Wernekink Commissure Syndrome: A Rare Cause of Bilateral Cerebellar Syndrome

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Abstract

Purpose: Focal signs are a big deal in neurology and are among the most important clues leading to diagnosis and localization. Wernekink commissure syndrome is due to lesions in the caudal paramedian midbrain involving the entire decussation of the superior cerebellar peduncles, resulting in the clinical hallmark of a bilateral cerebellar syndrome.

Case report: A 79-year-old man presented with sudden, severe unsteadiness associated with slurring of speech, binocular double vision, and bilateral hand tremor. Examination showed right INO, moderately severe dysarthria, bilateral dysmetria and dysdiadochokinesia, with severe truncal ataxia and bilateral upper and lower limb ataxia. Also, bilateral coarse tremor was noted in both hands which was present at rest, action and on reaching for objects. Brain MRI revealed an acute infarction involving the Wernekinck decussation in the right caudal midbrain and mesencephalo-pontine junction.

Conclusion: The differential of Wernekink Commissure Syndrome is complex, and localization and lateralization are extremely difficult owing to prominent bilateral cerebellar symptoms. The finding of an associated unilateral INO in some cases makes it possible to confidently narrow the list of differentials and localize the lesion to the paramedian tegmentum ipsilateral to the non-adducting eye.

Keywords: Stroke; Mesencephalon; Cerebellar Syndromes; Ocular Motility Disorders

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INTRODUCTION

Isolated midbrain ischemic stroke is rare (0.7-2.3% of posterior circulation strokes) owing to a complex and overlapping mesencephalic arterial blood supply[1]. Midbrain syndromes due to vascular insults, such as Weber, Benedict and Claude syndromes, usually affect the upper midbrain and result in clinical pictures dominated by oculomotor nerve palsy with contralateral hemiparesis, hemiparesis with involuntary movements, and ataxia respectively. Lower midbrain lesions are much less common and their symptomatology is less well recognized[2]. Wernekink commissure syndrome is an extremely rare syndrome due to a single, strategically placed lesion in the caudal paramedian midbrain that involves the entire decussation of the superior cerebellar

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peduncles, producing bilateral symptoms[3]. The clinical hallmark of this condition is a bilateral cerebellar syndrome, which may be associated with varying eye-movement signs and a possible tremor with or without late-onset palatal myoclonus[3]. Few cases of Wernekink commissure syndrome were reported since Lhermitte first described the condition in 1958[4]. A search in PubMed for variations of the word "Wernekink" revealed only 12 relevant results, and a recent literature review by Dong, et al. in late 2019 identified a mere 20 documented cases[5].

CASE REPORT

A 79-year-old man who is a known case of diabetes mellitus and hypertension presented to the ER with an 18-hour history of sudden, persistent unsteadiness associated with slurring of speech, binocular double vision worse upon looking to the left, and new bilateral hand tremor. He denied having dysphagia, limb weakness, sensory symptoms or decreased level of consciousness. Examination was remarkable for right internuclear ophthalmoplegia "INO", moderately severe dysarthria, bilateral dysmetria and dysdiadochokinesia, bilateral upper and lower limb ataxia, and severe truncal ataxia with a tendency to fall in either direction. Moreover, a bilateral irregular, medium amplitude, slow (3-4 Hz) tremor was noted in both hands, which was notably evident at rest and persisted with posture, during voluntary movements and on reaching for targets. There was no palatal myoclonus. Planter responses were flexor, and the remainder of the neurological examination was non-focal. Brain MRI revealed an acute infarction in the area corresponding to the Wernekinck decussation, involving the right caudal midbrain and mesencephalo-pontine junction (Figures 1-2). Another tiny lesion with fainter signal was also noted in the left basal ganglia. There were no cerebellar lesions. Upon follow up at the clinic after one month, the patient reported some improvement in his diplopia, unsteadiness and dysarthria. He also stated that his bilateral hand tremor has mostly disappeared except on certain tasks such as reaching for objects or feeding with a spoon. On examination, the patient could partially adduct his right eye, and improvement was noted in his gait, with mild residual truncal ataxia, upper and lower limb ataxia, dysmetria and intention tremor. There was no dysdiadochokinesia or palatal myoclonus. The patient was lost to further follow up.

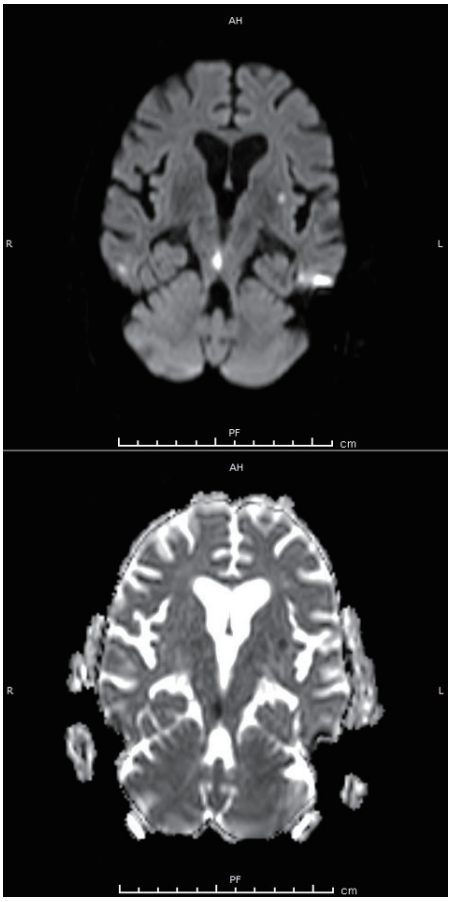

Figure 1. Diffusion and ADC map sequences demonstrating diffusion restricted lesions in the right caudal midbrain and the left basal ganglia. Device: 3-Tesla MRI. Slice thickness: 5 mm.

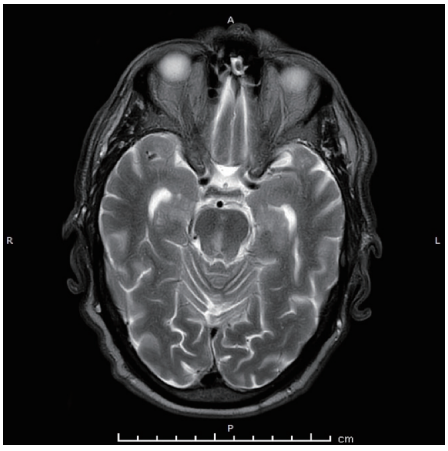

Figure 2. T2 weighted image from the same patient demonstrating the acute stroke in the right caudal midbrain. Device: 3-Tesla MRI. Slice thickness: 4 mm.

DISCUSSION

A prominent, bilateral cerebellar syndrome was present in 100% (n=21) of cases in the review by Dong, et al[5]. Such syndrome was predominated by the universal clinical findings of truncal ataxia, bilateral upper and lower limb ataxia, and dysarthria.

Eye-movement disorders were the second most consistent finding, being noted in 81% (n=17) of the cases. The most frequently reported eye-movement disorder was ophthalmoplegia, followed by nystagmus. INO was the commonest cause underlying ophthalmoplegia, being noted in 57% (n=12) of the cases, with unilateral INO (n=7) being slightly more prevalent than bilateral INO.

Palatal myoclonus, typically a late-onset manifestation, was seen in 14% (n=3) of cases, but this observation is limited by variation in follow up periods between some cases and lack of follow up in others.

Holmes tremor was noted in 19% (n=4) of the cases reviewed by Dong et al. Two of these cases presented with tremor at onset, similar to our case[5]. Also, cases presenting with Holmes tremor that had an early-onset or which was due to etiologies other than the classically recognized structural causes have been reported in the literature, which lends support to the opinion that Holmes tremor is heterogenous and describes a tremor phenomenology rather than a specific etiology[6].

Dong and colleagues also observed that impaired consciousness was relatively common in Wernekink Commissure Syndrome and was reported more frequently (33%, n=7) than either palatal myoclonus or tremor alone, however this was not a finding in our patient.

It is noteworthy that one case only[7] fulfils the classical triad first observed and reported by Lhermitte, which consisted of bilateral cerebellar syndrome, horizontal nystagmus, and late-onset myoclonus that manifested years later in a facial-pharyngo-laryngo-ocular distribution. However, while the finding of a bilateral cerebellar syndrome is uniformly present in all cases to date including ours, we believe that percentages pertaining to the prevalence of remaining clinical findings ought to be interpreted with caution due to the small number and varying methodology of the reported cases.

The differential diagnosis for a case presenting with prominent bilateral ataxia includes bilateral cerebellar strokes, acute cerebellitis, Miller-Fisher Syndrome, Wernicke's encephalopathy, and demyelinating lesions due to diseases such as multiple sclerosis[5,7]. Although the age and risk profile together with the sudden onset usually reported with vascular events collectively favor a stroke, predominantly bilateral cerebellar findings make neuroanatomical localization challenging in Wernekink commissure syndrome. In cases such as ours, the finding of a unilateral INO serves as a reliable clue for neuroanatomical localization and lateralization. Isolated INO localizes to the caudal or mid-pontine area, while the combination of ataxia and INO localizes to the paramedian tegmentum at the pontomesencephalic junction[8]. Lateralization is helped by noting the side of INO, which is always ipsilateral to the lesion. Due to the close proximity of the MLF to the midline, a large or occasionally bilateral lesion may produce bilateral INO and further obscure the clinical picture of an already peculiar syndrome.

Conflicts of interest: All authors declare that they have no conflicts of interest.

Ethical considerations: Consent for publication was obtained from the patient and identifying details were omitted from the manuscript.

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