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Predominant proximal upper extremity involvement in Hirayama disease

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Abstract

Hirayama disease usually selectively involves lower cervical myotomes (C8, T1). Thus, patients usually manifest with atrophy and weakness of small hand muscle. Predominant isolated involvement of proximal arm is rarely reported in Hirayama disease. Here, we report a case of Hirayama disease who had focal weakness and wasting, mainly confined to right biceps brachii muscle, with prominent shifting of dural sac in C4-5 segment by dynamic flexion magnetic resonance imaging (dfMRI), which may explain this unusual distribution of the disease.

INTRODUCTION

Hirayama disease[1], also known as "juvenile muscular atrophy of distal extremity", occurs more frequently in young men and is characterized by insidious asymmetrical upper extremities weakness and muscular atrophy.[2] The pathologic lesion is in the anterior horn cells of the spinal cord, most commonly at C7 and T1.[3] Some patients also manifest increased weakness during cold (cold paresis) and hand tremor.[4-6] A genetic contribution to the disease has also been suggested.[7] Some of the characteristics of this disease remains unexplained, including the male predominance.

Mechanism of Hirayama disease remains uncertain, but some authors have proposed an ischemic myelopathy as underlying pathogenesis, from anterior shift of cervical dural sac which subsequently lead to compression to anterior compartment of cervical cord. Recent technique of dynamic flexion magnetic resonance imaging (dfMRI) give support to this hypothesis.[5] Because the pathology usually occurs in the lower cervical segments (C8, T1 myotomes) of the spinal cord, patients with Hirayama disease usually present clinically with severe atrophy of intrinsic hand muscles. Thus, the main differential diagnosis clinically are ulnar neuropathy or cervical radiculopathy. Recently, we saw a Hirayama disease patient with focal atrophy of C5 segment, who clinically presented with a selective involvement of biceps brachii muscle. We report his clinical and radiological features here.

CASE REPORT

A 19-year-old man presented with progressive weakness and atrophy of right upper arm. He began to notice atrophy, confined only to his right biceps brachii, more than one year ago. He noted weakness of his arm flexion in the last 6 months. There was no notable event or systemic illness, such as infection, vaccine or surgery preceding the onset of the muscle atrophy and weakness, or a similar family history. He also recalled having involuntary muscle twitching of his right arm, which could be due to muscle fasciculation. He also complained of numbness and paresthesia in the right lateral arm, forearm, and thumb over the same period.

On neurological examination, the muscle power was 3/5 for the right elbow flexion with associated mild atrophy in right upper arm (Figure 1). The circumference of his right arm was much smaller than that of the left arm: 25 cm versus 32 cm. The intrinsic hand or forearm muscles were normal. Despite the subjective sensory complaints, sensory examinations were intact. The biceps and brachioradialis jerks of the right arm were absent and triceps jerk was decreased. Motor and sensory examination of the other extremities was normal. The various blood laboratory tests done were normal. Nerve conduction studies showed normal findings except mild prolongation of distal motor latencies in right median and ulnar nerves. Electromyography showed moderate increase of spontaneous activities in right biceps and deltoid muscle. The

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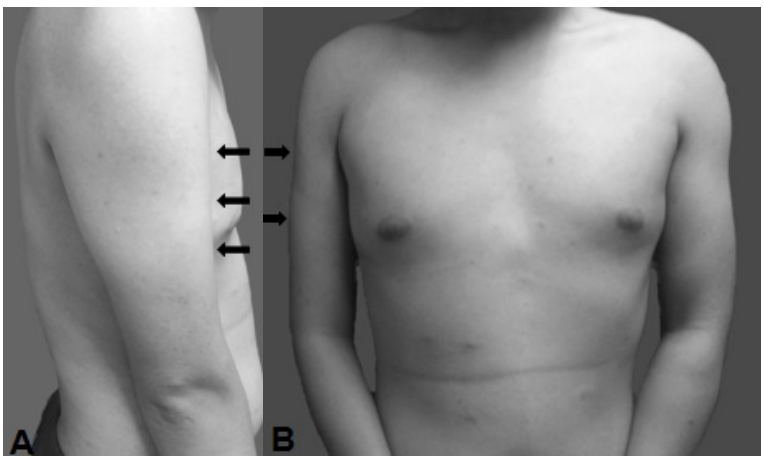

Figure 1: Photograph of the patient showing wasting in the right arm. The right lateral view (A) and front view (B) shows a remarkably asymmetrical atrophy in the right biceps brachii than that of the left. The circumferential length showed taken at the level indicated by the arrow showed a difference of 7 cm between the two sides (25 cm versus 32 cm)

right intrinsic hand muscles, including abductor pollicis brevis and first dorsal interossei, also showed mild increase of spontaneous activity. Electromyography of the left arm was normal.

Cervical MRI showed reversed curvature, focal cord atrophy, and detachment of posterior dura at C4 and C5 level (Figure 2A). An asymmetrical atrophy of the spinal cord and loss of attachment of the posterior dural sac (Figure 2B) were also seen. We performed dfMRI according to a previously suggested technique.[5,6] The dfMRI showed prominent anterior shift of dural sac and compression of cervical cord against vertebral body, maximally at the level of C4 and C5 (Figure 2C). We diagnosed the patient as having Hirayama disease with atypical manifestation of predominant C5 involvement.

The patient was treated with a supportive neck brace and bed rest. During follow-up, his sensory symptoms improved slightly. However, there was no improvement of motor weakness or muscle atrophy.

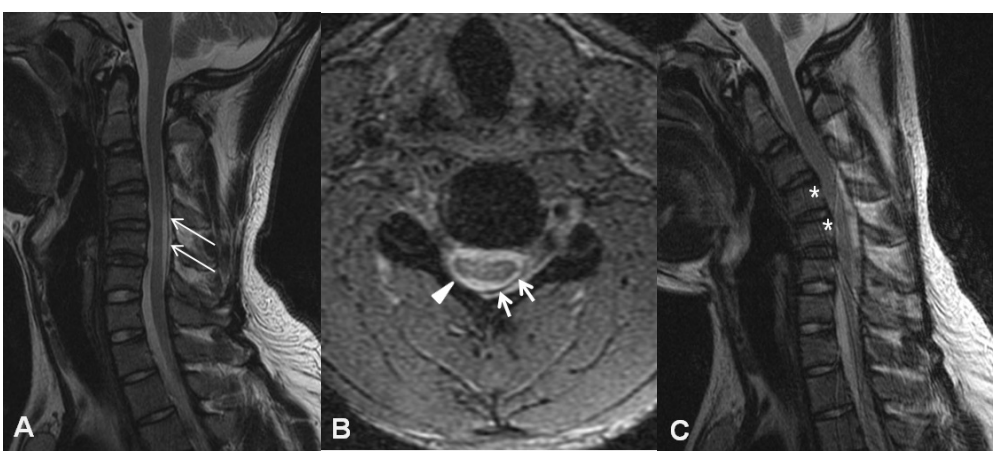

Figure 2: A. T2-weightedsagittal MRI taken in neutral position of the neck shows reversed curvature, focal cord atrophy and slight signal change of the spinal cord at C4 and C5 level. There is detachment of posterior dura shown by thin arrows. B. An axial gradient MRI at the level of C5 demonstrates an asymmetric atrophy of the spinal cord (arrowhead) and a loss of attachment of the posterior dural sac (thick arrows). Atrophy at the spinal cord is more in the right, whereas detachment at the dural sac is more in the left. C. T2 weighted sagittal MRI, acquired during neck flexion, shows exaggeration of detachment, anterior shift of posterior dural sac and compression of cord against the posterior border of vertebral body, also maximally in C4, C5 level (asterisks).

DISCUSSION

Hirayama disease is a benign self-limited disorder predominantly seen in young adults.[8] It was first reported by Hirayama *et al*.[1] Hirayama disease was known to usually involve lower cervical segments and manifests with atrophy, weakness of muscle in the distal portion of the upper arm. The patients may also complained of cold paresis and tremor in the hands muscles.[6] Hirayama disease often initially involves one upper limb[2], although it may progress to involve symmetrically both upper limbs.[9] Some authors, mainly from Asia, have reported rarepatients of proximal arm involvement[5,10], as is also the case in this report.

The recently proposed mechanisms of Hirayama disease is based on dfMRI studies. The hypothesis proposed is that repeated anterior shift of posterior dural sac compress the anterior part of the cervical cord resulting in ischemic myelopathy.[5,6] The juvenile onset of Hirayama disease was explained by the disproportionate growth between the vertebral column and the contents of the spinal canal, especially the dural sac during the juvenile growth spurt.[4,11] The asymmetric involvement of Hirayama disease was explained by the "posterior epidural ligament factor" with two separate posterior epidural ligaments being responsible for the adhesion of posterior dura matter to the ligamentum flavum. There was uneven distribution across spinal segments that may explain the asymmetric involvement.[2,12]

The explanation of dominant involvement of C8 and T1 myotomes is thought to be related to the maximal neck flexion in the lower cervical segments. The anterior shift of dural sac and subsequent compression of cord against the vertebral body is maximal in this regions.[4] Our patient had an anomaly of bony spine, which is not uncommon in Hirayama disease.[6] In our patient, the MRI in the neutral neck position showed maximal lordosis at C4-C5. Hence our patient may have maximal intramedullary pressure by anterior shifting of dural sac at the C4-C5, rather than C8-T1 levels.

In conclusion, we present a case that highlights that Hirayama disease can occur in higher cervical level and manifest as upper arm weakness.

DISCLOSURE

Conflicts of interest: None

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