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ORIGINAL COMMUNICATION

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Rotational vertebral artery syndrome

3D kinematics of nystagmus suggest bilateral labyrinthine dysfunction

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Abstract Whether the rotational vertebral artery syndrome (RVAS), consisting of attacks of vertigo, nystagmus and tinnitus elicited by head-rotation induced compression of the dominant vertebral artery (VA), reflects ischemic dysfunction of uni- or bilateral peripheral or central vestibular structures, is still debated. We report on a patient with bilateral high-grade carotid stenoses, in whom rightward head-rotation led to RVAS symptoms including a prominent nystagmus. Three-dimensional kinematic analysis of the nystagmus pattern, recorded with search coils, revealed major downbeat nystagmus with minor horizontal and torsional components. Magnetic resonance angiography demonstrated a hypoplastic right VA terminating in the posterior inferior cerebellar artery, a dominant left VA, and a hypoplastic P1-segment of the left posterior cerebral artery (PCA) that was supplied by the left posterior communicating artery (PCoA). The right PCA and both anterior inferior cerebellar arteries were supplied by the basilar artery. The right PCoA originated from the right internal carotid artery. Color duplex sonography showed severe reduction of diastolic blood flow velocities in the left VA during RVAS attacks. The nystagmus pattern can be best explained by vectorial addition of 3D sensitivity vectors of stimulated right and left anterior and horizontal semicircular canals with slightly stronger stimulation on the left side. We hypothesize that in RVAS, compression of dominant VA leads to acute vertebrobasilar insufficiency with bilateral, but asymmetric ischemia of the superior labyrinth. With regard to RVAS etiology, our case illustrates a type of pure vascular RVAS. Severity of attacks markedly decreased after successful bilateral carotid endarterectomy.

Key words vertebral artery compression · vertigo · labyrinthine ischemia · downbeat nystagmus

Introduction

Rotational vertebral artery syndrome (RVAS) is defined by recurrent attacks of vertigo, nystagmus, ataxia, and tinnitus which are elicited by fierce head rotation [1–4]. The presumed mechanism leading to RVAS is transient compression of the dominant vertebral artery (VA) by the rotational head movement, which leads to hemodynamic ischemia in the vertebrobasilar territory as the blood supply through the opposite hypoplastic VA is not sufficient. VA compression is often enhanced by anatomical obstacles such as cervical spondylosis or muscular insertions [1, 2, 5].

However, whether the RVAS results from ischemia of uni- or bilateral labyrinthine structures or of the central

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vestibular system remains unclear [2, 4]. Compression of the dominant VA is expected to impair the blood supply to the peripheral as well as the central parts of the vestibular system. On the other hand, clinical symptoms and signs of this syndrome suggest primarily a peripheral vestibular disorder. Indeed, recent ocular motor studies in patients with RVAS documented a mixed downward-horizontal nystagmus pattern during the attacks, suggesting a dysfunction of the superior labyrinth[1] on the side of VA compression [2–4]. But, since the labyrinth is irrigated by the labyrinthine artery (LA, also known as the internal auditory artery), which is a branch of the anterior inferior cerebellar artery (AICA), and since both AICA usually originate from the basilar artery (BA) [6], one would expect that compression of the dominant VA and the consecutive impairment of blood flow in the BA leads to bilateral labyrinthine dysfunction.

In the present paper, we report on a patient with a well-documented vascular-type RVAS. Kinematic analysis of three-dimensionally recorded nystagmus revealed a strong upward and only minor horizontal and torsional drift components. The observed drift pattern most probably reflects a bilateral excitation of the superior labyrinth with the stimulation being slightly stronger on the side of the VA compression. This concept of a bilateral, but asymmetric labyrinthine ischemia is plausible also in terms of the blood supply to the inner ear structures.

Case report

This 61 year old patient suffered from severe coronary artery disease and peripheral artery disease IIb. He underwent coronary stenting several times, and aortic valve replacement and coronary bypass surgery 3 years ago. Vascular risk factors included hypertension, diabetes mellitus, cigarette smoking, and hypercholesterolemia. Over the past 12 months the patient had experienced the following symptoms that stereotypically occurred when he turned his head more than 20° to the right side: Dizziness, blurring of vision, unsteadiness of gait, pressure-like headaches on the right side, and a feeling to start fainting. All symptoms ceased immediately upon moving the head back into the straight-ahead position. The provocative head position was not tolerated for more than a few seconds. The symptoms did not occur in any other head position. A week after onset of the RVAS attacks, the patient experienced an attack of amaurosis fugax. Color duplex sonography (CDS) revealed a high-grade stenosis at the origin of the right internal carotid artery (ICA). He underwent right carotid endarterectomy (CEA), and RVAS did not improve postoperatively.

Four months after CEA, the patient presented in our hospital because of the RVAS attacks. Neurological examinations performed between the attacks were unremarkable. The patient was repeatedly examined in the challenging head position and the followings symptoms and signs were always observed: A prominent downbeat nystagmus with – as seen from the patient – a left-beating horizontal and a counter-clockwise-beating torsional component (i.e. upper eye poles rotating towards the left ear) occurred after a few seconds. The nystagmus persisted over the few seconds, during which the patient was able to keep the challenging head position, and disappeared within one or two seconds after the patient had moved the head into the straight-ahead position.

As shown in Fig. 1, magnetic resonance angiography (MRA) demonstrated a dominant left VA and hypoplastic right VA that ended in the posterior inferior cerebellar artery (PICA). The left posterior cerebral artery (PCA) was supplied mainly by the left posterior communicating artery (PCoA) due to hypoplasia of the precommunicating (P1) segment of the left PCA, while the right PCA was supplied by the basilar artery (BA). The right PCoA was supplied by the right ICA. Both anterior inferior cerebellar arteries (AICA) originated from the BA.

While the patient turned his head into the challenging position and experienced the described symptoms, insonation of the pars transversaria of the left VA with a handheld linear probe revealed a resistance profile with absent diastolic blood flow velocities (Fig. 2A) compared to the normal blood flow velocities in the neutral head position (Fig. 2B).

An MRA in the challenging position of the head could not be performed, because the patient was unable to tolerate the RVAS symptoms for more than a few seconds. Cerebral MR imaging (MRI) showed general moderate brain atrophy, a few lacunar infarcts in both hemispheres and, to a lesser degree, in the brainstem. X-ray and computed tomography of the cervical spine showed mild osteochondrosis on segment C5/6, but no osteophytes compressing the VA.

About two months after our initial clinical examination, the patient experienced TIAs with weakness of the right arm and face. CDS diagnosed now bilateral high-grade carotid stenoses (left > 85%, right 70%) with cross-flow to the left MCA through the ophthalmic and the anterior communicating arteries, and cross-flow to the right MCA through the posterior communicating artery (PCoA). He underwent left CEA without complication, but the characteristics of RVAS attacks did not change postoperatively. Stenting of the asymptomatic right carotid restenosis was performed without complication five months later. CDS performed two days and three months after right ICA stenting revealed normal findings in the stented right carotid artery and a left

[1] The superior labyrinth comprises the anterior and horizontal semicircular canals and the utricle and is supplied by the anterior vestibular artery (AVA), a branch of the internal auditory artery (LA).

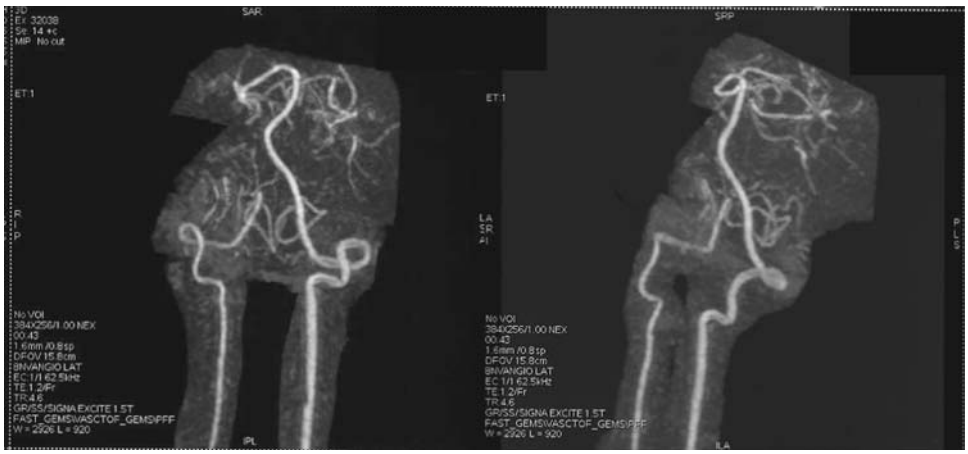

Fig. 1 Magnetic resonance angiography (MRA) of the cerebral arteries. The left vertebral artery (VA) is dominant; the right VA is hypoplastic and terminates in the posterior inferior cerebellar artery (PICA). The left posterior cerebral artery (PCA) is mainly irrigated by the left posterior communicating artery (PCoA); the right PCA and both anterior inferior cerebellar arteries are supplied by the basilar artery

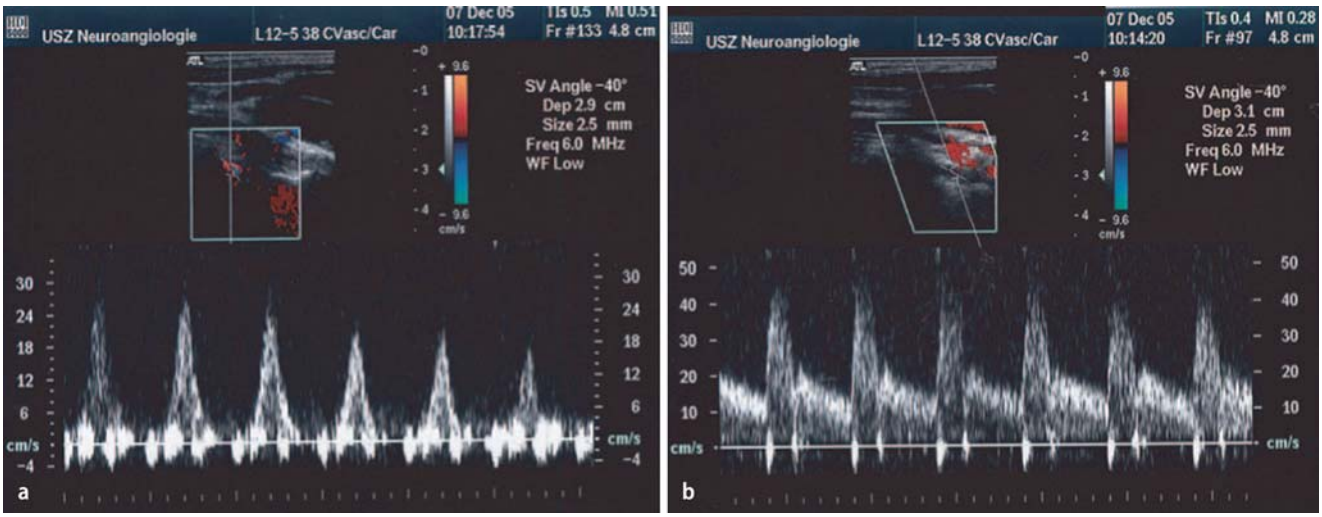

Fig. 2 Color duplex sonography of the pars transversaria of the left vertebral artery (VA) with normal spectral Doppler findings when the head is in neutral position (**a**). Head rotation to the right reduced blood flow to systolic spikes, indicating a strong increase of peripheral resistance (**b**), while the patient experienced the symptoms of the rotational vertebral artery syndrome

50% carotid stenosis as well as the disappearance of the right PCoA collateral, whereas the posterior cerebral circulation findings remained unchanged compared to the examination performed at presentation. However, only mild RVAS symptoms occurred postoperatively just when the patient turned his head maximally to the right, while CDS demonstrated a mild blood flow velocity reduction in both PCA.

Ocular motor findings

Eye movements were recorded with magnetic search coils after the first CEA of the right ICA. Fig. 3 shows horizontal (dark grey line), torsional (grey line) and vertical (black line) eye positions in the challenging head position. Positive values indicate, from the patient's point of view, upward, rightward and clockwise directions (i.e. upper eye pole rotating towards the right ear). While the patient noticed onset of typical symptoms, a strong downbeat nystagmus with a small left- and counterclockwise-beating component occurred. Fig. 4 depicts the time course of horizontal (dark grey triangles), torsional (grey squares), and vertical (black circles) ocular drift velocity (y-axis) with the head in the provocative position. Vertical ocular drift velocity increased considerably, while horizontal drift velocity only showed a moderate increase and torsional drift velocity almost remained stable.

Discussion

The RVAS comprises recurrent attacks of vertigo, nystagmus, ataxia, and tinnitus, which are elicited by head-rotation induced compression of the contralateral

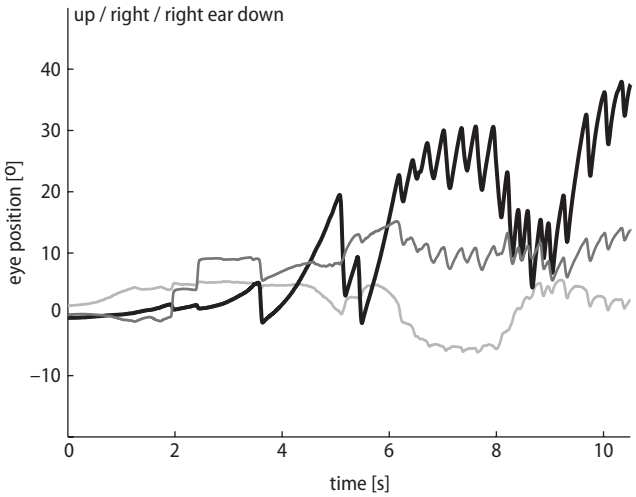

Fig. 3 Nystagmus during a typical rotational vertebral artery syndrome attack. Eye positions (horizontal: dark grey; torsional: grey line, vertical: black line) a few seconds after rightward head rotation. Positive values indicate, from the patient's point of view, upward, rightward and clockwise directions (i.e. upper eye pole rotating towards the right ear). Recording stops when the patient turned his head back to straight-ahead position, when he could not tolerate the provocating head position any longer

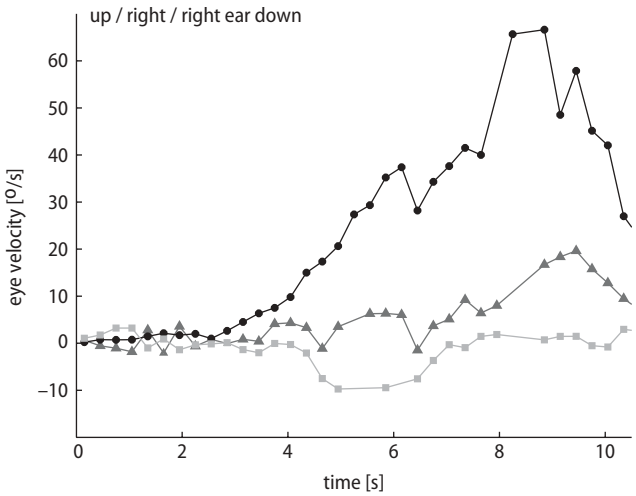

Fig. 4 Time course of ocular drift velocity during a typical rotational vertebral artery syndrome attack. Each point represents the mean velocity of one nystagmus slow phase. Positive directions as in Fig. 1. Y-axis: ocular drift velocity (deg/s; horizontal: dark grey triangles; torsional: grey squares, vertical: black circles); abscissa: time (s)

dominant VA, leading to acute reduction of blood flow in the vertebrobasilar territory [1–5, 7]. Whether the RVAS attacks are caused by ischemia-induced dysfunction of peripheral or central vestibular and auditory structures, is unresolved. We present a patient with RVAS attacks occurring during rightward head rotations that compressed the dominant left VA as documented by ultrasound (Fig. 2). Detailed kinematic analysis of the head rotation induced nystagmus, recorded with magnetic search coils, revealed prominent downbeat nystagmus (DBN), but only minor horizontal and torsional nystagmus components directed towards the side of VA compression (leftward and ipsitorsional). Our results suggest that RVAS is caused by bilateral, but asymmetric superior labyrinth dysfunction that was presumed to be more prominent on the side of the VA compression.

A similar head-rotation induced nystagmus pattern with a strong DBN and smaller horizontal and torsional components has been described in other RVAS patients [2, 4]. Eye movement recordings in these patients were performed with three-dimensional video-oculography. The pattern of the ocular drift was explained with a unilateral stimulation of the anterior and horizontal SCC on the side of the VA compression, most probably caused by a transient ischemia in the superior part of the respective labyrinth [2, 6]. From studies in the bullfrog it is known that ischemia depolarizes the membrane potential of SCC afferents, which initially leads to an excitation, but with persisting ischemia over minutes and further increase of the membrane potential, axons may become inexcitable [8]. Because in our patient as well as in earlier reported cases, RVAS symptoms in the challenging head position started only after a short delay of a few seconds, they are most probably a consequence of excitation, not to inhibition, of the SCC afferents [2]. The directional reversal of the nystagmus, which was observed in some patients who could maintain the provocative head position, may be explained by a transition from excitation to inhibition of the SCC afferents because of ongoing ischemia [4].

The presumed triggering mechanism underlying RVAS, i.e. compression of the dominant VA, will impair blood flow in the BA and therefore in both AICA and LA and may thus cause bilateral dysfunction within the central or peripheral vestibulo-auditory system. A central vestibular origin of the RVAS symptoms in our and other reported patients, however, is improbable for several reasons. First, with brainstem ischemia, additional neurological signs and symptoms are likely to occur, e.g. dysarthria or ataxia [5, 9]. Neither in our patient nor in other reported cases were there any clinical findings typical for a lower brainstem ischemia during the attacks [2].

The vascular anatomy of the peripheral and central vestibular system may explain the selective vulnerability of the labyrinth to ischemia. The VA irrigates the vestibular nuclei by its main branch, the posterior inferior cerebellar artery (PICA), and direct perforators [9]. The PICA irrigation of the vestibular nuclei is well collateralized. The labyrinthine artery (LA) is a branch of the anterior inferior cerebellar artery (AICA), which originates from the basilar artery (BA). The LA divides into the common cochlear artery, forming the posterior vestibular artery and the main cochlear artery, and the anterior vestibular artery, which supplies the superior

labyrinth. The latter artery has only sparse intraosseous collateralization, which may explain the susceptibility of the superior labyrinth to ischemia [6, 9, 10].

Hypothetically, since another end branch of the AICA supplies the cerebellar flocculus and since floccular lesions commonly lead to strong DBN [11], a floccular origin of the RVAS nystagmus has to be considered as well. However, the presence of considerable horizontal and torsional drift components during gaze straight-ahead speaks against this cerebellar mechanism, as floccular lesions typically evoke marked upward drift without consistent horizontal or torsional drift components in straight-ahead gaze [11]. Instead, the mixed nystagmus pattern observed in our patient is more characteristic for a peripheral vestibular origin (for review, see [12]) and can best be explained by a bilateral, but slightly asymmetric stimulation of the superior labyrinth: (1) Summation of upward drift components evoked by stimulation of left and right anterior SCCs accounts for the strong DBN. (2) The small leftward and counterclockwise directed torsional drift components indicate that stimulation of the horizontal SCCs and the utricles was slightly stronger on the left side. (3) A strict unilateral stimulation of the anterior and horizontal SCCs can not explain the drift pattern seen in our patient, since in this case, one would expect a mixed nystagmus with vertical and horizontal drift components being of similar magnitude [13]. (4) Finally, a symmetric bilateral labyrinth stimulation is unlikely in our patient, because this condition would evoke pure DBN, the horizontal and torsional components from both sides cancelling each other [13].

Our patient represents a rare case of a 'pure' vascular-type RVAS: As shown in the MRA (Fig. 1), both AICA are supplied by the BA, which is irrigated by the left VA. In case of head turning with compression of the left VA, blood flow to the BA and both AICAs is reduced, and cross-flow through the right PCoA to the BA might have been prevented or reduced due to the presence of the severe right carotid stenosis. After right carotid stenting, head-tilting induced reduction of vertebro-basilar blood flow might have been better compensated by cross-flow through the right PCoA to the BA.

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