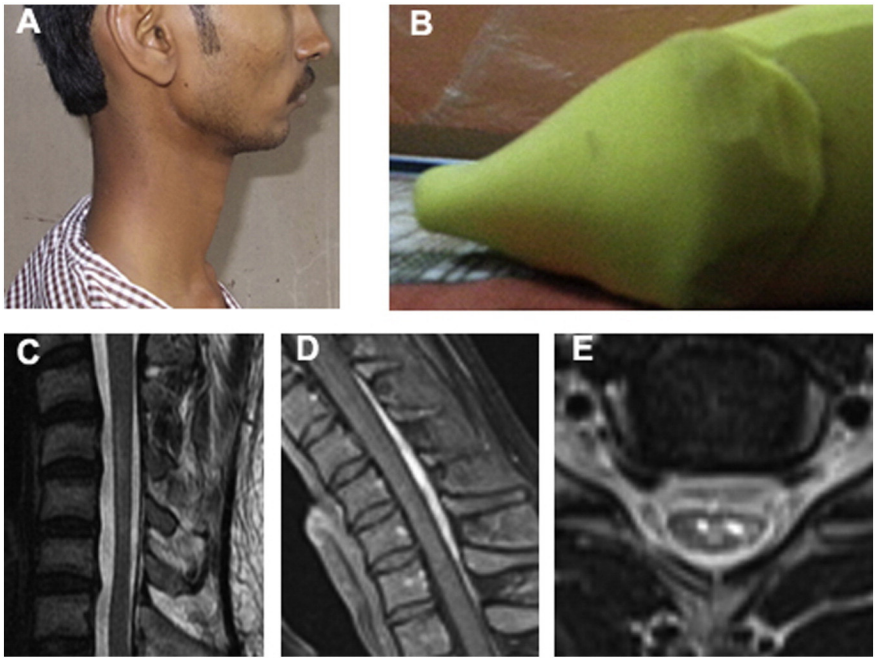

Fig. 1. Long neck [A], pillow designed to provide angle of 35° [B], MRI T2 sequences in neutral position showing cord atrophy [C], MRI T1 fat sag post-contrast sequence in flexion showing enhancement of posterior epidural space [D] and Axial T2 sequence showing intramedullary signals (snake eye appearance) [E].

history of subjective worsening of weakness on exposure to cold. In skeletal examination, particular attention was given to neck length and whole spine length. These were recorded as follows. Neck length was measured as linear measurement between external occipital protuberance and spinous process of C7 vertebra with neck held straight in neutral position. Whole spine length was measured from external occipital protuberance to tip of coccyx in sitting position. The ratios of whole spine length to neck length were calculated [5].

2.2.1. Electrophysiological studies

An 8 channel Neurosoft electromyography equipment was used to perform electrophysiological studies in all patients. Detailed nerve conduction studies including sensory and motor parameters were performed [6]. In electromyography, particular attention was given to C5–T1 segments, thoracic para spinals and lumbosacral segments.

2.2.2. Radiological studies

To achieve optimum flexion angle for eliciting changes of HD (35°) [7], a special pillow was constructed using polyurethane, which was MRI compatible. It provided a fixed angle of 35° of neck flexion [Fig. 1: B]. MRI was done using a 1.5 Tesla Siemens operating system. Following sequences were performed on cervical spine in neutral position 1) T1 weighted sagittal and axial 2) T2 weighted sagittal and axial MRI sequences. After achieving 35 degree neck flexion, 3) pre- and post-contrast T1 weighted fat suppression sagittal and axial images and 4) T2 weighted sagittal and axial images were obtained. Canal stenosis was defined as Torg ratio (sagittal spinal canal diameter to mid-vertebral body diameter) <0.8 on T2 sagittal cervical spine [8].

Patients who met the inclusion criteria were divided into two groups; HD and non-HD group.

2.2.3. HD group

Patients fulfilling clinical diagnostic criteria as proposed by Hirayama [9] and having dural changes [loss of attachment, engorgement of epidural venous plexus and contrast enhancement in epidural space] on 35 degree flexion MRI were included in HD group [Fig. 1: C–D].

2.2.4. Non-HD group [mimics]

Patients who did not fit the criteria for Hirayama disease (HD) were included in non-HD group. They were divided into the following subgroups: Canal stenosis, segmental distal SMA and segmental proximal SMA [10].

2.2.5. Controls

Unaffected males and females in age group from 18 to 22 years were recruited as controls. They underwent neck length and whole spine measurements.

2.3. Statistical tests

Mann Whitney, Chi square and one way Anova tests were applied. p value less than 0.05 was considered significant.

3. Results

All 40 were males. 26 (65%) patients fulfilled criteria of HD. Remaining 14 patients (35%) formed the non-HD group. Non-HD group comprised of

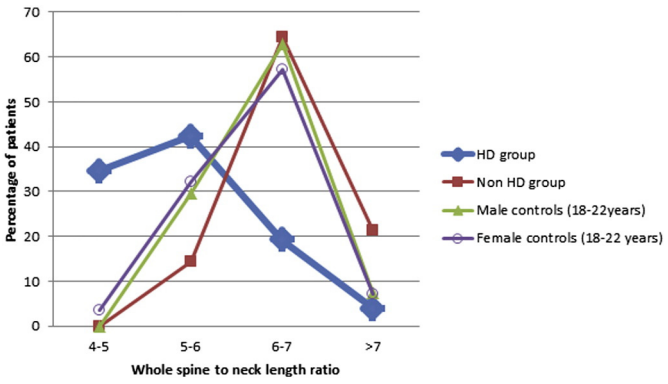

Fig. 2. Percentage distribution of whole spine to neck length ratio in HD, non-HD, male and female controls. HD group has significantly lower ratios [p = 0.001].