Visual Deficiency in Wallenberg’s Syndrome

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ABSTRACT

The aim of this case report of a 47-year-old woman who suffered from acute right-sided medullar ischemic stroke was to define the range of visual impairment in Wallenberg’s syndrome (WS). The patient complained of unbearable environmental tilt and rotating visual perception. On examination, 11 months following the stroke, the patient manifested rightsided postural inclination and gaze ipsipulsion. The fixation in primary position was unstable, after a conjugate ipsipulsion ensued, spontaneous corrective saccades and a horizontal-rotational jerking nystagmus beating away from the side of the lesion were generated. Monocular visual acuity (right eye: 0.4 logMAR distance and 0.2 logMAR near; left eye: 0.1 logMAR distance and 0.0 logMAR near) was significantly better than binocular (0.63 logMAR distance and near). Fluent reading was impossible. Contralateral smooth pursuits were more impaired. Saccades were defective manifesting right hypermetria and left hypometria. Visual field was constricted to central 10 – 20°. A diagnosis of Wallenberg’s syndrome was made. Occlusion was prescribed. Review of literature demonstrated lack of evidence-based guidelines for ophthalmic assessment and treatment of visual impairment in WS. Oculomotor abnormalities, oscillopsia and tilt illusion cause significant impact to daily life. Early post-stroke ophthalmological evaluation is thus mandatory in order to offer timing treatment.

Key words: lateral medullary syndrome, ischemic stroke, vision, nystagmus, saccades

Introduction

With an estimated prevalence of 65 %, poststroke visual impairment occurs commonly in an acute stroke population. Wallenberg’s (lateral medullary or retro-olivary) syndrome is caused by ischemia in the area supplied by posterior inferior cerebellar artery. It presents with vertigo and nassea, ipsilateral cerebellar signs and symptoms, Horner’s syndrome, palatal, pharyngeal, and vocal cord paralysis, dysphagia, and dysarthria, facial hypoalgnesia and thermanaesthesia and contralateral hypoalgnesia of trunk and extremities. Saccadic abnormalities (ipsipulsion and torsipulsion), skew deviation, nystagmus, smooth pursuit, and gaze-holding abnormalities and distortion of the visual field at 90° or 180° commonly occur in WS.

Case Report

A 47-year-old woman was referred to our Department in September 2020 due to unbearable environmental tilt and rotating visual perception. In October 2019 the patient suffered acute right medullar ischemia due to right vertebral artery dissection verified by MSCT. The functional outcome of the first brainstem infarction in 2010 was complete recovery. On examination, the patient tilted to the right describing a sensation of being pulled to the right side and manifested right upper lid ptosis, however, with no signs of Horner syndrome. She managed to keep the fixation at a distance only for a short time as conjugate right-sided ipsipulsion ensued, further compensated with corrective saccades and spontaneous horizontal-rotational jerking nystagmus beating away from the side of the lesion. These symptoms exaggerated while reading. Upon closing the right eye (RE) a moderate relief of symptoms occurred. Tracking of moving objects was severely compromised. When relaxed in the dark, a conjugate deviation of the eyes to the right was exhibited, followed by significant head and body tilt to the right side. Binocular visual acuity was 0.63 logMAR at distance and 0.63 logMAR at near. Monocular visual acuity was 0.4 logMAR distance and 0.2 logMAR near.
was significantly better: RE measured 0.4 logMAR at distance and 0.2 logMAR at near, while left eye measured 0.1 logMAR at distance and 0.0 logMAR at near. Fluent reading was impossible. Binocularly, the preferred position of gaze was of maximal left version, while during RE monocular fixation a strong face turn to the right side, and a pronounced inclination of the head and the body to the right ensued.

When performing saccades, right-sided hypermetria and left-sided hypometria were elicited. Vertical saccades manifested right-sided deflection. Smooth pursuit movements were performed easier to the right side, with the occurrence of corrective horizontal-torsional jerking nystagmus at about 10–15° from the midline and eyelid twitch. Kinetic visual field (VF) was difficult to perform because of the unstable fixation, however right and left VF were constricted to the central 10° and 20–25° respectively. Slit-lamp and fundus examination were unremarkable. A diagnosis of WS has been made and RE occlusion was prescribed.

Discussion

This is the first comprehensive report on visual impairment in WS. Wallenberg's syndrome is a unique but variable neurological entity. The most common causes of WS include ipsilateral vertebral artery or the posterior inferior cerebellar artery occlusion supplying the posteroinferior cerebellum. Although ischemia is the most common aetiology, demyelination is also acknowledged as the cause of WS.

A wide range of oculomotor and vestibular deficits encountered in WS include drift of the eyes towards the ipsilateral side, spontaneous, gaze-evoked and seesaw nystagmus, impaired contralateral pursuit eye movements, saccadic lateropulsion, and skew deviation, sometimes combined with an ipsilateral head tilt. A characteristic feature of these disturbances is lateropulsion, a directional preponderance of slow eye and saccadic movements towards the side of the lesion. Besides, patients with WS have gait ataxia and slant or lean ipsilaterally. The majority of signs and symptoms, such as vertigo, nystagmus, tilt illusion, skew deviation, the angular displacement of posture can be explained by vestibular dysfunction. However, the origin of other symptoms including ocular and saccadic lateropulsion, is much less certain since these symptoms may reflect either a vestibular imbalance or damage to other oculomotor and postural pathways in the inferior cerebellar peduncle.

It is advocated that infarction is limited to the lateral medulla disrupting afferent olivo-cerebellar climbing pathways. Saccadic ipsipsulsion is thus related to a general increase in spontaneous firing of Purkinje cells resulting in inhibition of the deep cerebellar nuclei. Hence saccadic dysmetria is the result of a functional but not structural cerebellar lesion. Deficits in gaze holding mechanisms, optokinetic responses, and vestibulo-ocular reflex suppression are consistent with lack of climbing fiber input to the (para)floculus which transmits specific image motion information.

With the loss of fixation, eyes tend to deviate toward the side of the lesion. If a patient is asked to fixate straight ahead and close the eyes, ipsipsulsion will be elicited, as in this case. This phenomenon can be induced even by blinking. Consequently, corrective saccades away from the side of the lesion occur to bring the eyes back toward the intended target. However, these corrective fast components are often inadequate to compensate for the slow components, and the average eye position slides toward the side of the lesion. This resulted in severely constricted visual fields in our patient.

As evidenced in this case, the lesion involving the climbing fibers within the right lateral medulla caused hypoactive leftward saccades (contralesional hypometria) and overactive rightward saccades (ipsilesional hypermetria), and ipsipsulsion. During vertical saccades, oblique or elliptic saccade directed toward the direction of lateropulsion appeared.

Also, the patient complained of an unbelievable sensation of environmental tilt, in which the entire room is tilted on one side or even upside down (“floor on ceiling” phenomenon). This phenomenon is also attributed to a disturbance of vestibular-otolith central connections. Binocular visual acuity was significantly worse compared to monocular due to horizontal-rotational jerking nystagmus beating away from the side of the lesion that was not pronounced on monocular viewing. In WS, nystagmus may be due to direct damage of the vestibular nuclei or their cerebellar, semi-circular canal, or otolthic connections most commonly presenting as horizontal or mixed horizontal-torsional beating away from the side of the lesion. It may not be a prominent feature of the syndrome because of the lateral and caudal location of the WS and sparing of the rostral and medial structures.

Further, our patient complained that the objects in motion create significant asthenopia and dizziness. It is evidenced that smooth pursuit eye movements that track targets moving away from the side of the lesion are also impaired in these patients.

In patients with WS, ipsilateral Horner’s syndrome is well recognized and associated with the damage of descending sympathetic fibers.

Conclusion

To conclude, a wide range of visual disorders occurs post-stroke, and WS is the most common syndrome related to intracranial vertebral artery occlusion. Despite that, there is a lack of evidence-based guidelines for ophthalmic assessment and treatment of visual abnormalities in WS. In this report, we have documented a plethora of disturbances of oculomotor control in a patient with WS. Spontaneous nystagmus, unstable fixation, smooth-pursuit, oscillopsia, and gaze-hold lateropulsion cause severe
visual symptoms that significantly impair quality of life. Furthermore, oculomotor abnormalities are often unappreciated and rehabilitation is largely limited to neurologi-
cal recovery. We recommend early post-stroke ophthalmo-
logical evaluation to offer appropriate and timing
treatment.

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WALLENBERGOV SINDROM I POREMEĆAJI VIDA

SAŽETAK

Cilj je ovoga prikaza slučaja 47-godišnje bolesnice s desnostranim medularnim ishemijskim moždanim udarom defini-
rati opseg oštećenja vida kod Wallenbergova sindroma (WS). Bolesnica se žalila na nesnosan osjećaj nagiba okoline i
rotirajuću vizualnu percepciju. Na kliničkome pregledu jedanaest mjeseci nakon moždanoga udara manifestirala je
desnostranu inklinaciju trupa i ipsipulziju pogleda. U primarnom položaju fiksacija je bila nestabilna, a slijedila ju je
konjugirana ipsipulzija te spontane korekcijske sakade i horizontalno-rotacijski trzajući nistagmus usmjeren suprotno
od strane lezije. Monokularna vidna oštrina (desno oko: 0.4 logMAR daljina i 0.2 logMAR blizina; lijevo oko: 0.1 logMAR
daljina i 0.0 logMAR blizina) bila je značajno bolja od binokularne (0,63 logMAR daljina i blizina). Fluentno čitanje nije
bilo moguće. Pokreti glatkoga praćenja u smjeru suprotnom od lezije bili su otežani. Sakade su manifestirale desnostra-
nu hipermetriju i lijevostranu hipometriju. Vidno polje bilo je suženo na središnjih 10 – 20 °. Postavljena je dijagnoza
Wallenbergova sindroma. Propisana je okluzija. Pregled literature pokazao je nedostatak smjernica medicine utemeljene
na dokazima za oftalmološku procjenu i liječenje oštećenja vida kod WS-a. Okulomotorne abnormlalnosti, oscilopsija i
iluzija nagiba značajno utječu na svakodnevni život. Zaključno, nakon moždanog udara nužna je rana oftalmološka
procjena kako bi se s liječenjem započelo na vrijeme.
