Original Contribution

BILATERAL VESTIBULAR LOSS IN VERTEBROBASILAR DOLICHOECTASIA

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Abstract — Bilateral vestibular loss (BVL) is a relatively uncommon syndrome that may produce problems of balance; unsteady gait, especially in the dark; and visual disorders and/or oscillopsia associated with walking and head movements. Sometimes patients with BVL remain asymptomatic. Ototoxic drugs are the most frequently identified cause of BVL, but there are many other possible causes. The aetiology remains unknown in a large percentage of patients. In some, vascular aetiology may be suspected. Here we report 4 cases of vertebrobasilar dolichoectasia (VBD) and symptomatic BVL. In 3 subjects, hearing was preserved, but in the 4th, there was retrolabyrinthine hearing loss. In our opinion, VBD may be the cause of BVL associated or not associated with hearing loss, the reason being that since the anterior vestibular artery is small and has no anastomoses, the horizontal semicircular canal is selectively susceptible to ischemia.

Keywords — vertebrobasilar dolichoectasia; loss of vestibular function; labyrinthine artery.

Introduction

Bilateral vestibular loss (BVL) is a relatively uncommon syndrome with an extremely variable clinical picture: some patients are asymptomatic, others experience mild loss of balance in the dark, others have visual impairment and/or incapacitating oscillopsia on active or passive movement of the head, and some suffer debilitating ataxia (1-3). The causes of BVL are numerous, the best-known being ototoxic drugs (gentamycin, streptomycin), bacterial meningitis, hydrops, and bilateral sequential vestibular neuritis (4-6). BVL may also be of familial congenital nature (7). The aetiology is often unknown, as suggested by several studies in which the idiopathic form always includes the largest group of patients (3,8). The possibility that BVL may be of vascular aetiology has been considered in very few cases (2,3).

Here we report the clinical features of 4 patients with bilateral loss of vestibular function associated with vertebrobasilar dolichoectasia (VBD) diagnosed on the basis of imaging studies.

Methods

All patients underwent CT brain scan and magnetic resonance (MR) study. One of those patients also underwent angiography. According to the criteria of Smoker and colleagues (9), the basilar artery (BA) was judged elongated if at any point along its course it lay lateral to the margin of the clivus or dorsum sellae or bifurcated above the plane of the suprasellar cistern. Ectasia was diagnosed if the diam-
eter of the artery was greater than 4.5 mm. MR was performed with a 0.5 T superconducting magnet unit using spin-echo, multi-echo, and FAST techniques. The images were obtained in sagittal, axial, and coronal planes. T2-, proton density, and T1-weighted images were obtained. Slice thickness was 5.0 mm with a 2.0-mm interval between successive slices.

Examination of vestibular function was carried out using Frenzel glasses for spontaneous-positional and positioning nystagmus and then by DC electro-oculography for gaze, saccades, smooth pursuit, and caloric test. Dynamic imbalance of the vestibular system was also assessed by head shaking and head thrust tests (10) and by dynamic eye chart testing which consisted of reading a standard Snellen eye chart at a distance of 3 m. Visual acuity was measured in static conditions and while the patient oscillated his or her head left to right in a 60° arc at a frequency of about 1 Hz (11). Eye movement recordings were performed with an automated electronystagmography package (Siter-Racia, Bordeaux). Gaze-evoked nystagmus was induced by having the patient fix gaze on a target (small red light-emitting diodes) 30 degrees to the right, to the left, above, and below the center position. Rebound nystagmus was also evaluated on returning the eyes to the primary position.

Saccadic movements were induced by random step changes in target position (small red light-emitting diodes). The resting time of the stimulus at each position was randomly varied in the range from 1 to 2.5 s. Parameters were accuracy and velocity of ±10° and ±30° horizontal saccades and ±10° vertical saccades. Five saccades for each direction were evaluated. Saccade accuracy was defined as saccade amplitude/target × 100. Dysmetria was diagnosed if saccade accuracy was less than 85% or more than 100%. Horizontal smooth pursuit movements were induced by a target moving sinusoidally ±20° from midposition at a frequency of 0.2 Hz. Normal range of gain (target velocity/eye velocity) was > 0.6.

Bithermal (30 °C and 44 °C) caloric testing was performed according to the Fitzgerald and Hallpike (12) technique. Slow phase velocity (SPV) and frequency of nystagmus were evaluated. Bilateral paresis was diagnosed when the SPV was less than 5°/s for all responses (13). Patients underwent postrotational stimulation (sudden stop after constant velocity rotation of 120°/s for 120 s) clockwise and counterclockwise. Rotational tests were performed in darkness with the subject's eyes open. Maximum SPV and the time constant of the response were evaluated (14).

Auditory function was evaluated by pure tone audiometry, auditory brain stem response (ABR) testing, and tympanometry and stapedius reflex (SR). ABR was performed with an Amplaid MK15 apparatus. Surface Ag/AgCl electrodes were placed on the brow (ground), mastoid (exploring electrode), and vertex (reference electrode). Single sine wave clicks of 100 μs duration and a frequency of 21 clicks/s were delivered through earphones. Stimulation was repeated 2,000 times. Analysis time was 12 ms. Criteria for abnormal ABR were I-III interpeak latency ≥ 2.5 ms; I-V interpeak latency ≥ 4.5 ms; III-V interpeak latency ≥ 2.3 ms.

Case Reports

Case 1

Male, 65 years of age, diabetic on oral hyperglycemia-reducing drugs for the last 10 years. Slowly progressive problems of balance, especially in the dark, in the last 2 years, with recurrent positioning vertigo (when rising and going to bed) in the last 6 months, and oscillopsia when walking or moving the head in the last 2 months. Recent transient episode of diplopia. Curiously, the first symptoms occurred while the subject was hunting: an ardent hunter, he noted having trouble following the sudden changes in direction of a fleeing hare. Neurological examination showed Romberg sign and severe gait ataxia that worsened with eyes shut. Imaging studies showed multiple bilateral lacunar infarcts in basal ganglia, white matter, and brain stem (midbrain and rostral pons). The BA was elongated, ectatic (diameter ranging from 4.8 to 5.8 mm) and tortuous, its proximal part being displaced to the
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left, its cranial part being in right paramedian position. The bifurcation of the BA indented and elevated the floor of the third ventricle (Figure 1).

Audiometric examination showed bilateral sensorineural hearing loss, reducing at higher frequencies and more accentuated on the left. ABR testing showed abnormal prolongation of the I–III and I–V intervals on the left. Bedside examination of vestibular function showed symmetric little gaze paretic nystagmus and limitation of upward eye movements. Rapid rotation of the head (head thrust) to either side revealed evident compensatory re-fixation saccades (Halmagyi’s sign). No nystagmus occurred with head shaking. Dynamic eye chart testing showed complete loss of visual acuity. ENG revealed symmetrical impairment of smooth pursuit reflex and vertical slowing of upward saccades with a peak velocity of about 100°/s, which is one-third of the normal value. There was no response to bithermal caloric stimulation, nor did ice water or post-rotatory stimulation reveal any nystagmus.

Case 2

Female, 63 years of age, with recurrent depression treated with amitryptyline. Problems of balance in the last 6 months, oscillopsia while walking (“objects move when I walk”) for the last 2 months. Neurological examination showed only gait ataxia. Imaging studies revealed multiple lacunar infarcts in the white matter of both hemispheres and mild cerebellar atrophy. The BA was elongated, ectatic (diameter ranging from 4.8 to 6.0 mm), and tortuous, passing from a right paramedian position in its proximal part to a position much more to the left in its cranial part and finally terminating 15 mm above the dorsum sellae. The bifurcation of the BA indented and elevated the floor of the third ventricle (Figure 2).

Tonal audiometry and ABR were within normal limits. Bedside vestibular examination revealed pathological response to bilateral head thrust test and a 5-line deterioration of visual acuity during oscillation of the head. No spontaneous or positional nystagmus was

Figure 1. Case 1. MR scan in sagittal plane (TR 400/TE 14 ms). The bifurcation of the basilar artery is compressing the floor of the third ventricle.
found. ENG showed symmetrical impairment of smooth pursuit reflex. Vertical saccades were slow and difficult to initiate. Bithermal caloric stimulation, including ice water and postrotational testing showed complete absence of horizontal vestibulo-ocular reflex (VOR).

Case 3

Male, 67 years of age. Three episodes of spinning vertigo with vomiting lasting several hours, over a period of 17 years. Last episode 6 months earlier. Sporadic episodes of positioning vertigo (on rising and going to bed) in the last 10 years and problems of balance, particularly at night, and blurring of vision without definite oscillopsia in the last 5 months. Vestibular examination 5 years earlier was normal, and one performed at the last episode of dizziness showed right-beating spontaneous nystagmus and caloric left vestibular paresis. Neurological examination was negative. Imaging studies revealed multiple lacunar infarcts in the white matter of both hemispheres and mild cerebellar atrophy. The BA was elongated, ectatic (diameter ranging from 4.8 to 5.4 mm), and tortuous, its proximal part being displaced considerably to the left, its cranial part being in right paramedian position. The bifurcation of the BA was at the level of the floor of the third ventricle.

Bilateral sensorineural hearing loss with features of presbyacusia was found. ABR was abnormal on the left, with prolonged I-III and I-V intervals. No spontaneous or positional nystagmus was found. Head thrust provoked corrective saccades, and oscillation of the head caused a 3-line deterioration of visual acuity. Saccades were normal, but smooth pursuit was symmetrically impaired with the highest frequency stimulus. Bithermal caloric stimulation evoked no response, whereas ice water and postrotation showed a few beats of nystagmus.

Case 4

Male, 72 years of age. In the last 7 years, 5 reversible ischemic attacks with right hemifacial paraesthesia, hyposthenia of right limbs, and dysarthria. Recurrent episodes of spinning
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vertigo and continuous imbalance, especially in the dark, for the last 2 months. Neurological examination showed Romberg sign and severe gait ataxia. Imaging studies showed multiple bilateral lacunar infarcts in the basal ganglia and white matter. The BA was elongated, ectatic (diameter ranging from 5.6 to 7.8 mm), and tortuous, its proximal part being displaced to the right, its intermediate part considerably to the left, and its more cranial part to left paramedian position. The bifurcation of the BA indented and elevated the floor of the third ventricle. Angiographic examination also showed ectasia of the internal carotid and vertebral arteries (Figure 3).

Audiometric examination showed mild bilateral symmetrically sloping sensorineural hearing loss above 2000 Hz. ABR were normal. Vestibular examination revealed clearly positive bilateral Halmagyi's sign. No head shaking or spontaneous or positional nystagmus was found. Saccades and pursuit were normal. Caloric responses, including to ice water, were absent bilaterally. Post-rotatory response was almost absent (a few beats of nystagmus).

Discussion

The diagnosis of BVL is relatively simple. Bithermal caloric stimulation shows a reduction or absence of the expected nystagmus. When the response is completely absent, it is necessary to use more intense stimulation of the ear with ice water (15). Although caloric unresponsiveness generally indicates a functional defect, it does not necessarily indicate an effective absence of vestibular function, as caloric stimulation is equivalent to low frequency stimulation and only enables a narrow range of vestibular dynamics to be explored (16). Moreover, patients with caloric areflexia may also have a normal VOR for low frequencies of rotatory stimulation (5,8,17,18). Although we could not perform rotatory testing at variable frequencies, the post-rotatory stimulus we used (an acceleration of about 100°/s²) was sufficient to confirm the diagnosis of BVL in our patients, particularly in view of the symptoms and the results of the other clinical tests.

All patients complained of imbalance, especially in the dark. Two had typical oscillopsia with any movement of the head, and one complained of blurred vision when he moved. Dynamic oscillopsia, illusory movement of the visual scene during head movements, is due to failure of the VOR, which does not maintain the image of a stationary target on the retina. VOR is the largest contributor to compensatory eye movements for head movements having a sine wave frequency from 1 to 4 Hz, which is the frequency band of head movement during walking and running (19).

It is known that BVL does not necessarily accompany oscillopsia and that the symptoms may attenuate or resolve in time. This may be due to adaptive mechanisms such as potentiation of the cervico-ocular reflex, preprogramming of compensatory eye movements, and perceptual changes (1,19–21).
Head thrust testing and dynamic testing of visual acuity are two simple and reliable bedside tests that enable evaluation of vestibular function. With head thrust, the 4 patients showed clear compensatory saccades. The test, described by Halmagyi and Curthoys (10), is of proven reliability in cases of severe monolateral and bilateral canal paresis (3). The rapid passive acceleration of the head (head thrust) causes corrective saccades, easily detected by the examiner, towards the fixation target. Of the 4 patients, 2 also showed marked deterioration of visual acuity with dynamic eye chart testing, confirming the VOR deficit (5,11). One patient had a 3-line decrease, which in our experience is borderline. In one patient, the test was not performed.

The best-known cause of BVL is ototoxic damage, but in the majority of cases, no cause can be identified. The possibility of vascular causes is controversial. Brandt (3) maintains that some cases of idiopathic BVL may be ischemic in character, whereas Baloh and colleagues (2) state that “the complete loss of vestibular function with preserved auditory function is difficult to explain on an ischemic basis”. In any case, among the 22 patients with idiopathic BVL described by Baloh, I had severe occlusive pathology of both vertebral arteries. In the series of 22 patients with BVL described by Banshali and colleagues (5), I had dolichoectasia of the intracranial vertebral artery (sudden left deafness and left facial weakness). Büttner and colleagues (22) recently described a case of sequential bilateral loss of cochlear and vestibular function attributable to bilateral ischemia as a complication of VBD, and Demanez and Demanez (23) described 1 case of BVL in 7 cases of VBD and otovestibular symptoms. Other authors indicate vertebrobasilar failure as a possible cause of BVL (3,5,11,15).

Vertebrobasilar dolichoectasia may cause a series of clinical manifestations by mechanisms such as ischemia and compression. Compression may induce isolated or multiple cranial nerve involvement (9,24-26), dysfunction of the caudal brain stem (9,25,27), and hydrocephalus (28); ischemia may produce a variety of brain stem and cerebellar symptoms and signs (29-32).

In our patients, BVL can hardly be attributed to bilateral compression of the vestibulocochlear nerve. In these cases the mechanism responsible may be central dysfunction due to ischemia or a pressure effect of the ectatic basilar artery on the brain stem, but a more likely cause is bilateral impairment of perfusion of labyrinth structures. The membranous labyrinth and its neural structures are irrigated by the labyrinthine artery that, shortly after entering the inner ear, divides into two branches, the common cochlear artery (CCA) and the anterior vestibular artery (AVA). The AVA supplies the upper part of the vestibular labyrinth including the horizontal semicircular canal and the utricular macula. It is smaller than the CCA and has no anastomoses, so that the horizontal semicircular canal is selectively susceptible to ischemia (33,34).

In cases 1 and 2, vestibular loss probably developed from slowly progressive impairment of blood perfusion of the two AVA, without causing any asymmetry between the two vestibular inputs, as suggested by the absence of episodes of vertigo. The episodes of positional vertigo experienced by patient 1 (and patient 3) may have been due to the detachment of otoconia from the utricular macula on an ischemic basis leading to cupulolithiasis or canalolithiasis, as the posterior semicircular canal was still functional. However, since positional vertigo is very common, the apparent relationship may be a coincidence. In patients 3 and 4, the episodic symptoms of vertigo suggest that BVL developed as a consequence of sequential ischemic episodes in districts supplied by the AVA. The alteration of upward eye movements in patients 1 and 2, of ABR in cases 1 and 3, and of smooth pursuit in cases 1, 2, and 3 suggest the coexistence of brain stem/cerebellar dysfunction, as can be expected in patients with this type of vascular disease.

In conclusion, our study suggests that BVL is not a rare finding in patients with VBD. In patients, especially elderly ones, who have a clinical picture compatible with BVL, it is worth considering the possibility that the complaint may be due to vertebrobasilar dolichoectasia. This is also of therapeutic importance because, besides vestibular rehabilitation (35), patients...
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with BVL associated with VBD may also require treatment to prevent ischemic complications and to improve microcirculation.

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REFERENCES