Neuro-ophthalmic Manifestations of Cerebellar Disease

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KEYWORDS

• Cerebellum • Neuro-ophthalmology • Eye movements • Nystagmus • Saccades • Smooth pursuit • Vestibuloocular reflex • Vergence

KEY POINTS

- The cerebellum is intimately involved in all classes of eye movements.
- Ocular motor findings can help localize the cerebellar structure that is dysfunctional.
- The 3 regions of the cerebellum involved in ocular motor control are (1) the oculomotor vermis (lobule VI and VII) and fastigial nuclei, (2) the uvula and nodulus, and (3) the flocculus and paraflocculus. Lesions to each region result in a unique clinical syndrome.
- Certain cerebellar disorders may result in abnormalities of the afferent visual pathway.
- Oscillopsia may arise from nystagmus or saccadic intrusions, and be highly disabling in some patients. Certain therapeutic measures can dampen the nystagmus or saccadic intrusions to improve the visual acuity of these patients.

INTRODUCTION

The cerebellum is responsible for sculpting and refining ocular movements to ensure their precision and accuracy (to bring images to the fovea and keep them stable there), thereby guaranteeing the best possible visual acuity and clarity despite changes in body or head positions or movement of the object of interest. Cerebellar lesions do not abolish eye movements but cause them to become coarse, slow, imprecise, and unreliable, leading to degradation in the quality of vision.

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Case Vignette

A 60-year-old man complains of a bouncing of his vision when he walks or drives. As a result, he has to stop in order to read road signs or to look at the facial features of others. He also reports progressively worsening gait imbalance, but denies any cognitive deficits, autonomic symptoms, or parkinsonian features. He has no family history of similar complaints. Examination reveals gaze-evoked nystagmus, rebound nystagmus, and saccadic pursuit. Ophthalmoscopy revealed that his discs were unstable and moved with head oscillations. He lost more than 5 lines of acuity when the examiner oscillated his head in the horizontal and vertical planes. He had some mild dysmetria, dysdiadochokinesia, and heel-shin ataxia. His gait was wide based and he was not able to maintain his stance when his eyes were closed. He had diminution of sensation over his shoulders, posterior torso, and proximal lower extremities. Magnetic resonance imaging (MRI) revealed cerebellar atrophy, most noticeably over the anterior and dorsal vermis. What is his likely diagnosis? What electrophysiologic test can help confirm this diagnosis?

The cerebellum is involved in the control of all classes of eye movements, both in their real-time, immediate modulation, and in their long-term adaptive calibration.[1] When approaching cerebellar neuro-ophthalmic abnormalities, 3 important caveats should be remembered. First, the cerebellum is able to adapt and compensate for lesions, and therefore the ocular motor manifestations of acute cerebellar lesions can change, and usually improve with time. Second, because of the intimate relationship between the brainstem and cerebellar pathways that mediate eye movements, lesions affecting the cerebellar peduncles, or the cerebellar brainstem projections (both efferent and afferent), can also give rise to clinical signs and symptoms suggestive of cerebellar dysfunction. The presence of such phenomenology does not automatically indicate damage to the cerebellum. Furthermore, in disorders that affect both the brainstem and cerebellum, it may be impossible to accurately localize the source of a specific ocular motility disorder. In addition, although eye movement abnormalities are the most prominent neuro-ophthalmic manifestation of cerebellar disease, afferent visual pathway disorders may coexist with some cerebellar disorders; the presence of such findings often provides vital clues regarding the diagnosis.

This article begins with an overview of the role of cerebellar structures in modulating various classes of eye movements, including gaze holding, the vestibular ocular reflex (VOR), saccades, vergence, smooth pursuit, and the optokinetic reflex (OKR). This overview is germane to the later discussion of the different ocular motor abnormalities that can arise from cerebellar disorders. In addition, some treatment options for oscillopsia, a common symptom of cerebellar disease, are discussed.

THE ROLE OF THE CEREBELLUM IN OCULAR MOTOR CONTROL

The goal of all eye movements is to direct and maintain the angle of gaze (ie, the line of sight of the fovea) on an object of interest, to ensure the best visual acuity and clarity. Three distinct mechanisms help the visual system achieve this goal: (1) fixation, which detects (and corrects for) any retinal image drift, and suppresses unwanted saccades; (2) the VOR, by which eye movements compensate for head perturbations at short latency, ensuring visual acuity during locomotion; and (3) the gaze-holding system, which counteracts the elasticity of orbital tissue.[1–3]

The cerebellum continuously uses visual input to calibrate and optimize all categories of eye movements. Neurons in the dorsolateral pontine nuclei and climbing fibers from the inferior olivary nuclei (ION) convey visual signals to the cerebellum.[1,3,4] Another important aspect of the role of the cerebellum in calibrating eye movements