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Bilateral ptosis with wall-eyed bilateral internuclear ophthalmoplegia and vertical gaze paralysis

Josip Ljevak¹, Antonija Mišmaš¹, Zdravka Poljaković^{1,2}, Mario Habek^{1,2}

¹ University Hospital Center Zagreb, Department of Neurology, Referral Center for Demyelinating Diseases of the Central Nervous System, Zagreb, Croatia

² School of Medicine, University of Zagreb, Zagreb, Croatia

Corresponding author:

Mario Habek, MD, PhD

University Department of Neurology, University Hospital Center Zagreb

Kišpatićeva 12

HR-10000 Zagreb

Croatia

Phone/Fax: +38512376033; e-mail: mhabek@mef.hr

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Case presentation

A 53-year old patient presented in the neurological emergency department complaining of sudden onset of blurry vision, as well as difficulty opening his eyes. He had no previous medical history, and of significance was his body mass index of 48. Initial neurological examination showed binocular vertical double vision, bilateral ptosis with exotropia of the left eye, bilateral adduction deficit combined with abducting nystagmus (wall-eyed bilateral internuclear ophthalmoplegia, WEBINO) and vertical gaze paralysis. Bed side examination showed normal acuity and no visual field deficit. During the examination patient quickly deteriorated, becoming comatose, tetraplegic, with unreactive pupils, anisocoria (right pupil wider) and ataxic breathing. After lowering his blood pressure from 260/160 mmHg to 160/90 mmHg, his condition improved to somnolence, dysarthria, bilateral ptosis, anisocoria with medium sized fixed pupils (R>L), bilateral adduction deficit with abducting nystagmus and exotropia (more pronounced on the left eye), skew deviation (hypertropia of the left eye, presence of incyclotorsion was uncertain in the setting of bedside examination in emergency department) and vertical gaze paralysis (Figure 1, Video 1). There was no compensating head tilt and no ocular counter-roll, while vertical VOR were absent.

Urgent cerebral CT and CT angiography were normal, but MRI showed small areas of restricted diffusion in medial portions of both thalami, as well as in rostral part of mesencephalon (characteristic V sign absent) (Figure 2), all consistent with acute infarction.

Question:

Occlusion of which artery is consistent with presented clinical and MRI features?

Discussion

WEBINO is a rare syndrome consisting of bilateral exotropia, bilateral internuclear ophthalmoplegia and impaired convergence. Anatomic basis of this condition is believed to be a combination of bilateral medial longitudinal fascicle (MLF) and medial rectus subnucleus (MRSN) damage, but is still unproven (1). This most often occurs in mesencephalic stroke. Literature on this syndrome is scarce, but other etiologies are described (2), most often in young adults with multiple sclerosis.

Bilateral thalamic damage has broad differential diagnosis, including both arterial and venous stroke, tumors, infections and inflammatory lesions. Thalamus is supplied by multiple small arteries originating from posterior communicating artery (PcoA) and posterior cerebral artery (PCA), with significant variability and overlapping. For this case, it is interesting to remember a rare anatomic variant called artery of Percheron (AOP) which is a single thalamoperforating artery supplying bilateral medial thalami and sometimes rostral midbrain. Another point of interest is anterior thalamic territory, which is usually supplied by thalamotuberal arteries. However, they can also be absent, in which case AOP can also supply blood for anterior thalami as well. Lesions in this area have been reported as cause of ptosis.

Occlusion of AOP produces 4 distinctive ischemic patterns: bilateral paramedian thalamic with and without midbrain, bilateral paramedian and anterior thalamic with or without midbrain (3). In case of AOP infarction with mesencephalic involvement MRI typically shows "V sign" - axial FLAIR and DWI hyperintensity along the pial surface of midbrain in the interpeduncular fossa (3). In this case however, it was absent.

Classically, bithalamic infarctions present with triad of vertical gaze impairment, amnesia and mental changes. Additionally, as a rule of thumb, any ocular movement disorder without clear

anatomical correlation should raise suspicion for possible thalamic infarction (4), possibly with mesencephalic ischemia.

In the case described, initial symptoms seemed atypical for stroke (bilateral ptosis and oculomotor deficit, WEBINO), while sudden (albeit transient) deterioration suggested a large territory posterior circulation stroke, or even basilar artery occlusion. After reviewing the literature, and given the patency of large arteries of the brain as demonstrated on CTA, we could speculate that this deterioration was a more profound loss of consciousness caused by bithalamic ischemia. In the presence of bilateral ptosis and pupilar abnormalities, it is difficult to differentiate between WEBINO and bilateral oculomotor nerve palsy. However, rostral mesencephalic infarction caused both bilateral oculomotor nerve damage (bilateral ptosis - damage to central caudal nucleus, anisocoria), but also a lesion of MLF with clinical signs of WEBINO and combined up and down gaze paralysis. This case expands the clinical manifestations and aids in improving recognition of AOP infarction.

References

1. Chakravarthi S., Kesav P., Khurana D. (2014) Wall-eyed bilateral inter nuclear ophthalmoplegia with vertical gaze palsy. *Q J Med* 107:165.
2. Chen CM, Lin SH. (2007) Wall-eyed bilateral internuclear ophthalmoplegia from lesions at different levels in the brainstem. *J Neuroophthalmol* 27:9–15.
3. Lazzaro NA, Wright B, Castillo M, Fischbein NJ, Glastonbury CM, Hildenbrand PG, Wiggins RH, Quigley EP, Osborn AG. (2010) Artery of percheron infarction: imaging patterns and clinical spectrum. *AJNR Am J Neuroradiol* 31:1283-9.
4. Adamec I, Barun B, Mahovic Lakusic D, Ozretic D, Brinar VV, Habek M. (2011) Neuro-ophthalmologic manifestations of thalamic stroke. *Neuro-Ophthalmology* 35:121-124.

Figures

Figure 1. Screenshot of the eye movements; arrows indicate the direction of the movements.

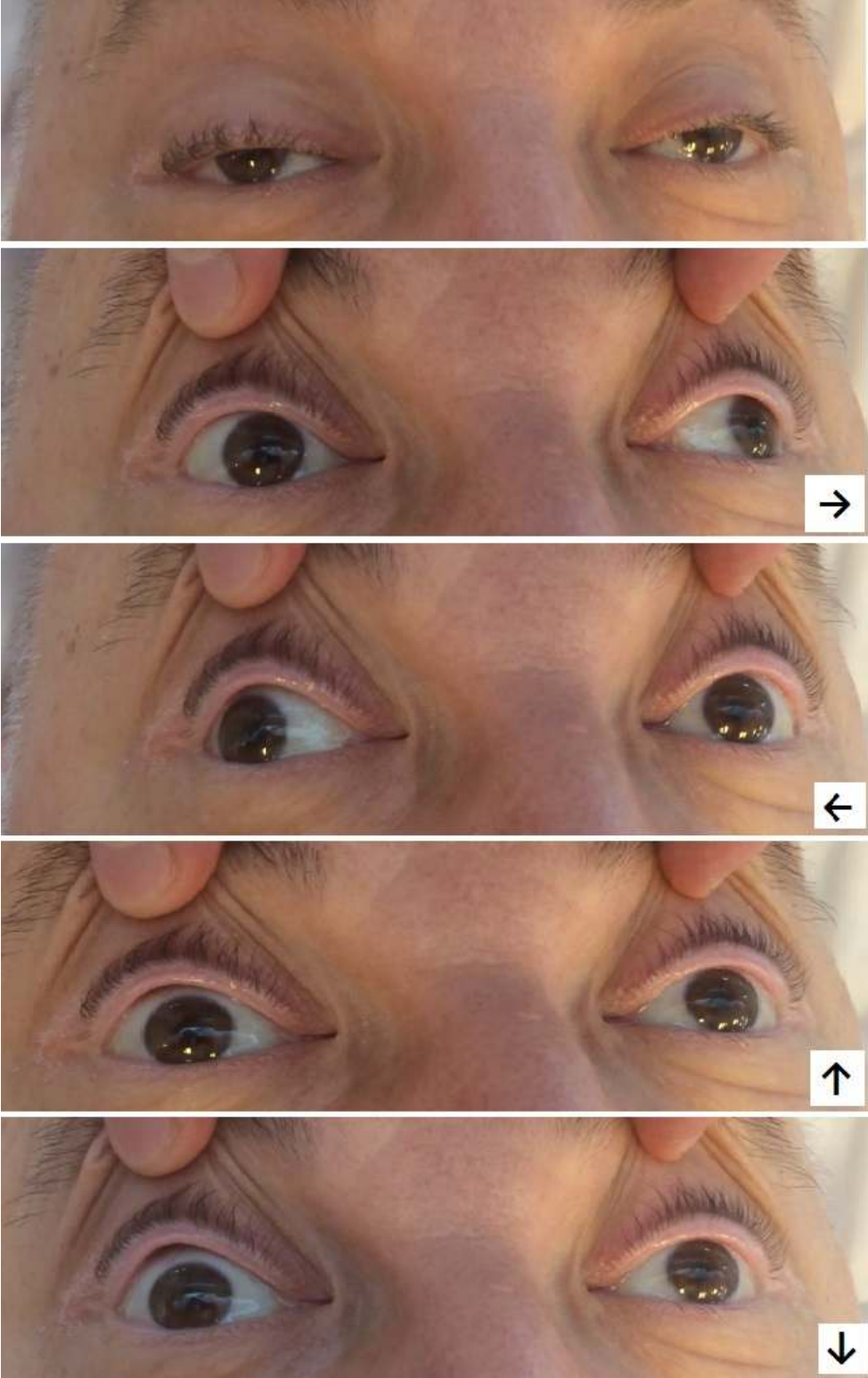
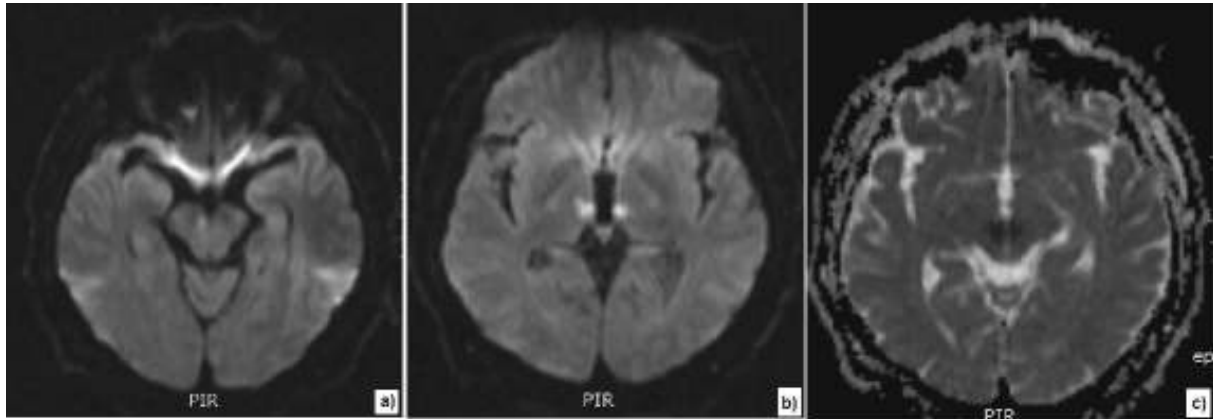


Figure 2. Brain MRI (diffusion-weighted imaging) showing acute diffusion restriction in mesencephalon (a) and both thalami (b) (hypointensities on ADC (c)).



Videos

Video 1. Showing the eye movements in the presented patient. At the beginning of the video the patient is looking straight ahead, then he is looking right, left, up and down.