Webino Syndrome Accompanied by Vertical Gaze Paralysis Related to Multiple Hemorrhagic Brainstem Foci Detected by SWI MRI

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Abstract

A 68-year-old male patient was diagnosed with the Wall-eyed bilateral internuclear ophthalmoplegia (WEBINO) syndrome accompanied by vertical gaze paralysis due to multiple hemorrhagic foci in the mesencephalon and pons as a result of hypertensive microangiopathy. The hemorrhagic foci could not be detected with standard MRI sequences, but were shown with the susceptibility-weighted imaging (SWI) sequence. Vertical gaze paralyses are not classic signs of the WEBINO syndrome but some cases with this sign have been reported. As far as we know, hypertensive microangiopathy-related hemorrhage has been identified in the etiology of the WEBINO syndrome for the very first time.

Keywords: Wall-Eyed Bilateral Internuclear Ophthalmoplegia Syndrome; Hypertensive Microangiopathy; Brain Stem Multiple Hemorrhagic Foci; SWI

Introduction

The Webino syndrome is an ocular motility disorder characterized by bilateral adduction defect, bilateral nystagmus on abduction and a large angle exotropia in primary gaze [1]. Mesencephalon or pons infarct, demyelinating diseases, toxic causes, neurodegenerative diseases, trauma, neoplasm, surgical intervention, nutritional causes, metabolic diseases, inflammation and infections can be included in the etiology [2]. Simultaneous horizontal and vertical gaze paralyses are very rare as the control centers of these eye movements are anatomically in different locations [3]. Conventional MRI and diffusion-weighed MRI (DWI) images of the midbrain and pons are helpful in the diagnosis [3,4]. We present this quite rare case of WEBINO syndrome together with vertical gaze limitation and ptosis in the right eye, caused by hemorrhagic foci at the mesencephalon or pons due to hypertensive microangiopathy that could not be detected with standard imaging sequences but were demonstrated with the SWI sequence.

Case

A 68-year-old male patient presented to the emergency service with symptoms of double vision, drooping right eyelid, headache around the right eye and in both temporal regions and dizziness that he had first noticed two days ago when he woke up in the morning. His blood pressure was 250/130 mmHg in the emergency room. Since no pathology was found in the conventional cranial MRI and DWI requested by the emergency service, the patient’s blood pressure was regulated and he was discharged. The patient who then presented to our outpatient department due to the continuation of his symptoms was hospitalized for further investigation and treatment. The personal history of the patient revealed hypertension for 15 years and one pack/day smoking for 45 years (he had quit 2 years ago). He was using valsartan+ hydrochlorothiazide 160/12.5, nebivolol 5 mg, amlodipine 5 mg and acetylsalicylic acid 150 mg once a day. His fam-
Family history revealed hypertension in both his siblings. The physical examination was normal. Neurological examination revealed isocoric pupils, light reaction +2/+2, and large angle exotropia in primary gaze. On right gaze, the right eye showed irregular nystagmus on full abduction while the left eye remained at the midline. On left gaze, the left eye showed irregular nystagmus on full abduction while the right eye had an adduction defect. Limitation of vertical gaze in both directions was present, more prominent with upward gaze, together with a conjugate gaze paralysis. Ptosis was present in the right eye (Figure 1). Other neurological system examinations were normal. Whole blood count, biochemistry, thyroid function tests, vitamin B12, folate, hemoglobin A1C and tumor markers were within normal limits. Carotid vertebral artery doppler was normal. Multiple hemorrhagic foci that could not demonstrated on conventional MRI sequences and the DWI sequence but could be seen due to a sensitivity artifact in the SWI sequence and that suggested hypertensive microangiopathy due to the location (basal ganglia, cerebellum, pons and mesencephalon) were found (Figure 2). He was diagnosed with bilateral medial longitudinal fasciculus (MLF) involvement accompanied by vertical gaze paralysis (WEBINO syndrome) due to multiple hemorrhagic foci in the mesencephalon and pons.

**Figure 1**: Ocular motility on admission. Right ptosis and wide-angle strabismus in the primary position. While the abducted eye deviates on horizontal gaze, the adducted eye does not pass the midline. Vertical gaze limitation that is more significant when looking up.

**Figure 2**: Bilateral basal ganglions and millimetric microhemorrhage foci creating sensitivity artefacts in the thalamus, left half of the pons, left cerebral peduncle and right cerebellum are observed in the SWI sequence (A, B, C). Existing foci cannot be distinguished on T1 axial, DWI and T2 axial sequences (D, E, F).
Discussion

We presented a male patient diagnosed with the WEBINO syndrome due to a bilateral MLF lesion that was caused by multiple mesencephalon and pons hemorrhages as a result of hypertensive microangiopathy. The patient clinically had nystagmus that had appeared with a bilateral adduction defect and large angle exotropia in primary gaze, together with limited upward and downward gaze and right ptosis. Other clinical signs such as vertical gaze paralyses, upbeat nystagmus and skew deviation have been reported in the WEBINO syndrome [5,6]. Vertical gaze abnormalities of varying degrees in the WEBINO syndrome are explained with involvement of the MLF in the midbrain-thalamic region accompanying the affected rostral interstitial nucleus or Cajal's interstitial nucleus [5,6]. Ptosis occurs with the 3rd nerve nucleus being affected by lesions of the midbrain-thalamic region and is accompanied by mydriasis in such cases [6]. The ptosis in the right eye was not accompanied by mydriasis in our case. This suggests a subnuclear lesion and can be explained by a right-sided midbrain lesion [7]. It has been reported that MRI may not be able to demonstrate similar functional abnormalities. Although changes in consciousness or other neurological changes such as hemiplegia have been reported in WEBINO syndrome cases due to brainstem cerebrovascular diseases, no focal neurological sign other than the ocular findings was found in our case [8]. Midbrain infarcts are the most commonly reported causes in the etiology, followed by multiple sclerosis [9]. As far as we know, hypertensive microangiopathy-related hemorrhage has been identified as an etiological factor in the WEBINO syndrome's etiology for the first time. Conventional cranial MRI and DWI are the most common imaging methods for demonstrating the midbrain lesions for the diagnosis of the WEBINO syndrome and are sufficient to determine the etiology in many cases [10]. While the multiple midbrain microangiopathy-related hemorrhages could not be observed in the MRI and DWI sequences, they were seen in the SWI sequence in our case. Although the prognosis of the WEBINO syndrome varies based on the underlying etiology, spontaneous improvement is possible in multiple sclerosis-related cases [11]. Our case was able to adduct better in the left eye on right gaze and in the right eye on left gaze at the end of the first month as well. Strabismus surgery and botulinum toxin injection have been reported as treatment options for the management of diplopia in persistent cases [11].

Briefly, the WEBINO syndrome can be accompanied by vertical gaze paralysis and multiple midbrain hemorrhages can be included in the etiology. Conventional MRI sequences as well as an SWI sequence should therefore be added when investigating the etiology of the WEBINO syndrome.

Conclusion

Here we described the case of 68-year-old man with the WEBINO syndrome accompanied by vertical gaze paralysis due to a bilateral MLF lesion that was caused by multiple midbrain hemorrhages as a result of hypertensive microangiopathy. Vertical gaze paralyses are not classic signs of the WEBINO syndrome but some cases with this sign have been reported. This finding in the WEBINO syndrome are explained with involvement of the MLF in the midbrain-thalamic region accompanying the affected rostral interstitial nucleus or Cajal's interstitial nucleus. Midbrain infarcts are the most commonly reported causes in the etiology of WEBINO syndrome, followed by multiple sclerosis. As far as we know, hypertensive microangiopathy-related hemorrhage has been identified as an etiological factor in the WEBINO syndrome's etiology for the first time. Conventional cranial MRI and DWI are the most common imaging methods for demonstrating the midbrain lesions for the diagnosis of the WEBINO syndrome. The hemorrhagic foci could not be detected with standard MRI sequences, but were shown with the SWI sequence.

In summary, the WEBINO syndrome can be accompanied by vertical gaze paralysis and multiple midbrain hemorrhages can be included in the etiology. Conventional MRI sequences as well as an SWI sequence should therefore be added when investigating the etiology of the WEBINO syndrome.

Bibliography

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