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Letter to the Editor

Evaluation of two female patients of Hirayama disease supports the longer neck hypothesis

Dear Editor,

Hirayama disease (HD) or Juvenile muscular atrophy most often affects young asian males in their teens and female patients are seen only exceptionally [1]. Patients present with asymmetric wasting and weakness of the distal upper limb musculature, the oblique amyotrophy. MRI shows dural changes in flexion [2]. Various causative explanations have been put forth. Recently, longer cervical segments have been documented in male patients with HD while patients with non HD causes of upper limb amyotrophy do not have long necks [3]. As females are rarely affected by HD, the study of neck length in them assumes importance. Here we describe two female patients with characteristic findings of HD who had long cervical segments.

1. Case description

Case 1: An 18 years old female presented with slowly progressive thinning of left hand and left middle finger drop of six months duration. She noted tremulousness of the weak digits. There were neither sensory symptoms nor any twitching of muscles. She did not complain of any cranial nerve dysfunction and had no symptoms pertaining to the lower limbs. The motor system examination showed wasting of first dorsal interosseus and abductor digiti minimi muscles on the left side. There was weakness of small muscles of both hands affecting extensors more than flexors, being more prominent on the left side. Polyminimyoclonus was seen on both sides. Muscle strength was normal in the proximal arms and in both the legs. All deep tendon stretch reflexes were normal. Sensory examination did not reveal any abnormalities. Thus, clinically she had a pure motor, asymmetric, progressive lower motor neuron type distal upper limb weakness with wasting and polyminimyoclonus. This pointed to an anterior horn cell disorder. Considering the age and the distribution, HD appeared more likely than other causes of young onset anterior horn cell diseases and spinal cord pathologies. Cervical spine MRI was done with contrast administration which revealed asymmetric atrophy of left hemicord from C4 to C7 segments, with dural changes of HD in flexion, showing prominent posterior epidural space. (Fig. 1, C–D). Nerve conduction studies confirmed chronic motor axon degeneration of the C7, 8 and T1 segments, being more severe on the left side. The diagnosis of HD was thus confirmed and the measurements of cervical segment of the spine vis a vis the whole spine were documented.Neck length

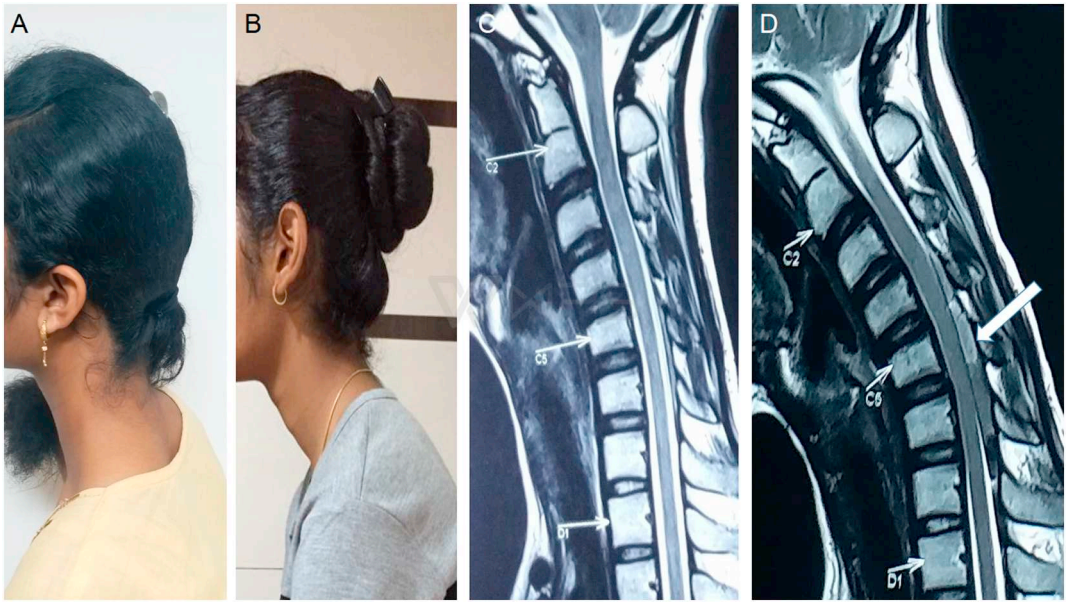

Fig. 1. Patient 1(A) and 2(B) showing long necks, MRI cervical spine T2 sequence in neutral (C) and flexion positions (D) showing prominent posterior epidural space (white thick arrow) in flexion.

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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. [3] Interestingly, patient has long neck(Fig. 1,A) with spine to neck ratio (57cm/14cm) of 4.14.

Case 2: The second patient ,an 18 years old girl presented with progressive pure motor dysfunction of C7,C8 and T1 segments with characteristic MRI changes of HD. She also had long neck (Fig. 1,B) and her neck measurements were spine length 74cm and neck length 14cm with spine to neck ratio of 5.28.

2. Discussion

Assessment of these two female patients makes an important observation.

HD is outstandingly common in males and descriptions of females having HD are rare. Large cohort studies have documented this male preponderance (China: $n = 192$, m:f = 7:1 [4], Japan: $n = 333$, m:f = 8:1 [5],India $n = 224$, m:f = 9:1 [6]. The reasons for such stark male preponderance are largely unknown.

In males with HD, the proportion of whole spine to its cervical segment has been documented to be lower, i.e. the necks longer. This study [3] compared the proportion of cervical segments of patients of HD with non HD and controls. Results demonstrated longer cervical lengths in HD patients, with lower whole spine to neck ratios (mean 5.61) when compared to controls (mean 6.20 in males and 6.24 in females).While the frequency of this association is very high, its causative relationship with the dural changes is presently speculative.It has been proposed that disproportionate growth of the vertebral column and the contents of the spinal canal during the juvenile growth spurt causes HD [7]. The shortening of dura relative to the vertebral column and resultant anterior shift of dura during neck flexion is believed to cause dynamic compression of cord leading to degeneration of anterior horn cells of cervical cord.

The explanation for the male preponderance could be the different growth rates in males and females. A Japanese study of the normative data documented that the average velocity curve for females peaks at 11 years of age, two years earlier than for males, and the annual average increase in height at the peak is about 8 cm, which is 2 cm less than for Males [6]. The slope of the average growth velocity curve is also less steep in females when compared to males.

The documentation of longer cervical segments in these two female patients is relevant as it further substantiates its bearings on the pathophysiology of HD.

Authors contributions

Satish Vasant Khadilkar: Design, text preparation, figure selection and final editing.
Madhu Bala Singla:Data collection, text and figure preparation.
Pawan Ohja:Data collection and figure preparation.
Sunila Jaggi: Figure selection and legend preparation.

Conflict of interest

No.

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