

Bilateral Cerebellar Ataxia with no Apparent Cerebellar Lesions: A Case of Wernekink's Commissure Syndrome

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Abstract

Wernekink commissure syndrome was first described in 1941 by Lhermitte, but it wasn't until 1958 that it made its first appearance in a publication at the 22nd International Neurological Meeting. This syndrome takes its name from the first illustrations made in 1840 by Franz Joseph Julius Wilbrand, the successor of Friedrich Christian Wernekink (1798-1835). Wernekink's commissure syndrome is characterized by the combination of bilateral cerebellar ataxia with dysarthria or anarthria, associated with occasional internuclear ophthalmoplegia and delayed-onset palatal myoclonus (Holmes tremor), secondary to caudal paramedian midbrain infarction.

Keywords: Wernekink's commissure syndrome • Midbrain infarction • Cerebellar ataxia • Claude syndrome • Benedikt syndrome • Weber syndromee

Introduction

Wernekink commissure syndrome was first described in 1941 by Lhermitte, but it wasn't until 1958 that it made its first appearance in a publication at the 22nd International Neurological Meeting [1]. This syndrome takes its name from the first illustrations made in 1840 by Franz Joseph Julius Wilbrand, the successor of Friedrich Christian Wernekink (1798-1835) [2-4]. Wernekink's commissure syndrome is characterized by the combination of bilateral cerebellar ataxia with dysarthria or anarthria, associated with occasional internuclear ophthalmoplegia and delayed-onset palatal myoclonus (Holmes tremor), secondary to caudal paramedian midbrain infarction. Extremely rare, it represents less than 1% of cerebral infarctions and its clinical manifestation is very little known unlike other mesencephalic syndromes such as Weber syndrome, Claude syndrome or Benedikt syndrome [5,6]. We report the observation of a patient with Wernekink's commissure syndrome related to a midbrain infarction.

Case Presentation

A 59-year-old right-handed patient, having a cardiovascular risk factors; active chronic smoking, high blood pressure and unbalanced type II diabetes, presented to the emergency reception service thirty hours after the acute onset of intense crisis of rotary vertigo associated with a balance disorder, gait disturbance with an inability to stand without assistance and a speech disorder. He also reported having had two episodes of vomiting that reportedly occurred at home.

On admission, he had high blood pressure at 159/88 mmHg and the rest of the vital parameters were normal. He was conscious and his speech was very dysarthric, the word wasn't rhythmic, explosive, poorly articulated and elocution was slowed down.

The Deep tendon reflexes (patellar and tricipital) were pendular (hypotonia) and the plantar reflex was flexed bilaterally. He had a very marked

clumsiness of four limbs with a severe dysmetria of them more marked on the right during the finger to nose and heel to knee maneuvers, as well as dysdiadochokinesia (rapid alternating movements) and asynergy. In addition, he had a left horizontal nystagmus, associated with a limitation of the abduction of the left eye and the abduction of the right eye. Convergence was discreetly altered and pupillary light reflex was preserved.

The ataxic table is in favour of a bilateral cerebellar syndrome prevailing on the right associated with left internuclear ophthalmoplegia evocative of a Wernekink's commissure syndrome. A Computed Tomography Scan (CT) made on admission was without anomaly.

Brain Magnetic Resonance Imaging (MRI) performed the day after admission revealed a lesion in the left lower paramedian midbrain and left upper pons, suggesting an infarction in looks at the Wernekink commissure and some signal anomalies of the deep white substance appearing in hypersignal T2 and FLAIR (Fluid Attenuated Inversion Recovery) (Figure 1).

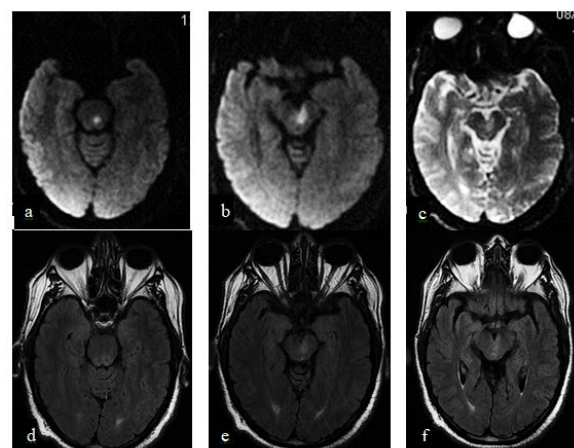


Figure 1. (a) Brain magnetic resonance imaging in axial cuts showing

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a restriction of diffusion in the pontic paramedian region (b) left mesencephalic (c) T2-weighted axial MRI showing a left mesencephalic paramedian hypersignal (d) (e) (f) pontic hypersignal, mesencephalic and leucoencephalopathy in FLAIR (Fluid Attenuated Inversion Recovery).

Results

Transthoracic echocardiogram showed concentric left ventricular hypertrophy, without mural thrombus. The supra-aortic trunk echo-Doppler was without anomaly. The Holter monitoring did not reveal any anomaly of repolarization, or rhythm disturbance. The CT angiography of the neck and cerebral vessels did not objectify anything. Blood tests had objectified a diabetic imbalance with glycosylated haemoglobin (HbA1c) at 9.9% and dyslipidemia. The patient was treated with Platelet antiaggregants (Acetyl salicylic acid), an ACE inhibitor (perindopril), a lipid-lowering agent (simvastatin), Long-acting insulin and balance rehabilitation.

Discussion

We have only been aware of very few reports of midbrain infarction characterized by Wernekink's commissure syndrome. Indeed, this is an extremely rare syndrome, justifying the fact that only a few series of cases and isolated cases have been reported [7–17]. Its incidence remains very low, representing 0.6% of the total number of patients admitted for ischemic stroke. The rarity of midbrain infarction is explained by the fact that the arterial blood supply to the midbrain is complex, compared to that of the pons and the medulla. Also, there are overlaps between arterial territories of sub-tentorial structures and individual variations [18, 19].

Wernekink's commissure involves the decussation of the superior cerebellar peduncle in the midbrain. To better understand the symptomatology related to mesencephalic impairment, let's first address the neurovascular anatomy of the midbrain. The midbrain extends from the ponto-midbrain junction to the diencephalon. The midbrain is divided into two parts (rostral midbrain and caudal midbrain). One just below the lower thalamus (rostral midbrain) and the other just above the isthmus (caudal midbrain).

The caudal paramedian mesencephalon includes small, extremely medial parts, the cerebral peduncles (frontopontine tract), extremely medial portions of the substantiae nigrae, the superior cerebellar peduncles (Wernekink decussation), the central tegmental tract (the component of the Guillain and Mollaret triangle) and its decussation, the medial longitudinal fasciculus (MLF), the nucleus of the trochlear nerve (IV cranial nerve), the short segment of IV fibers and the reticular structure. The rostral midbrain includes corticospinal tract, substantia nigra, red nucleus, oculomotor nucleus and Edinger-Westphale nucleus, periaqueductal gray.

Anatomically, the decussation of the superior cerebellar peduncles and the central tegmental tract constitute the commissure of Wernekink in "horseshoe shape" which owes its name to the German anatomist Friedrich Wernekink [20]. The arterial territories of the midbrain are classified according to the literature and consist of four groups: a prominent anteromedial group (paramedian), a large anterolateral group, a small lateral group and a small dorsal group [21]. The paramedian territories of the midbrain and thalamus are provided by interpeduncular perforating branches.

Pedroza et al have described the interpeduncular branches in three groups of vessels which come from the last 5 mm of the basilar artery, the initial 7 mm of the two Superior Cerebellar Artery (SCA) and the initial segment (segment P1) of the Posterior Cerebral Artery (PCA) [22]. The perforated interpeduncular branches show great variability in number, size, origin and territorial contribution to the midbrain and thalamus.

Small infarctions of the brainstem and of the midbrain in particular, often lead to various clinical syndromes, in particular the syndromes of Weber, Benedikt and Claude, which, unlike the syndrome of the commissure of Wernekink, are well known. However, note that lacunar stroke of the

midbrain have a wide range of clinical expressions and is sometimes difficult to diagnose.

The bilateral cerebellum dysfunction characterized by a disorder of elocution (a dysarthric speech), truncal and gait ataxia, and ataxic movement of the four limbs, is explained by Infarction of paramedian mesencephalic that can be seen on MRI. This infarction involves the decussation of the superior cerebellar peduncles and explains the symptomatology because a unilateral lesion at this level can be interrupted the spinocerebellar and dentato-rubro-thalamic tract just before and after decussation. On the other hand, the partial implication of the decussation of brachium conjunctivum explains the asymmetric distribution of cerebellar signs and the different degrees of disability. Our patient had a right predominance of ataxia. Internuclear ophthalmoplegia is due to the attainment of the Medial Longitudinal Fasciculus (MLF).

A median lesion, if it is located at the decussation of the brachium conjunctivum, can cause bilateral olivary degeneration if there is damage to the right and left dentato-olivary fibers during their crossing, and result in a palatal myoclonus generally delayed [23].

The observation made in our patient does not reveal palatine myoclonus, because this observation is made in the acute phase. However, it is well known that the time interval between the appearance of a lesion within the dentato-rubro-olivary system and the clinical manifestations of palatal tremor is very wide [24], and the data in the literature indicate also that palatal tremor does not develop in all patients with olivary degeneration [25].

In the face of bilateral cerebellar ataxia, it is appropriate to discuss, on the one hand, the bihemispheric cerebellar lesions (vascular, infectious: Acute viral cerebellar and inflammatory), on the other hand, lesions of the middle cerebellar peduncles (at the level of the pons).

However, the existence of at least one of the signs of internuclear ophthalmoplegia is useful for topographic diagnosis (differentiates the lesion territory). It is the MRI that allows the precise diagnosis [26].

In 2019, Zhou et al proposed a classification of the etiologies of mesencephalic infarction. This classification is based on the results of Magnetic Resonance Angiography (MRA), carotid ultrasonography, and echocardiograph/electrocardiography. They classified them as large artery atherosclerosis disease (41.7%), small vessel disease (33.3%), cardiac embolism (16.7%), or stroke of undetermined etiology (8.3%) [27]. Our patient had a lacunar stroke due to damage to the small paramedian arterial vessels.

Conclusion

Lacunar infarcts (small infarction) in the midbrain have a wide spectrum of clinical expressions and are sometimes difficult to diagnose. Wernekink's commissure syndrome is a rare midbrain syndrome. It is the result of a lesion of the commissure of Wernekink involving the decussation of the superior cerebellar peduncle in the midbrain. This syndrome can present a clinical picture characterized by the association of bilateral cerebellar ataxia, internuclear ophthalmoplegia and palatal myoclonus. Although rare and little known, it is essential not to ignore it.

Authorship

All authors contributed to the design of the manuscript and approved its publication.

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Conflicts of Interest

We declare that we have no conflicts of interest in connection with this manuscript.

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Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

Informed consent

Written informed consent was obtained from the patients for their anonymized information to be published in this article.

References.

1. Lhermitte, F. "Anatomy of clinical study in adults. Report on the cerebellum of the XXII international neurological Meeting." *Rev Neurol* 98(1958):435-477.
2. Bogousslavsky, J, Maeder P and Regli F. "Pure midbrain infarction: Clinical syndromes, MRI, and etiologic patterns." *Neurology* 44(1994):2032-2040.
3. Mossuto, Agatiello L. "Caudal paramedian midbrain syndrome." *Neurology* 66(2006):1668-1671.
4. Wilbrand, FJJ. "Anatomy and physiology of the central structures of the nervous system." *Neurology* 47(1997):3032-3040.
5. Kim, JS and Kim J. "Pure midbrain infarction: Clinical, radiologic, and pathophysiologic findings." *Neurology* 64(2005):1227-1232.
6. Adeva, MT, Gomez Sanchez JC and Marcos MM. "Triple association of mesencephalic syndromes." *Rev Neurol* 28(1999):403-404.
7. Chenguang, Z, Yuanhong H, Zhiwen C and Yinghui Z, et al. "Wernekink commissure syndrome secondary to bilateral caudal paramedian midbrain infarction presenting with a unique heart or V appearance sign: Case report and review of the literature." *Front Neurol* 8(2017):376.
8. Cerrato, P, Lentini A, Colonna R and Bosco G, et al. "Gait and bilateral limb ataxia as isolated feature of a lower midbrain tegmental infarction." *J Neurol* 255(2008):290-291.
9. Okuda, B, Tachibana H, Sugita M and Maeda Y. "Bilateral internuclear ophthalmoplegia, ataxia, and tremor from a midbrain infarction." *Stroke J Cereb Circ* 24(1993):481-482.
10. Zhou, C, He Y, Chao Z and Zhu Y, et al. "Wernekink commissure syndrome secondary to bilateral caudal paramedian midbrain infarction presenting with a unique heart or V appearance sign: Case report and review of the literature." *Front Neurol* 35(2017): 368-376.
11. Zhu, Y, Liu HN and Zhang CD. "Wernekink commissure syndrome is a pure midbrain infarction." *J Clin Neurosci* 17(2010):1091-1092.
12. Mullaguri, N, Battineni A and Chuquilin M. "Wernekink commissure syndrome with palatal myoclonus at onset: a case report and review of the literature." *J Med Case Rep* 12(2018):113.
13. Sheetal, S and Byju P. "Wernekink commissure syndrome." *Neurol India* 64(2016):1055-1057.
14. Dai, AI and Wasay M. "Wernekink commissure syndrome: A rare midbrain syndrome secondary to stroke." *J Pak Med Assoc* 56(2006):289-290.
15. Sato, S, Toyoda K, Kawase K and Kasuya J, et al. "A caudal mesencephalic infarct presenting only with tetra-ataxia and tremor." *Cerebrovasc Dis* 25(2008):187-189.
16. Kim, TW, Yoo S and Koo J. "Wernekink commissure syndrome secondary to ischemic stroke: Severe dysarthria is one of the main characteristics of this syndrome." *Neurol Sci* 35(2014):1475-1477.
17. Spengos, K. "Bilateral paramedian midbrain infarct: an uncommon variant of the "top of the basilar" syndrome." *J Neurol Neurosurg Psych* 76(2005):742-743.
18. Castaigne, P, Lhermitte F, Gautier JC and Escourolle R, et al. "Arterial occlusions in the vertebro-basilar system: A study of 44 patients with post-mortem data." *Brain* 96(1973):133-54.
19. Burger, KM, Tuhim S and Neidich TP. "Brainstem vascular stroke anatomy." *Neuroimag Clin N Am* 15(2005):297-324.
20. Voogd, J and Baarsen KV. "The horseshoe shaped commissure of wernekink or the decussation of the brachium conjunctivum methodological changes in the 1840s." *Cerebellum* 13(2013):113-120.
21. Tatu, L, Moulin T, Bogousslavsky J and Duvernoy H. "Arterial territories of human brain brainstem and cerebellum." *Neurol* 47(1996):1125.
22. Pedroza, A, Dujovny M, Ausman JI and Diaz FG, et al. "Microvascular anatomy of the interpeduncular fossa." *J Neurosurg* 64(1986):484-493.
23. Agatiello, LM. "Caudal paramedian midbrain syndrome." *Neurology* 66(2006):1668-1671.
24. Lapresle, J. "Palatal myoclonus." *Adv Neurol* 43(1986):265-273.
25. Matsuo, F and Ajax ET. "Palatal myoclonus and denervation super sensitivity in the central nervous system." *Ann Neurol* 5(1979):72-78.
26. Lee, H. "Caudal paramedian midbrain syndrome." *J Neurol Neurosurg Psych* 74(2003):1445.
27. Zhou, C, Xu Z, Huang B and He Y, et al. "Caudal paramedian midbrain infarction: A clinical study of imaging, clinical features and stroke mechanisms." *Acta Neurol Belg* 8(2019):65-97.

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