Hirayama disease

Hirayama disease, or Juvenile amyotrophy of the distal upper extremity (JADUE), is a type of cervical myelopathy related to flexion movements of the neck. It is considered a benign motor neurone disorder with a stationary stage after a progressive course.

It is associated with epidural venous engorgement and spinal canal narrowing. It has been unclear whether dural venous pressure influences spinal cord injury in this illness.

It was initially described Keizo Hirayama et al. in 1959 [1].

Epidemiology

It typically occurs mainly in young males between the ages of 15-25 years [2].

Pathology

Chronic microcirculatory changes in the territory of the anterior spinal artery induced by repeated or sustained flexion account for the necrosis of the anterior horns of the lower cervical cord, which is the hallmark of pathology [3].

Clinical features

Hirayama disease manifests as slowly progressive unilateral or bilateral hand weakness and atrophy, predominantly unilateral upper extremity weakness and atrophy, cold paresis, and no sensory or pyramidal tract involvement. It is also characterised by muscle weakness and atrophy in the hand and forearm with sparing of the brachioradialis, giving the characteristic appearance of oblique amyotrophy that affects the C7, C8 and T1 myotomes [4] [5]. The amyotrophy is unilateral in most patients, asymmetrically bilateral in some and rarely symmetric [6].

Diagnosis

In flexion MR studies, we can see the striking and pathognomonic picture of anterior shifting of posterior dura at the lower cervical spinal canal. In nonflexion studies, we find that asymmetric cord atrophy, especially at the lower cervical cord, though subtle, is highly suggestive of Hirayama disease. When it is seen, a flexion MR study is warranted to prove this diagnosis [7].

On myelograms and flexion-extension MR images, there can be a forward migration of the posterior

wall of the dura mater. The posterior epidural space becomes enlarged with flexion and is seen as a crescent of high signal intensity on T1- and T2-weighted MR images, with or without epidural flow voids. Uniform enhancement of this epidural space occurs with administration of contrast material.

At the site of maximal forward shift of the posterior dural sac the spinal cord is dynamically compressed with a reduction in the AP diameter of the cord compared to neutral imaging [8].

Differential diagnosis

Anterior cervical epidural spinal cysts should be considered in the differential diagnosis in patients who present with slowly progressive hand weakness [9].

Case series

2018

Spinal cord injury may cause cortical reconstruction. Wang et al., therefore, explored the changes in cortical activation before and after anterior cervical decompression and fusion surgery in patients with Hirayama disease (HD).

17 HD cases were performed anterior cervical approach for decompression and fusion surgery. BOLD-fMRI scan was performed preoperatively, 3 months, 6 months and 1 year after surgery. Activated voxels were compared between both hands after adjusting for head motion, slice timing, spatial normalization and image smoothing. Grip strength was also tested in both hands.

A retrospective review indicated that the grip strength of the asymptomatic hand was significantly stronger than the symptomatic hand at the time point before the surgery, three months after surgery, six months after surgery and one year after surgery ($p< 0.001$). The grip strength of both symptomatic and asymptomatic hands continuously increased within six months after surgery ($p<0.05$), but it stopped at one year after the surgery. The symptomatic limb tends to produce bilateral activation in M1 during motor tasks. Both contralateral and ipsilateral M1 activation were stronger in symptomatic hand tasks preoperatively ($p<0.05$). Both contralateral and ipsilateral activation in M1 during symptomatic hand tasks began to reduce after surgery and statistical significance was observed six months after surgery ($p<0.05$). Contralateral activation was relatively even over six months of the surgery($p>0.05$).

Following surgery pathological reconstruction may have occurred in the primary motor cortex. Recovery of motor function in the symptomatic limb was accompanied by decreased ipsilateral and contralateral M1 activation, as well as symptom improvement. These findings suggested that postoperative cortical activation changes may reflect functional recovery in HD [10].

2010

From May 2002 through December 2006, 6 young patients with cervical flexion myelopathy were seen in the Department of Neurosurgery at Chang Gung Memorial Hospital. The neurological and radiological findings in all 6 patients met the criteria for Hirayama disease. All patients had evidence

of a tight dural canal or forward migration of the posterior wall of the dural canal in dynamic MR imaging studies. Five patients were treated with surgical decompressive procedures (4 anterior and 1 posterior) and 1 patient received conservative treatment. Duration of follow-up ranged from 13 months to 4 years.

Motor function improved in 3 of 5 surgically treated patients and sensory function improved in 2. Neurological symptoms were unchanged in the conservatively treated patient. During follow-up MR imaging in the surgical group, anterior effacement during neck flexion was noted in 1 patient treated with a posterior approach.

Hirayama disease is so rare that it is easily misdiagnosed. Diagnosis is achieved via clinical presentation, neurophysiological examination, and neuroradiological imaging studies (dynamic MR imaging). The anterior decompressive approach may be better for patients showing anterior effacement and severe cervical kyphosis during neck flexion in MR imaging [11)].

Neuroradiologic examinations including myelography, CT myelography, and MRI in a fully flexed neck position were performed on 73 patients with this disorder and on 20 disease control subjects.

A distinctive finding in the disorder was forward displacement of the cervical dural sac and compressive flattening of the lower cervical cord during neck flexion. The forward displacement was significantly greater in patients with disease duration less than 10 years than in age-matched control subjects and patients in a late, nonprogressive stage.

Radiologic abnormalities of the lower cervical dural sac and spinal cord support the hypothesis that this disorder is a type of cervical myelopathy [12)].

Case reports

2012

A 34-year-old man presented with a 5-year history of slowly progressive hand and forearm weakness and atrophy. Nerve conduction studies demonstrated low median and ulnar motor amplitudes, and EMG demonstrated fibrillation potentials and long-duration, high-amplitude motor unit potentials in C6-T4-innervated muscles. MRI demonstrated a longitudinally extensive anterior spinal epidural cyst extending from C2 to L1. The patient had improved hand strength after surgery.

Anterior cervical epidural spinal cysts should be considered in the differential diagnosis in patients who present with slowly progressive hand weakness [13)].

2009

A previously healthy 19-year-old man presented with insidious onset of weakness in the left forearm and hand for the past year. On MRI, during neck flexion, the posterior dura showed anterior displacement that compressed the cervical spinal cord. The dura was opened linearly from C3 to C6, observing the herniation of the spinal cord through the opening. Duraplasty was performed in order to increase the room of the spinal cord. No spinal fusion was performed.

The postoperative course was uneventful. Clinical deterioration stopped following operation and two years later unchanged as compared to the preoperative one [14].

An 18-year-old man presented with profound hand weakness and atrophy that had developed over a period of 1 year. Electromyographic, magnetic resonance imaging, and angiographic findings are presented.

The epidural space was accessed using a microcatheter technique. Pressure measurements were recorded with and without Valsalva maneuver in the inferior vena cava, vertebral veins, and epidural space at C4 and C6 in both the flexion and neutral positions. Cervical epidural venous pressure measurements in flexion and neutral positions are presented. The patient underwent duraplasty with C4 to T1 laminectomies and fusion using lateral mass screws and facet arthrodeses.

Lack of significant pressure change with neck flexion suggested that dural venous engorgement is passive and not the direct cause for spinal cord injury. Data presented herein and review of the literature suggest that surgical treatment targeting the underlying pathophysiological mechanism in Hirayama disease can benefit patients, especially early in the course of the disease [15].

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