Hirayama Disease: Imaging Profile of Three Cases Emphasizing the Role of Flexion MRI

ABSTRACT

We report three cases of Hirayama disease, cervical flexion myelopathy presenting as unilateral or bilateral asymmetric muscular atrophy of forearm and hand involving C7 - T1 myotomes in young males. MRI revealed asymmetric cord atrophy, altered signal intensity of cord, posterior dural detachment and enlarged posterior epidural space with multiple flow voids. This article emphasizes the role of Flexion MRI in diagnosing Hirayama disease.

CASE REPORT

Case 1

A 30-year-old male presented with weakness of both the upper limbs. On examination, there was wasting and reduced power of both forearms and small muscles of hand with more severe involvement of the left upper limb. There was no sensory system involvement. Routine MRI of the cervical spine was unremarkable except for subtle cord atrophy at C6 to C7 vertebral levels and posterior dural detachment of more than 33% from the lamina [Table/Fig-1a,b]. Considering the clinical features and imaging findings, possibility of Hirayama disease was suspected. Flexion MRI was performed which showed extensive detachment of posterior dura from lamina, its anterior displacement causing enlarged posterior epidural space and engorged epidural veins. The spinal cord was compressed against the posterior margin of vertebral body [Table/Fig-1c,d].

Case 2

A 25-year-old male presented with reduced bulk of left hand muscles for six months associated with resting tremor and reduced grip. On examination, there was wasting and reduced power of small muscles of hand. No sensory system involvement was noted. Routine MRI of the cervical spine showed mild kyphosis of cervical spine, mild asymmetric cord flattening and atrophy. There was evidence of mild posterior dural detachment from the lamina extending from C4 to C6 vertebral levels [Table/Fig-2a,b]. Flexion MRI revealed anterior displacement of the cord with obliteration of anterior subarachnoid space, enlarged posterior epidural space with engorged epidural veins and compression of spinal cord against vertebral body [Table/Fig-2c,d].

Case

A 28-year-old male referred as anterior spinal muscular atrophy presented with weakness of right forearm and hand for five years. He had history of recent onset of weakness of left upper limb. His weakness was accentuated on exposure to cold. No evidence of sensory loss was noted. Routine MRI of cervical spine showed kyphotic curvature of spine, cord atrophy from C4 to C7 vertebral levels and linear hyperintensities of the cord in the region of anterior horn cells on T2 weighted images which appeared as an Owl's eye in axial section [Table/Fig-3a,b]. Considering the age, pattern of involvement and anterior horn cell involvement flexion MRI was performed to rule out Hirayama disease. Flexion MRI revealed detachment of posterior dura from the lamina compressing the spinal cord against vertebral body, enlarged posterior epidural space with engorged epidural veins [Table/Fig-3c,d].
Hirayama disease is a frequently undiagnosed condition with simple and inexpensive treatment. Clinicians must have a high index of suspicion of Hirayama disease when a young male presents with distal upper extremity weakness. Dynamic flexion MRI is the investigation modality of choice which reveals the typical features of the disease and clinches the diagnosis.

**REFERENCES**