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The role of imaging in Hirayama disease

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In the article "imaging in Hirayama disease: A case series," the authors reported three cases of young Indian male patients with Hirayama disease (HD). They all presented the typical clinical features of the disease (i.e., gradual onset and progressive course of unilateral or asymmetric muscular weakness and atrophy in the hand, and ulnar side of the forearm muscles, without sensory or pyramidal signs).[1]

Moreover, sagittal magnetic resonance imaging (MRI) scans of cervical spine in neutral position showed focal area of cord atrophy and during neck flexion, an anterior displacement of the posterior dural wall was present with enlargement of the posterior epidural space, which showed contrast-enhancement. After the diagnosis, all patients were conservatively treated with a favorable outcome in two out of three.

Through the description of these cases, the authors pointed out the importance of MRI features in the diagnosis of the disease, underlying the challenge for neurologists and neuroradiologists to early identify this condition.

HD was described in 1959 by Hirayama as a rare, self-limiting juvenile spinal muscular atrophy of the distal upper limbs. The disease predominantly affects young male adults of Asian origin and is characterized by asymmetric motor dysfunctions that may initially be misinterpreted as an atypical form of amyotrophic lateral sclerosis.[2]

In HD, symptoms generally progress for 3–4 years after the onset and are followed by a stationary stage; however, the early arrest of the progression is essential for any possibility of improvement.

The pathogenesis of the disease seems to be due to the disproportionate growth of the vertebral column compared to the spinal cord and the following cervical cord compression during neck flexion that induce microcirculatory disturbances responsible of ischemic necrosis of the anterior horn cells, as demonstrated by autoptic findings.[2]

The presence of classically described MRI features unequivocally confirms the diagnosis of HD.

It has been demonstrated that in neutral neck position MRI, the detection of segmental, inferior cervical spinal cord atrophy has a diagnostic value with the sensitivity of 59% and the specificity of 100%.[3] Furthermore, a "sand-watch" – like pattern (i.e., the fact of localized atrophy being between regions of preserved cord architecture) on sagittal views, should lead to further investigations.[4]

The diagnosis of HD is straightforward at flexion MRI studies, which often reveals posterior dural detachment from lamina with anterior shift causing spinal cord compression and enlarged posterior epidural space with congested epidural veins. The posterior epidural space is a crescent-shaped space that appears isointense in T1-weighted and hyperintense in T2-weighted images. Uniform contrast-enhancement of this space has been described.[3] Moreover, the venous engorgement of the posterior epidural plexus during active flexion of the cervical spine has

been angiographically proven.[5]

Conventional MRI may underestimate the extent of cord tissue damage, as demonstrated by Gallo *et al*., who used nonconventional MRI techniques to assess a patient with HD. Using magnetization transfer and diffusion-weighted MRI of the cervical cord, they found that cord damage in HD extends beyond that seen on routine MR imaging scans. In fact, as a consequence of the chronic ischemic damage in the anterior spinal artery territory, also the corticospinal tracts can be damaged because of their location in the border zone, but remains asymptomatic thanks to the brain cortical reorganization.[6]

HD can be treated either conservatively or nonconservatively, and MRI has a crucial role also in the treatment decision. The application of a cervical collar can minimize neck flexion and prevent the progression of muscle weakness and atrophy in the early stages, while surgical decompressive treatment should be applied only to patients with progressive worsening despite conservative treatment, whose radiological findings include compression of the middle and lower cervical cord against the vertebral body, narrowing of the ventral subarachnoid space, and a severe forward shift of the posterior wall of dura during neck flexion.[7,8]

The evidence within this article strongly supports the idea that an early diagnosis and a prompt therapeutic intervention of HD patients may induce a premature arrest of symptoms progression. The disease-specific features on MRI in neutral position, using both conventional and nonconventional techniques, can help to exclude differential diagnosis (such as compressive myelopathy, radiculopathy, and intrinsic cord pathology) and reveal changes suggestive of the disease that can subsequently be confirmed on dynamic fully-flexed acquisition MRI.

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