

'Snake-Eye' Appearance in Cervical Cord Posterior Horn in Adult-Onset Spastic Paraparesis: Rare Observation

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Abstract

Adult-onset progressive spastic paraparesis occurs commonly due to spinal cord pathology. Here, we report a middle-aged man who presented with adult-onset progressive spastic paraparesis of 10 year duration and MRI showed 'snake-eye' appearance in cervical cord posterior horn region. The probable cause for spastic paraparesis was sporadic Hereditary Spastic Paraparesis (HSP).

Introduction

Adult-onset progressive spastic paraparesis occurs commonly due to spinal cord pathology. It is characterised by bilateral upper motor neuron signs, sensory deficits, and bladder/bowel/sexual dysfunction that evolves over a few months to years. The clinical symptoms and signs are due to the interruption of ascending sensory and descending motor pathways in the spinal cord [1]. The lesions on the Magnetic Resonance Imaging (MRI) may be extradural; intradural/extramedullary; intramedullary; intramedullary/tract-specific; spinal cord atrophy; or normal-appearing spinal cord. Here, we report a middle-aged man who presented with progressive spastic paraparesis of 10 year duration and MRI showed 'snake-eye' appearance in cervical cord posterior horn region. The probable cause for spastic paraparesis was sporadic Hereditary Spastic Paraparesis (HSP).

Case Report

A 42-year-old man presented with history of difficulty in walking of 10 year duration. The first symptom noticed by patient was that he used to trip and fall frequently with injuries over toes. Gradually, he noticed that his walking speed has come down with feeling of tightness in both lower limbs. The symptom slowly progressed over 10 years and was still ambulating with one cane support at the time of presentation to us. He had urinary frequency and urgency. There was no sensory symptoms, upper limb or cranial nerve symptoms. His cognition was normal. His perinatal history was unremarkable. There were no similar complaints in his family. He did not consume cassava or chicken peas. On examination, higher mental functions were normal. Speech and cranial nerves were normal. Motor examination showed normal tone and power with mild hyperreflexia in upper limbs. There was spasticity in both lower limbs with hyperreflexia and ill-sustained ankle clonus. Jaw jerk was not elicitable but abdominal reflex was preserved. Sensory examination was normal with no cerebellar signs. Plantar response was extensor on both sides. His gait was spastic requiring one cane support to ambulate. Complete hemogram, renal, hepatic and thyroid functions were normal. Serological testing for Human Immunodeficiency Virus (HIV) and venereal disease research laboratory was non-reactive. Cervical spine MRI showed T2 hyperintense signal changes in posterior horn region resembling 'snake-eye' on axial sections and two linear hyperintensity on coronal sections (Figure 1). Brain MRI was normal. Cervical spine X-ray did not reveal cranio-vertebral junction abnormalities. Nerve conduction studies were normal. Median nerve Somatosensory Evoked Potentials (SSEP) was normal; however waveforms were absent in the posterior tibial SSEP. Cerebrospinal fluid (CSF) examination was within normal limits. Vitamin B 12 and E, copper and tandem mass spectroscopy were normal. Serum/ CSF human T lymphotropic virus type 1 (HTLV-1) antibodies level assessment and molecular genetics for HSP could not be done due to non-availability. A diagnosis of adult-onset spastic paraparesis due to sporadic hereditary spastic paraparesis was made.

Discussion

Adult-onset progressive spastic paraparesis is due to a large number of causes and often poses a diagnostic challenge to the clinicians. The multitude of aetiologies of adult-onset progressive spastic paraparesis can be classified into six patterns based on the MRI abnormalities. The six patterns include: extradural; intradural/extramedullary; intramedullary; intramedullary/tract-specific;

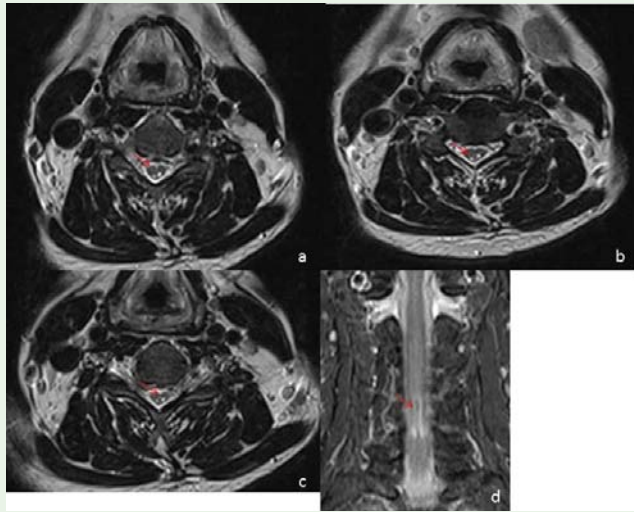


Figure 1: Cervical spine MRI T2 axial view (a), (b) & (c) shows hyperintense signal changes in posterior horn region resembling 'snake-eye' (red arrow); (d) two linear hyperintensity on coronal sections (red arrow).

spinal cord atrophy; or normal-appearing spinal cord. The extradural aetiologies include tumors/metastases, spondylotic myelopathy, metabolic/toxic like fluorosis, Paget's disease, congenital spinal anomalies, infection like tuberculosis, syphilis, and immune-mediated like rheumatoid arthritis. The intradural/extramedullary aetiologies include pachymeningitis, tumors/metastases, superficial siderosis, congenital cysts like neuroenteric, arachnoid and epidermoid cysts, vascular like spinal Arterio-Venous Malformation (AVM), infection like tuberculosis, syphilis, and immune-mediated like sarcoidosis. The intramedullary aetiologies include immune mediated like sarcoidosis, paraneoplastic; tumor/metastases like ependymoma, astrocytoma; infection like tuberculosis, syphilis; vascular like AVM, congenital cysts like arachnoid and epidermoid cysts; syringomyelia; and toxic/metabolic like vitamin B12 deficiency, radiation induced. The intramedullary/tract-specific aetiologies include immune mediated like paraneoplastic; infection like HIV, toxic/metabolic like vitamin B12 and E deficiency; and congenital like homocysteine remethylation defects, hypomyelination with brainstem and spinal cord involvement and leg spasticity. The spinal cord atrophy aetiologies include immune mediated like sarcoidosis, lupus, multiple sclerosis; infection like HIV, HTLV; toxic like radiation myelopathy; and congenital like HSP, adrenomyeloneuropathy, adrenolukodystrophy. The Normal appearing spinal cord aetiologies include immune mediated like paraneoplastic; infection like HTLV; congenital like HSP, phenylketonuria, Krabbe disease and toxic like neurolethyrism [1].

Our patient had adult-onset sporadic progressive spastic paraparesis of 10 years duration. There was no evidence of extradural or intradural/extramedullary compression of the cord on MRI, infection like HIV, syphilis, metabolic like B12, E and copper deficiency and toxic like lathyrism. A diagnosis of sporadic HSP was made. Genetic confirmation could not be done due to non-availability. MRI showed tract-specific intramedullary signal changes involving the posterior horns appearing as 'Snake-eye'.

Hereditary spastic paraparesis is a disease which is compatible with a normal life expectancy but carries significant functional disability. The age at onset can range from infancy to eighth decade [2,3]. The main clinical abnormalities include spasticity of lower limbs, hyperreflexia and extensor plantar response. Hyperactive bladder symptoms in the form of frequency, urgency and hesitancy may be present [4]. HSPs can occur sporadic due to de novo mutations or can be transmitted in an autosomal dominant, autosomal recessive, X-linked or mitochondrial inheritance. The main neuropathological abnormalities include axonal degeneration involving corticospinal tracts and dorsal column tracts within the spinal cord. Spinocerebellar tracts are also reported to be affected. A process of "dying back" occurs wherein the degeneration begins in the distal part of long axons of corticospinal, dorsal column and spinocerebellar tracts and proceeds towards the cell body [5]. Axonal loss is accompanied by demyelination and gliosis. Dorsal root ganglia, dorsal roots, anterior horn cells and peripheral nerves are normal. Spinal cord appears atrophied on MRI in pure HSP.

'Snake-eye' appearance on MRI has been described in relation to abnormal bilateral hyper intensities of the anterior horns on axial spinal cord imaging. It has been described in association with quite a few lower motor neuron syndromes that includes spinal cord infarction, fibrocartilaginous emboli in to anterior spinal artery, resolving cord contusion due to gliosis, poliomyelitis, spondylotic myelopathy (bulging disc causing mechanical compression over radicular artery leading to chronic ischemia and gliosis in the region of anterior horn cells), radiation myelopathy and brachial monomelic amyotrophy [6].

Conclusion

The appearance of 'snake-eye' on MRI involving the posterior horn region has not been described in adult-onset progressive spastic paraparesis due to sporadic HSP so far. The possible mechanism for these hyper intensities may be gliosis involving posterior horn cells due to 'dying back' process involving the corticospinal tracts and the posterior column.

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