Cervical spine MRI in Hirayama disease: practical aspects

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Learning objectives

To understand etiology, pathogenesis, clinical and MR-signs of Hirayama disease (HD);

To learn how the standard cervical MRI protocol can be modified in order to confirm HD in clinically supposed case.
Background

Juvenile muscular atrophy of unilateral upper extremity with benign course was reported by Keizo Hirayama in 1959\textsuperscript{1}. Decreased anteroposterior diameter of the lower cervical in these patients was demonstrated by computed tomography with intrathecal contrast. In 1987 results of biopsy investigation of spinal cord were published. Authors showed decrease number of both large and small nerve cells and degenerative changes and suggest a circulatory insufficiency in the lower cervical cord as pathogenesis\textsuperscript{2}.

There are follow clinical characteristics of Hirayama disorder: greater prevalence in Far East and Asia (Japan, China, India, some published cases in Europe, Australia and America), males 15-25 years old suffering with 1-5 years of progression and then stabilization of neurological deficit\textsuperscript{3-5}. Predominantly it takes one upper extremity, but involvement of both upper extremities and pyramidal signs occur\textsuperscript{6}.

The gradual onset and progressive course of the disease, isolated motor neuron involvement may mimic early stage of the amyotrophic lateral sclerosis. In view of different prognosis in these pathologies early diagnosis is very important. Moreover early recommendation of cervical collar wearing can prevent increasing weakness in arm.

HD was thought to be not endemic for Russian population and we didn’t find publications about HD cases in Russia in PubMed, ScienceDirect and e-library (Scientific electronic library in Russia), but only during year 2015 we investigated 6 HD patients in Research Center of neurology in Moscow. So it is important to be familiar with MR signs of this pathology in order to be able to recognize it.
Findings and procedure details

Case 1

A 20-year-old male, Russian, had weakness of the left hand and forearm, periodic trembling of left hand fingers at the time of consultation. He had pituitary dwarfism with somatotropin insufficiency and took hormone therapy since 4 to 13 years old. 16-year-old he noted weakness of the left hand. Weakness and wasting of the hand were progressing during the year and than plateauing over 4 years.

Neurological examination revealed marked hypotrophy of left hand and distal left forearm. Right hand was slight hypotrophical (Fig. 1). Power was reduced with left finger abduction and flexion. Left carporadial reflex was hypoactive, another limb reflexes were all normal.

Neutral-position cervical spine MRI showed abnormal cervical curvature, loss of attachment (LOA) between the posterior dural sac and subjacent lamina, asymmetrical spinal cord flattening at C5-C7 vertebral body level. In flexion position anterior shifting of the posterior dura and increase of spinal cord compression were visualized (Fig. 2B).

According to long period (4 years) of stable symptoms only recommendation to prevent of long periods of neck flexion was given. In 3 months the patient was examined again and signs of HD progression were not revealed.

Case 2

Patient 2 is a 21-year-old male of Caucasian (Chechen) descent who presented with a 27 month history of left upper limb weakness. Initially, the illness progressed slowly and patient didn't take care about it. In the August 2013 he began to train a lot, exercises with flexion neck position were also performed. In the August 2014 patient noticed wasting of the left hand and forearm, which worsened in cold weather and progressed.

Neurological examination revealed hypotrophy of left hand and left forearm, posterior interossei muscles of the right hand. Power was reduced with left finger and hand flexion and extension. Upper limb reflexes were reduced on the left.

Normal-position cervical spine MRI revealed abnormal straightening of the cervical lordosis and flattening spinal cord at the C6-C7 levels with some intramedullary hyperintensity. MRI in flexion showed anterior dural displacement and spinal cord contact, loss of posterior dural contact with posterior epidural flow voids due to venous plexus engorgement (Fig.3,c2).

Soft cervical collar and avoiding of flexion neck position were recommended. Patient informed us about increase of power in the left hand in 3 months.

Case 3
A 19-year-old male, Russian, had weakness of both hands and forearms, more expressed in the left hand. 15-year-old he noted weakness and wasting of the right arm. MRI at that time revealed lesion in C5-C6 levels of cervical spine. Weakness of the left hand acceded when he was 17-year-old.

Neurological examination revealed hypotrophy and severe weakness in hands and forearms. Upper limb reflexes were reduced. In the down limb reflexes were increased.

Cervical spine MRI in neutral position revealed flattening of the spinal cord and intramedullary hyperintensity at the levels C5-C7; cervical lordosis was normal and posterior epidural space was not increased. In flexion position anterior dura displacement was visualized, leading to the spinal cord contact against C5-Th1 vertebrae bodies and posterior epidural space enlargement with engorges venous plexus (Fig. 3,c3).

HD was diagnosed. Retrospectively details of anamnesis were cleared: in the last years patient spent a lot of time on the computer with flexion position of the neck. Soft cervical collar and avoiding of flexion neck position were recommended. Patient informed us about increase of power in the left hand in 3 months.

Case 4

A 30 y.o. male complaining about reduced left hand movement. The first signs of hand muscle hypotrophy were noticed at the age of 23 with further slow progression at the left forearm and arm levels.

Normal-position cervical spine MRI revealed abnormal cervical kyphosis and flattening spinal cord at the C5-C7 level with some intramedullary hyperintensity (anterior horns). MRI in flexion showed anterior dural displacement with posterior epidural flow voids due to venous plexus engorgement; spinal cord attachment to the posterior vertebral bodies surface (Fig.3,c4)

Case 5

A 20-year-old male, Russian, had weakness of both hands and forearms. 18-year-old he noted weakness and wasting of the left hand, in 6 months weakness of the right hand occurred. Neurological examination revealed hypotrophy and severe weakness in hands and forearms (left more than right).

Cervical spine MRI in neutral position revealed asymmetry flattening of the spinal cord and intramedullary hyperintensity at the levels C5-C7 (left more than right). In flexion position anterior dura displacement was visualized, leading to the spinal cord contact against C5-Th1 vertebrae bodies and posterior epidural space enlargement with engorges venous plexus (Fig. 3,c5).

Case 6
The only one female case, which is extremely rare. A 25 y.o. female with left upper limb weakness and hypotrophy. 16-year-old she noted weakness of the left arm, with further progression for the next 7 years.

Neurological examination revealed hypotrophy and severe weakness in left hand and forearm. Upper limb reflexes were reduced.

Cervical spine MRI in neutral position revealed flattening of the spinal cord and left-side intramedullary hyperintensity at the levels C5-C7; there was abnormal cervical kyphosis. In flexion position anterior dura displacement was visualized, leading to the spinal cord contact against C5-C5 vertebrae bodies and posterior epidural space enlargement with engorges venous plexus (Fig. 4).
Fig. 1: Oblique paresis of the left arm in case 1.

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Fig. 2: Healthy volunteer cervical spine MRI in a neutral and flexion positions (A, upper and lower rows) compared to signs of HD in neutral (upper B) and flexion (lower B) positions MRI in case 1. Sagittal and axial T2-weighted imaging.

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Fig. 3: Signs of HD in neutral (upper) and flexion (lower row) positions MRI in cases 2-5. Sagittal and axial T2-weighted imaging.

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**Fig. 4:** Signs of HD in 25 y.o. female (case 6). Sagittal (neutral and flexion positions) and axial T2-weighted imaging.

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The pathogenesis of HD is still debated. In a report of Hirayama et al\textsuperscript{2} there is a conclusion that dynamic spinal cord compression at neck flexion with forward displacement of the posterior dura is an unequivocal finding in the progressive stage of this disease. Furthermore, this finding is absent in elderly patients who have reached a stable stage in the progress of this disease. This observation suggests that dynamic compression of the spinal cord may be an important finding in the diagnosis of this disease and that a "tight" dural canal during flexion of the neck is due to a disproportional length between the vertebral column and the dural canal. The mechanism of myelopathy may involve ischemic changes or chronic trauma inflicted by repeated neck flexion.

HD is very rare condition in Russia, but it has common signs described by K. Hirayama and detected on MRI. Considering the importance of early diagnosis neurologists and MRI-specialists should be more informed about HD.

MRI of the cervical spine in neutral and flexion position is the way to confirm HD in clinically supposed case. There are signs of this pathology:

In neutral position

1. - abnormal cervical curvature (Fig.5),
2. - loss of attachment (LOA) between the posterior dural sac and subjacent lamina,
3. - localized lower cervical cord atrophy
4. - asymmetric spinal cord flattening and/or hyperintensity at the C5-T1 levels

In flexion images anterior shifting of the posterior dura and posterior epidural space enlargement can be visualized\textsuperscript{7}
Fig. 5: Signs of physiological cervical lordosis and abnormal cervical curvature

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References


