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Clinical short communication

Are basketball players more likely to develop Hirayama disease?

Corrado Cabona[a,*], Alessandro Beronio[b], Ilaria Martinelli[c], Chiara Briani[c], Caterina Lapucci[a], Carlo Serrati[d], Stefano Delucchi[e], Antonio Mannironi[b], Luana Benedetti[a]

[a] Department of Neuroscience, Rehabilitation, Ophthalmology, Genetics, Maternal and Child Health (DINOGMI), University of Genova and IRCCS, Policlinico San Martino, Genova, Italy
[b] Department of Neurology, Osp. S. Andrea, La Spezia, Italy
[c] Department of Neurosciences, University of Padova, Italy
[d] Department of Neuroscience, Ospedale Policlinico San Martino, Genova, Italy
[e] Department of Neuroradiology, Osp. S. Andrea, La Spezia, Italy

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ABSTRACT

Hirayama disease is a rare neurological disease affecting primarily men in the second to third decades. To date there are only few reports from Italy.

We report the case of three young basketball players who presented with clinical, electrophysiological and MRI findings suggestive for Hirayama disease.

Although the pathophysiology of the disease is still unknown, several hypotheses have been suggested and two of these are the disproportionate growth of cervical spine and cervical cord/roots during adolescence and the chronic traumatism.

We think that, in our cases, the height of basketball players combined with the constant stress caused by the sport, could have contributed to the development of the Hirayama disease.

With this report we would stress the importance to be careful to consider this pathology in order to avoid misdiagnosis predictive of poor prognosis in young patients.

1. Introduction

Hirayama disease is a rare neurological disease characterized by insidious unilateral or bilateral muscular atrophy and weakness of the forearms and hands, without sensory or pyramidal signs [1]. The disease affects primarily young men, progresses for a mean of 4–5 years and spontaneously arrests several years after the onset. Several cases have been reported from Asian countries [1,2], but only few from Italy [3–9].

We report on three young basketball players with Hirayama disease.

1.1. Patient 1

A 17-years-old basketball player, 1.95 m tall, came to our attention for progressive hyposthenia and hypotrophy of the intrinsic muscles of the left hand. Weakness worsened during exposure to cold temperature. Neurological examination revealed weakness of interosseous, abductor pollicis brevis and flexors and extensors of the fingers muscles without pyramidal and sensory signs.

1.2. Patient 2

A 16.years-old basketball player, 1.98 m tall, presented with a 5-months history of weakness and hypotrophy of the distal portion of the left upper limb. Neurological exam evidenced weakness of left interosseous, abductor digiti minimi and opponens pollicis muscles without pyramidal signs or sensory symptoms. A fascicular twitching of fingers in the extended position was present.

1.3. Patient 3

A 16-years-old basketball player, 1.97 m tall, complained of 4-months history of slowly progressive hyposthenia and hypotrophy of the last two fingers of the left hand, symptoms more evident at cold

* Corresponding author.
E-mail address: corrado.cabona@gmail.com (C. Cabona).

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Table 1
Clinical, neurophysiological and MRI characteristics of the patients.

Features	Patient 1	Patient 2	Patient 3
Age	17 years	16 years	16 years
Sex	Male	Male	Male
Height	1.95 m	1.98 m	1.97 m
Affected side	Left	Left	Left
Electrodiagnostic evaluation	EMG: a chronic and active (fibrillation potentials) neurogenic pattern at C7-C8-T1 myotomes*. NCS: a low amplitude of cMAP of ulnar nerve with a low persistence of the F wave to ulnar and median nerves. SEP: normal.	EMG: signs of acute and chronic denervation (fibrillation, positive sharp waves, large amplitude, polyphasic potentials) at C7-C8-T1 myotomes*. NCS: low amplitude of ulnar cMAP without conduction blocks and absent F wave to ulnar nerve. Median nerve normal SEP: normal.	EMG: signs of chronic denervation (polyphasic potentials, large amplitude) at C7-C8 myotomes*. NCS: ulnar cMAP with low amplitude. No conduction blocks detected. Median nerve normal SEP: not done.
Dynamic cervical MRI	Localized lower cervical cord atrophy, asymmetric cord flattening, abnormal cervical curvature and loss of attachment between the posterior dural sac and subjacent lamina in neutral position, forward migration of the wall of the dura mater with a hyperintense enlarged posterior epidural space showing curvilinear flow voids and uniform enhancement after administration of contrast in fully flexed position		
Other diagnostic tools	Brachial plexus ultrasound and MR: negative	Anti gangliosides antibodies: negative	Anti-gangliosides antibodies: negative
Follow-up	28 months from symptoms onset. Progressive course for 11 months, then clinical stability.	15 months from symptoms onset. Steady state reached in 13 months.	12 months from symptoms onset. Progressive course for 11 months, then clinical stability.

EMG: Electromyography; NCS: Nerve Conduction Study; SEP: Somatosensory Evoked Potential; cMAP: compound muscle action potential; *EMG altered muscles: first dorsal interosseous, abductor pollicisbrevis, extensor digitorum, triceps brachii (C7-C8-T1 myotomes).

temperature. No fasciculations, cramps or sensory abnormalities were present. Neurological examination revealed weakness of extensors and flexors carpi, extensors and flexors of fingers, long and brief abductors pollicis, opponent pollicis, interosseum, abductor of V finger. Deep reflexes were preserved.

All the patients underwent nerve conduction studies and electromyography (Table 1). Dynamic cervical MRI revealed localized lower cervical cord atrophy, asymmetric cord flattening, abnormal cervical curvature and loss of attachment between the posterior dural sac and subjacent lamina in neutral position and forward migration of the wall of the dura mater (with a hyperintense enlarged posterior epidural space showing curvilinear flow voids and uniform enhancement after administration of contrast) in fully flexed position (Fig. 1). The diagnosis of Hirayama disease was made. All patients discontinued basketball and underwent physiotherapy; two of them also used cervical collar. After a mean of 12 months of slow progression from the onset, the deficits became stable.

2. Discussion

Slowly progressive upper limbs distal weakness and hypotrophy is a possible manifestation of both multifocal motor neuropathy and amyotrophic lateral sclerosis, but fortunately in our patients the clinical and electrophysiological patterns fulfilled almost all Hirayama disease diagnostic criteria [10].

MRI findings (in dynamic acquisition) supported the diagnosis. Indeed cervical MRI in neutral and flexed position is now recommended for the diagnosis and asymmetrical atrophy of the lower cervical segments is the most prominent abnormality associated with forward displacement of dural sac and flattening of the lower cervical cord in a fully flexed position of the neck [11].

The majority of the described Hirayama patients are form Asia [1,2], but cases also from America and Europe have been described [3–9].

Although in the literature there are reported cases of Hirayama disease in amateur athletes, this is the first case series in agonists basketball players [12–14].

The etiology is unknown but it has been attributed to several factors, mainly trauma and ischemia [1,15]. A possible mechanism of lower cervical anterior horn damage may be a chronic circulatory insufficiency or chronic trauma induced by repeated or sustained neck flexion. Kikuchi et al. proposed that an imbalanced growth causes disproportion in length of the vertebral column and spinal canal contents, resulting in a “tight dural sac.” As a consequence, a short dura cannot compensate for the increased length in flexion and so it is displaced anteriorly, with resultant spinal cord compression [16], phenomenon that could be accentuated during juvenile growth spurt [17].

Our patients' height could be related to the disproportion in the lengths of the vertebral column and the spinal canal contents, potentially constituting a risk factor for the onset of the disease, whereas basketball might have contributed to sustained trauma and neck flection, favoring its progression. Interestingly, as previously reported [12], also in two of these patients a growth of 15–20 cm has been reported in the 2–3 years preceding the onset of Hirayama disease, supporting the role of the strong growth in a short time as an additional risk factor.

Considering the short duration of the illness and the repeated and sustained neck flection of the patients, in accord with published data [18], a conservative approach with cervical collar was tried with benefit (arrest of disease progression).

3. Conclusion

Hirayama disease is a rare neurological disease, affecting primarily young Asian patients, that may be overlooked in western countries. It presents a diagnostic challenge, especially in the differential diagnosis with motor neuron disease.

Hirayama disease should then be considered in front of young tall patients exposed to neck chronic trauma also because an early diagnosis is crucial for a proper therapy and behavioral changes.

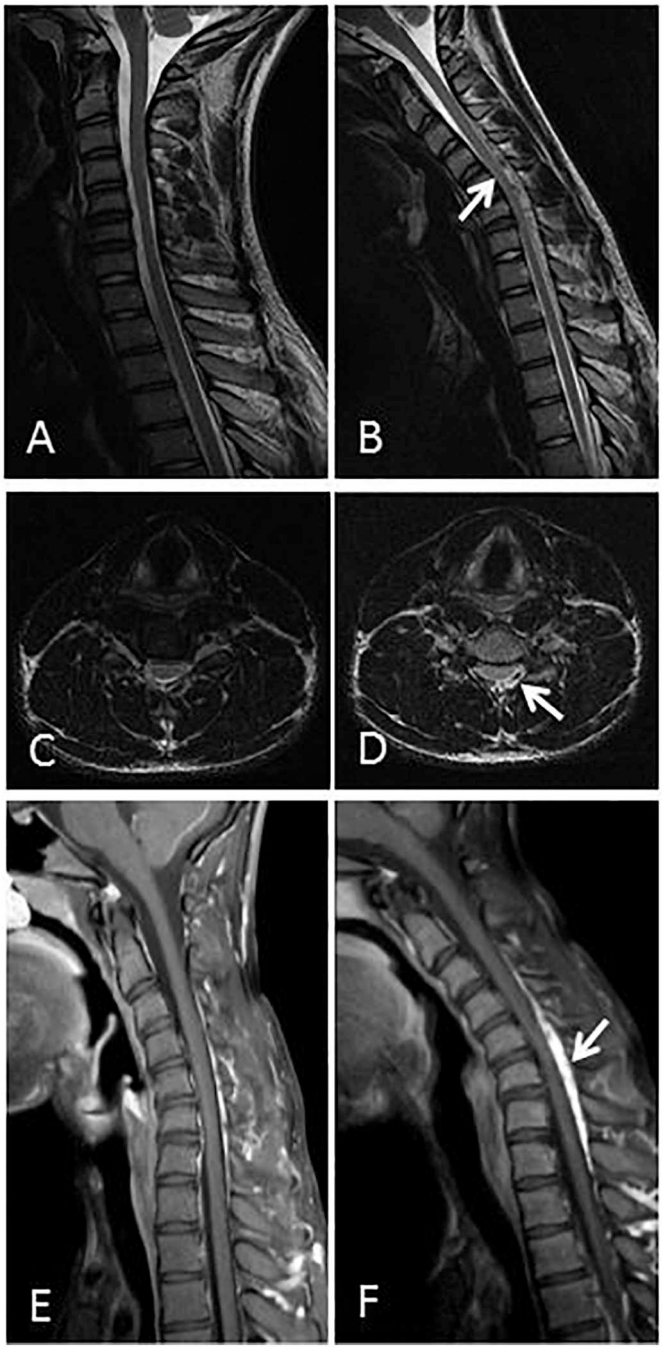

Fig. 1. Spinal MRI in neutral position and in hyperflection. Fig. A and B (T2w sagittal sections in neutral and flexion position) and Fig. C and D (T2w axial sections in neutral and flexion position) evidence the loss of attachment between the posterior dural sac and subjacent lamina in neutral position and forward migration of the wall of the dura mater in hyperflection (see the arrows). Fig. E and F (pre and post-contrast T1w sagittal sections) show a hyperintense enlarged posterior epidural space with a curvilinear flow voids and a uniform enhancement after administration of contrast in fully flexed position (see the arrow).

Conflict of interest

The authors report no conflict of interest.

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