SAJEM EMERGENCY MEDICINE JOURNAL

Case Report

Hirayama disease- A case report

Salman Mansoor[1], Kevin Murphy[2], Mohammad Hijaz Adenan[3], Eimear Joyce[4], Orla Hardiman[5], Michael Hennessey[6], Siobhan Kelly[7]

Authors Affiliation

Sligo University Hospital, Sligo, Ireland[1,2,3,4,7]

National Neuroscience Centre, Trinity College Dublin, Ireland[5]

University Hospital Galway, Galway, Ireland[6]

Correspondence to

Salman Mansoor
salmanmansoor.dr@gmail.com

ABSTRACT

Hirayama disease is a rare neurological entity which can present in any setting emergency departments included. These cases are usually diagnosed after carefully excluding other conditions. We report the case of a 23-year old man who presented with an 18-month history of numbness, tingling and painless curling of the 4th and 5th digits. The symptoms did not progress beyond the 12-month period. The right side and bulbar function remained unaffected throughout. Based on the age of onset, arrest of progression after a period of deterioration, imaging features and neurophysiological findings a diagnosis of Hirayama Disease was made.

KEY WORDS

Hirayama Disease, Monomelic Amyotrophy, Motor Neuron Disease

BACKGROUND

Hirayama disease is a rare condition which is sometimes considered a variant of anterior horn disease. (1) The mechanism has been postulated to be due to a dynamic anterior compression of the posterior internal vertebral venous plexus resulting in hypoxic damage to the vulnerable anterior horn cells. (2) Intraspinal lesions (e.g., syringomyelia, syringobulbia, or tumor) can present with symptoms that can mimic Hirayama disease.

CASE SUMMARY

We report the case of a 23-year old Irish man who presented with an 18- month history of numbness, tingling and painless curling of the 4th and 5th digits on the left hand. He found it difficult to hold heavy items in his right hand, however there was no difficulty in opening jars.

His birth history was complicated due to a twin pregnancy. He was preterm born at 28 weeks via C-section and developed a post-partum intra-cerebral hemorrhage. He had delayed milestones with single word speech at 2 years and walking at 2.5 years. He managed to complete his leaving certificate with special needs assistance. His history was noncontributory for any neurological conditions.

He was in good state of health without any functional impairment or residual weakness before his symptoms started, 18 months back. He was referred by his General physician for a neurologist opinion to further investigate the true cause for his presentation.

Pertinent findings on his examination were: atrophy of the left upper limb extending from the shoulder, arm and forearm. He had marked atrophy in the intrinsic hand muscles affecting the thenar, hypothenar eminences and 1st dorsal interosseous and clawed 4th and 5th fingers. There was weakness of wrist flexion, finger extension and finger flexion. A mild tremor in the left thumb was also noted. There was no winging of scapula or any discernible fasciculations. Reflexes were pathologically brisk in the left upper and lower limb with bilaterally down going plantar. His symptoms progressed over a period of 12 months and he developed extensive left upper limb and left pectoral muscle wasting.

Baseline blood tests, Autoimmune Screen and metabolic profile were within normal ranges. MRI of his cervical spine showed thinning of the cord at C6-7 level and flexion and extension of neck didn't reveal significant compression as shown in Figure 1. Nerve conduction studies showed unilateral marked prolonged latency of median and ulnar F- waves on the left with mild slowing in motor conduction velocities in ulnar and median nerves as shown in Table 1 and 2. EMG showed chronic partial denervation in Extensor digitorum, left first dorsal interosseous (FDIO) and Left biceps with some occasional fasciculation in FDIO on the left only.

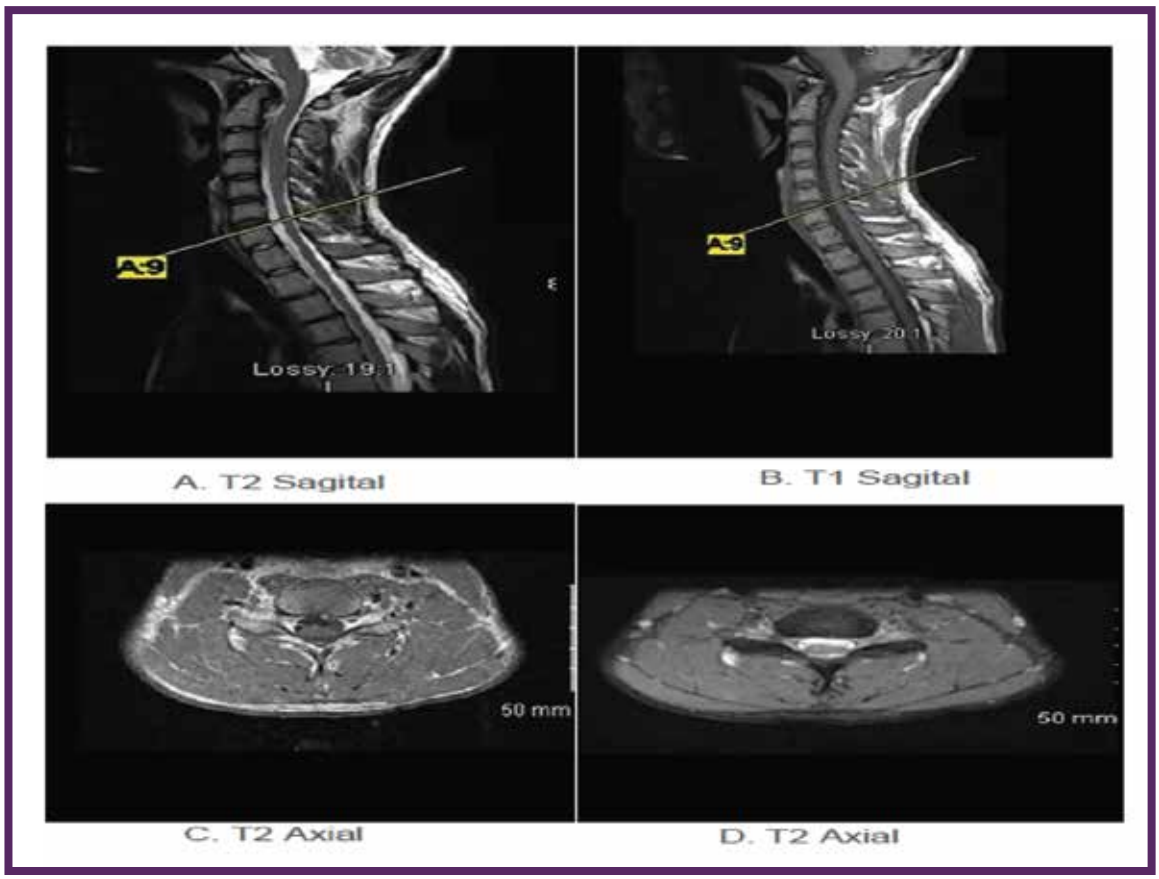

Figure 1: MRI of cervical spine

Left Median Motor	Latency (ms)	Amplitude mV	Conduction Velocity m/s
Wrist-APB	4.73	1.92	--
Pos. 2-Wrist	10.3	2.0	43.1
Elbow-Bl. elbow	13.2	3.8	--
Axilla-Ab. elbow	16.0	2.6	--

Table 1: Nerve Conduction study Left Median Motor Nerve

Left Median Motor	Latency (ms)	Amplitude mV	Conduction Velocity m/s
Wrist-ADM	3.21	2.5	--
Bl. Elbow -Wrist	7.23	2.2	52.2
Ab. Elbow -Bl. elbow	9.14	2.1	52.4
Axilla-Ab. Elbow	11.4	1.88	--
Pos. 5-Axilla	14.6	1.82	--

Table 2: Left Ulnar Nerve Motor Nerve Conduction Study

DISCUSSION

Based on the age of onset, arrest of progression after a period of deterioration, imaging features and neurophysiological findings a diagnosis of Hirayama disease was made. Other diagnostic possibilities were carefully excluded including the structural and functional causes.

It may be that these patients are predisposed by imbalanced growth in their vertebral column compared to their spinal. A clinical pattern of isolated arm (bilateral or unilateral) atrophy over several years, often with subsequent plateauing is classically seen. It is therefore considered a relatively benign condition which may be treated by limiting neck flexion. [3] It is rare however and importantly needs delineation from motor neuron disease with its graver prognosis and other, potentially treatable conditions on this list. Electromyography (EMG) and flexion MRI cervical spine can help make the diagnosis but follow up to ensure that it is not the early stages of MND are time dependent.

His symptoms have remained static after the initial deterioration for 12 months presenting to the neurology outpatient department.

CONCLUSION

Our case highlights the importance of considering Hirayama disease as a diagnostic possibility in young men. Although it is a self-limiting condition but early detection may help in managing the condition with a neck collar which might halt progression. [4] It should be considered especially when the symptoms and clinical signs present in young males involving upper limbs, asymmetrical, and plateau after a deteriorating initial phase.

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