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EDITORIAL

Vertigo in Cerebellar Disease—Do the Eyes Have It or Is There More to Perceive?

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The vertigo is a sensation characterizing the erroneous motion of the self or surround. The disabling percept is often like a spin or a sway. The false sensation of motion impairs daily life activities, leading to substantial disability in gait and other volitional movements. Such profound deficit in self-motion is often seen in rather benign conditions such as benign paroxysmal positional vertigo (BPPV) or in serious central disorders ranging from chronic dizziness featuring degenerative cerebellar conditions to dramatic presentation of acute cerebellar stroke.

Regardless of its etiology, the examination of eye movements plays a substantial role in thorough clinical investigation of a dizzy patient. On the flip side, the detailed ocular motor examination had often pointed the clinicians in the right direction when the diagnosis of the cerebellar disorder was under a conundrum. Gait and appendicular ataxia are other common coexisting features when the cerebellum is affected. Historically, although the reports of degenerative cerebellar disorders have frequently included ataxia, inclusion of eye movement deficits has not been so common in the past years. The best example is depicted in Fig. 1 where the trend of peer-reviewed publications on PubMed where cerebellar degeneration was one of the key words (black line, Fig. 1) in either the title or the abstract is compared with the number of publications with key words cerebellar degeneration plus ataxia (blue line, Fig. 1), plus nystagmus (gray line, Fig. 1), plus any eye movement deficit (pink line, Fig. 1), and plus gait disorder (green line, Fig. 1). It is surprising and unexpected from the standpoint of clinical wisdom that the reports of eye movement or gait disorders along with cerebellar degeneration are rare compared to the combination of ataxia with cerebellar degeneration. Such trend of published literature poses an important question—how vital the eye movement or gait examination is—although clinical wisdom has always suggested their critical role in diagnostics of the cerebellar conditions?

An elegant and rigorous study conducted by the group of investigators from the Ludwig-MaxMillan University systematically assessed the ocular motor deficits in the large cohort of the cerebellar patients with primary presentation of vertigo [1]. In this extensive dataset, the authors excluded patient with rather rare, focal, and well-defined lesions of the cerebellum, but analyzed relatively common but frequently disregarded chronic or episodic vertigo with no obvious structural deficits. The authors found a robust link to the cerebellum. Careful clinical examination frequently revealed cerebellar deficit, and often it categorized the symptoms to specific posterior fossa region [1]. Of all patients with cerebellar degenerative disorders, 81% had vertigo and 95% of vertigo patients had eye movement abnormality. The trend was similar in paraneoplastic or immune-mediated forms or hereditary cerebellar disorders. From the physiological standpoint, it is expected that sensation of vertigo should correlate with the presence of nystagmus during primary gaze since the constant movements of the eyes (hence retinal image motion) leads to perception of rotation. Although the eye movement deficit, such as ocular pursuit abnormality, was quite common and was present in 95% of these patients, only 24% had central fixational nystagmus. Therefore, while it is convincing that eye movements represent an important "marker" for the cerebellar involvement, it is still unclear why abnormal perception of surrounding motion leading to vertigo was present in almost three times more number of patients. We interpret that the presence of nystagmus is not an absolute requirement for perception of vertigo.

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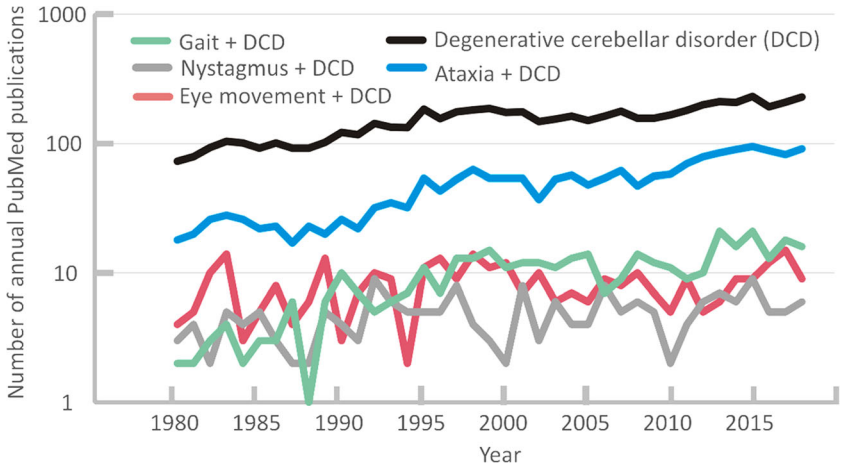

Fig. 1 Analysis of number of peer-reviewed publications on cerebellar degeneration with or without various co-morbidities in human patients. The number of annual PubMed publications was extracted with PubMed search of the keyword "cerebellar degeneration." Black trace depicts annual number of publications from 1980 through 2018. Blue trace is the number of annual publications where cerebellar degeneration and ataxia is reported, green trace is when cerebellar degeneration is associated with gait disorder, gray trace nystagmus, and pink trace eye movement disorder. It is noteworthy that ataxia is by far the commonly co-reported symptom with degenerative cerebellar disorder compared to ocular motor or gait deficit. The trend remains about the same through nearly four decades

This study brings a fundamentally important question, why the cerebellar patients with putative vermis involvement have false perception of rotation (i.e., vertigo) even when nystagmus is not present? The question brings us to scientifically novel area of investigation where the role of cerebellum was examined in the physiology of motion perception. Contemporary literature had asked whether the human cerebellum plays a role in neural processing beyond motor control, including perception of the environment and others. Does the cerebellum influence higher brain function through multiple cerebello-cerebral loops? What are the neural strategies for the cerebellar processing of self-motion perception and perception of objects moving around? In natural behavior, the brain computes a precise internal estimate of body movement that ensures appropriate reflexive behavior and accurate perception of self-motion. Classic vestibular and eye movement studies delineated basic principles of central motion processing in pathways responsible for the reflexive eye movements—the vestibulo-ocular reflex (VOR). Perceived angular motion may also utilize the same basic principles with some fundamental differences especially in dynamic properties of perceived motion as compared to that provoked due to eye movements [2–4]. One may consider several theoretical constructs to explain differences of perceived motion without the influence of eye movements. One obvious factor is the influence of prediction in perception [5]. Predictive behavior relies on the memory of the preceding sensations and the influence of the reaction (or percept) to the ongoing sensory input. In other words, the reaction (or percept) of ongoing sensory signal could represent the average of the predicted (cognitively determined) and current sensory signals [6, 7]. This downstream signal processing could include the weighted averaging of the prospective estimate of future movement (prediction based on the memory of the immediately preceding movement—cognitive or predictive component) and the current signal received from the sensory organs. During impoverished or disease-affected conditions, such as in patients with diffuse cerebellar insult, the cognitive or predictive component of motion cues might have a stronger influence on the computation of perceived motion [7]. Such a mechanism might also disrupt the perceptual physiological response to rapidly changing unexpected movements, leading to perception of sways and imbalance during locomotion as well as vertigo without nystagmus. Key questions need to be addressed: what is the pattern of activation of the cerebellum during the observation of others moving around? Does the cerebellum participate actively in action perception? What is the link with the processing of internal/external kinematics at the level of cerebellar cortex and cerebellar nuclei? What is the role of the cerebellar node(s) in the processing of sways/balance in the context of social interactions, a fundamental element of human behavior?

To epitomize, this editorial highlights a key study where eye movements were identified as a critical "marker" of cerebellar dysfunction in patients with vertigo. We believe in the authors' claim that eye movements should be carefully examined not only to identify cerebellar involvement, but also to categorize cerebellar dysfunction to particular posterior fossa syndrome or Schmahmann's syndrome. Such categorization (in future) may lead to proper identification of pharmacotherapy targets. Finally, we also emphasize the importance of the cerebellum on abnormally perceived motion even in the absence of corresponding movement. Latter deficit is organic and is far more common than clinically appreciated; it leads to substantial impairment in morbidity and quality of life in affected individuals.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Ethical Committee Approval Not applicable.

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References

1. Feil K, Strobl R, Schindler A, Krafczyk S, Goldschagg N, Frenzel C, et al. What's behind cerebellar vertigo and dizziness? Cerebellum. 2019. https://doi.org/10.1007/s12311-018-0992-8.
2. Shaikh AG, Palla A, Marti S, Olasagasti I, Optican LM, Zee DS, et al. Role of cerebellum in motion perception and vestibulo-ocular reflex-similarities and disparities. Cerebellum. 2013;12(1):97–107.
3. Shaikh AG, Straumann D, Palla A. Motion illusion-evidence towards human vestibulo-thalamic projections. Cerebellum. 2017;16(3):656–63.
4. Tarnutzer AA, Shaikh AG, Palla A, Straumann D, Marti S. Vestibulo-cerebellar disease impairs the central representation of self-orientation. Front Neurol. 2011;2:11.
5. Lee DN. A theory of visual control of braking based on information about time-to-collision. Perception. 1976;5(4):437–59.
6. Blumle A, Maurer C, Schweigart G, Mergner T. A cognitive intersensory interaction mechanism in human postural control. Exp Brain Res. 2006;173(3):357–63.
7. Jurgens R, Becker W. Perception of angular displacement without landmarks: evidence for Bayesian fusion of vestibular, optokinetic, podokinesthetic, and cognitive information. Exp Brain Res. 2006;174(3):528–43.