Case Report

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Wernekink commissure syndrome as the presenting feature in multiple sclerosis

Sheetal Sasikumar[1], Jeethu TJ[2], Chris Alvis Shaji[3], Aswathy Sasidharan[4]

[1]Associate Professor, Department of Neurology, Pushpagiri Institute of Medical Sciences and Research, Tiruvalla, Kerala, India

[2] Senior Resident, Department of Neurology, Pushpagiri Institute of Medical Sciences and Research, Tiruvalla, Kerala, India

[3] Junior Resident, Department of Medicine, Pushpagiri Institute of Medical Sciences and Research, Tiruvalla, Kerala, India

[4] Assistant Professor, Department of Neurology, MES medical college, Malappuram,Kerala, India.

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Abstract

Wernekink commissure syndrome is a rare syndrome characterized by bilateral cerebellar signs, internuclear ophthalmoplegia and delayed palatal myoclonus, due to involvement of the caudal paramedian midbrain. It has been almost exclusively described in strokes involving the midbrain. We hereby report the case of a gentleman who presented with features of Wernekink commissure syndrome and on evaluation was diagnosed to have multiple sclerosis. This is the first reported case of a patient with multiple sclerosis presenting with Wernekink commissure syndrome.

Keywords: Wernekink commissure syndrome; caudal midbrain; internuclear ophthalmoplegia; bilateral cerebellar signs; multiple sclerosis

Introduction

Wernekink syndrome is an uncommon midbrain syndrome characterized by bilateral cerebellar signs, internuclear ophthalmoplegia (INO) and delayed palatal tremor.[1] The Wernekink commissure refers to the decussation of the superior cerebellar peduncle.[2] Wernekink syndrome has been described in strokes involving the caudal paramedian midbrain, which houses the Wernekink commissure and the adjacent structures including the medial longitudinal fasciculus (MLF). It is rarely reported in demyelinating disorders. There is only one reported case of Wernekink syndrome in a patient with Neuromyelitis optica spectrum disorder (NMOSD).[3] It has not been reported as the presenting symptom of multiple sclerosis so far.

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Address for correspondence:
Sheetal Sasikumar
Associate Professor, Department of Neurology, Pushpagiri Institute of Medical Sciences and Research, Tiruvalla, Kerala, India
Email: sheetalrehaan@gmail.com

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Case report

A 55-year-old gentleman, with history of type 2 diabetes mellitus and systemic hypertension, presented with complaints of binocular double vision, with horizontal separation of images, on looking to either sides, for the past eight months. He denied history of diminution in vision, drooping of eyelids, facial deviation, dysphagia, dysarthria or limb weakness. He had no history of fever, vomiting or headache. He also complained of unsteadiness on walking, with swaying to either side for past 8 months. He had no history of memory impairment or executive dysfunction. He had no sensory, bowel or bladder symptoms. On examination, he had a pulse rate of 88/ minute and a blood pressure of 140/80 mm of Hg. Fundus examination was normal. He had normal visual acuity in both eyes. His pupils were symmetric and readily reacting to light. There was no ptosis. He had impaired adduction in both eyes, with nystagmus of the abducting eye, suggestive of bilateral inter nuclear ophthalmoplegia(INO). Cranial nerve examination was otherwise normal. Motor system examination revealed normal tone, power and deep tendon reflexes with bilateral flexor plantar response. He had positive finger nose test bilaterally and impaired tandem walking. Sensory system examination was normal. Magnetic resonance imaging (MRI) brain showed a T1hypointense, T2/FLAIR hyperintense lesion in the midbrain tectum, towards peri-aqueductal grey (PAG) with subtle post contrast enhancement and no perilesonal edema or mass effect (Figure 1 & 2). The presence of bilateral INO and bilateral cerebellar ataxia, along with MR evidence of caudal midbrain

involvement suggested the diagnosis of Wernekink commissure syndrome. Serum ACE, C-ANCA, P-ANCA and ANA-negative were negative. CSF analysis showed no cells, with normal CSF sugar and protein values. CSF OCB was positive, suggesting the diagnosis of multiple sclerosis. Serum anti - MOG antibody, and aquaporin 4 antibody were negative. MRI of the spine was normal. He was initiated on injection Methyl Predisolone, which was given for 5 days followed by Dimethyl fumarate. He had significant improvement in symptoms, on review after one month.

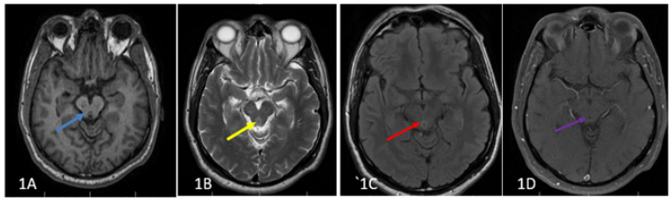

Figure 1: Axial MRI brain showing lesion in the midbrain tectum, towards peri-aqueductal grey (PAG) which is hypointense on T1 (1A), hyperintense on T2 (1B), hyperintense on FLAIR (1C), with minimal post contrast enhancement (1D)

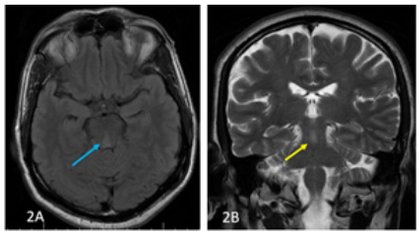

Figure 2: Axial FLAIR (2A) and coronal T2 (2B) MRI brain images showing hyperintense lesion in bilateral paramedian midbrain.

Discussion

Wernekink commissure syndrome is a rare midbrain syndrome characterized by internuclear ophthalmoplegia, bilateral cerebellar signs and delayed palatal myoclonus.[1] The Wernekink commissure is a horse shoe shaped commissure located anterior to the cerebral aqueduct in the caudal paramedian midbrain and is formed by the decussation of the superior cerebellar peduncle. The structures located near this commissure are the medial longitudinal fasciculus and the central tegmental tract; the characteristic clinical features of Wernekink commissure syndrome can be explained due to the involvement of these structures.[2] The involvement of the decussation of the superior cerebellar peduncle results in bilateral cerebellar signs, MLF involvement results in internuclear ophthalmoplegia and the involvement of the central tegmental tract can result in palatal myoclonus. If the involvement is bilateral, patients can present with bilateral internuclear ophthalmoplegia.[4]

In most of the cases of Wernekink commissure syndrome reported so far, the etiology has been stroke involving the caudal paramedian midbrain. It is rarely described in demyelinating disorders. Zou et al have described Wernekink commissure syndrome in a patient with neuromyelitis optica spectrum disorder (NMOSD).[3] Though bilateral INO has been frequently reported in multiple sclerosis, the constellation of symptoms of Wernekink commissure syndrome has not been reported in multiple sclerosis so far.[5] Our patient had bilateral INO and bilateral cerebellar signs. Palatal myoclonus was not seen in our patient on review. As per the neuroradiological criteria published by the same authors, the diagnosis of Wenekink commissure syndrome can be made if all three essential criteria including ipsilateral INO, unilateral or bilateral ataxia and neuro-radiological evidence of caudal midbrain involvement are present, with or without the presence of the supportive criteria, which includes rubral tremor and palatal myoclonus.[2]

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