CASE REPORT

Subclavian steal syndrome: neurotological manifestations

La sindrome del furto dell’arteria succavia

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SUMMARY

The subclavian steal syndrome is characterized by a subclavian artery stenosis located proximal to the origin of the vertebral artery. In this case, the subclavian artery steals reverse-flow blood from the vertebrobasilar artery circulation to supply the arm during exertion, resulting in vertebrobasilar insufficiency. As the vertebrobasilar arterial system feeds both the peripheral and central auditory and vestibular systems, in subclavian steal syndrome, neurotological symptoms are expected because of the vertebrobasilar insufficiency. In this report, we describe three patients suffering from subclavian steal syndrome, who presented with isolated dizziness, recurrent vertigo, hearing loss and tinnitus. In two of the three cases, a positional nystagmus was detected, which was vertical in two. Abnormal saccades were documented in one, and the auditory brainstem responses were pathological in all three patients; the caloric response was reduced in only one case. Upon magnetic resonance imaging, ischaemic lesions were observed in two patients, in the brainstem and in the hemispheres, respectively. These findings suggest that the central auditory and vestibular system is more likely to be involved in the pathogenesis of neurotological symptoms in subclavian steal syndrome. Patients complaining of numbness of the upper arm and isolated neurotological symptoms should be thoroughly examined for subclavian steal syndrome. Furthermore, regular follow-up must be undertaken in order to prevent other neurological deficits in the vertebrobasilar arterial territory.

KEY WORDS: Vertigo • Sensorineural hearing loss • Subclavian steal syndrome • Vertebrobasilar insufficiency

Introduction

The subclavian steal syndrome (SSS) refers to a vascular disorder in which occlusion or stenosis of the subclavian artery proximal to the vertebral artery origin (which is the subclavian artery) causes altered vascular haemodynamics that result in retrograde blood flow in the ipsilateral vertebral artery toward the upper arm, distal to the subclavian artery narrowing, where decreased blood pressure had been established. Subclavian artery lesions are usually asymptomatic because of the abundant collateral blood supply in the head, neck, and shoulder. However, these lesions produce neurologic symptoms when compensatory flow to the subclavian artery from the vertebral artery diverts too much flow toward the arm and away from intra-cranial structures leading to vertebrobasilar insufficiency (VBI).

The most important collateral circulation to the posterior fossa is through the circle of Willis, principally through the posterior communicating artery. When this communication is absent or inadequate, possibly due to concurrent
extracranial carotid stenosis, then vertebrobasilar symptoms become manifest. The rate of SSS is estimated at 1.3% (324 cases in 25,000 persons) in European patients referred for carotid and vertebral artery Doppler ultrasound, most frequently in Caucasians because of the increased incidence of atherosclerosis in this population. SSS generally occurs in patients > 55 years of age and has a 2:1 male-to-female ratio. Clinically, SSS may be suspected when the difference in blood pressure between the two arms exceeds 20 mmHg, but the diagnosis is essentially confirmed by Doppler ultrasound, although magnetic resonance imaging (MRI) with or without magnetic resonance angiography (MRA), computed tomography (CT) scan of the brain and digital subtraction angiography can also be used. Symptomatic patients require surgical intervention such as carotid-subclavian bypass, axillo-axillary bypass, or percutaneous transluminal angioplasty of the subclavian artery with stent placement. Thus, the goals of surgical treatment consist in restoration of the antegrade vertebral artery flow, alleviation of cerebral hypoperfusion, and improvement of arterial perfusion to the upper arm. Moreover, aggressive management of risk factors, such as hypertension, diabetes and tobacco use, is also essential for successful treatment of this syndrome.

The present report deals with 3 patients suffering from SSS, who were admitted to our unit on account of neurotological symptoms.

Case reports

Case 1

A 68-year-old white male, one week after a cardiac bypass surgery, developed numbness in the left upper arm and, two months later, unsteadiness and instability especially while walking, which still persists to this day. He had no auditory disturbance or tinnitus. Digital subtraction angiography revealed SSS on the left side, stenosis at the origin of the left vertebral artery and stenoses of both the carotid bifurcations. At neurological examination, when performing Unterberger testing, he deviated towards the left side. Audiometric evaluation showed a relatively symmetric high-frequency sensorineural hearing loss in contrast to the patient’s impression and the neurological Auditory Brainstem Responses (ABR) using 120 dB SPL click stimulus demonstrated a delayed wave V and increased I-V latency on the left side (Fig. 1). In the electronystagmography (ENG), there was no spontaneous nystagmus, but in the position of Rose (supine position, head hanging down and rotated to the left, middle, right, with eyes closed), a positional vertical up-beating nystagmus was present (Fig. 2). Caloric testing was normal on both sides. MRI revealed ischaemic lesions in the area of the brainstem (Fig. 3).

Case 2

For 3 years now, a 53-year-old male has been complaining of tinnitus in the left ear and slight unsteadiness. At the same time, he noticed a sensation of numbness on the right upper arm. Digital subtraction angiography revealed SSS, on the right side. His past history revealed...
that he had experienced an episode of dizziness 5 years earlier, lasting for 3 months, related to the movements of his head, with difficulty in visual fixation. At neurological examination, the patient exhibited a slight deviation towards the right side upon the Unterberger testing. Pure tone audiogram was within the normal limits and the tinnitus was located at the low frequencies in the left ear. During ENG, no spontaneous nystagmus was found and, in the position of Rose 4, a positional vertical downbeating nystagmus was noted with the head turned left, middle and right. The horizontal saccades were abnormal (Fig. 4), and the caloric response was bilaterally normal. Neurological ABR using 130 dbSPL click stimulus showed remarkably desynchronized traces, on both sides. The MRI demonstrated distinct lesions in the white matter of the hemispheres, possibly of vascular origin.

Case 3
A 59-year-old female was submitted to surgery, in February 2001, for herniated disk at C-5 level and, 6 months later, she felt fatigue both while having a bath and getting dressed. On echo-Doppler, the scalenus anticus syndrome (SAS) was diagnosed bilaterally, as a partial obliteration of radial pulse was observed when the arm was elevated to 80° on the right side and 90° on the left side. The following month she was admitted to our unit for recurrent vertigo, accompanied by hearing loss and pulsatile tinnitus, in the left ear. SSS was revealed by digital subtraction angiography. at this time, the tonal audiometry showed a moderate low-frequency sensorineural hearing loss, on the left side. At ENG testing, no spontaneous nystagmus was present but, in the Rose position, a positional nystagmus was noted with the horizontal component beating to the right (Fig. 5). A vestibular left paresis, on caloric testing, was also found. No spontaneous nystagmus was recorded. On the contrary, the neurological ABR showed an inconsistent shape and prolonged latency wave V on the left side. Cerebral MRI and MRA did not show any abnormalities. This year the patient has not complained of vertigo, but rather of a permanent disturbing pulsatile tinnitus. The hearing level has slightly deteriorated, in the left ear.

All patients refused surgical treatment to repair SSS.

Discussion
In SSS, occlusion or marked stenosis are present, usually due to atherosclerosis, of either the subclavian or innominate artery proximal to the origin of the VA. The established difference of pressure between the subclavian and basilar
artery may lead to a siphoning effect with blood flowing from the opposite VA across the basilar artery and down the VA on the occluded side. This results in a steal blood phenomenon at the expense of the basilar artery, as blood flow is reversed to the ipsilateral occlusion VA, which leads to a hypoperfusion state in the vertebrobasilar arterial (VBA) system and thus vertebrobasilar insufficiency (VBI). In our third case, SAS coexisted with SSS, as already reported in another study, without excluding the possibility that SSS was secondary to SAS when lifting the arm. The possible mechanism would be the anterior scalenus muscle compressing the subclavian artery medially to the vertebral column, resulting in its obstruction.

SSS is classified as asymptomatic, oligosymptomatic, if only neurological symptoms or upper limb ischaemia are present and complete when both symptoms are found. When symptomatic, the SSS can be manifest with a variety of VBI symptoms such as headache, blurred vision, diplopia, impairment of consciousness, dysarthria, and facial paraesthesiae. Neurotological symptoms, such as vertigo, are also mentioned in VBI and are considered to be of central origin. Our patients suffering from SSS and thus from VBI presented with unsteadiness (cases 1, 2), recurrent vertigo (case 3), hearing loss (case 3) and tinnitus (cases 2, 3). Although VBI is related to disturbed haemodynamics in the posterior cerebral circulation, neurological symptoms may be isolated, without any other neurological signs of brainstem ischaemia. Baloh and Halmagyi reported that a high incidence of isolated episodic vertigo was found in patients with VBI and it was not clear whether vertigo originated from ischaemia of the labyrinth, brainstem, or both structures. In all three cases, a positional nystagmus was revealed, being vertical in two (up-beating in case 1, down-beating in case 2). No cases of spontaneous nystagmus were observed. The positional nystagmus could be an isolated finding, in VBI, even if there were no other neurological signs. Moreover, positional vertical down-beating nystagmus has already been described in VIB; positional vertical up-beating nystagmus is attributed to brainstem lesions located at the ponto-mesencephalic and ponto-medullary junctions; accordingly, in case 1 ischaemic lesions were located in the low brainstem. Some studies have reported that the most common finding in VBI is the unilateral vestibular paresis to caloric testing. In our study, in only 1 out of 3 cases was the caloric response reduced; it is currently accepted that caloric testing remains normal in central lesions. Abnormal oculomotor testing, such as smooth pursuit, is frequently seen in VBI patients, in our case 2, abnormal saccades were detected, indicating a central nervous system involvement. In 2 out of 3 of our patients, sensorineural hearing loss was demonstrated, unilateral in one and bilateral in the other. Unilateral hearing loss, attributable to VBI is not so frequent, estimated at approximately 20%, according to Yamashoba et al. in a series of 70 patients suffering from VBI; bilateral, sudden sensorineural hearing loss, although rare, is also described in VBI, in isolation or accompanied by neurological symptoms. ABRs were abnormal in all cases, including prolonged wave V and increased wave I-V latency interval on the side of SSS (cases 1, 3) and complete desynchronized traces in case 2. Similar findings have been reported in patients affected by SSS, even in asymptomatic patients and after vertebrobasilar transient ischaemic attacks and they are compatible with retrocochlear lesions affecting both the acoustic nerve and the central auditory pathways.

Since the VBA system feeds both the peripheral and central region of the auditory and vestibular systems, it is difficult, in SSS, to precisely localize the site of insult. The presence of positional vertical nystagmus (cases 1, 2) and prolonged wave V and increased wave I-V latency interval in ABRs, the finding of abnormal saccades (case 2) and the radiological...
Clinical evidence of cerebral ischaemic lesions (cases 1, 2) support the hypothesis that, in SSS, the central auditory and vestibular system is implicated in the pathogenesis of neurotological symptoms. However, as already mentioned, subclavian artery lesions are usually asymptomatic because of the abundant collateral blood supply in the head, neck, and shoulder. It is important to note that when the syndrome becomes symptomatic, then vertigo is the most frequent symptom, of which a neurotologist should be aware. According to a large study on 168 patients suffering from SSS, vertigo was present in 52% of cases and tinnitus in 4%. Thus, in patients suffering from SSS, who developed dizziness, hearing loss or tinnitus, careful neurological examination and follow-up are mandatory in order to prevent other neurological deficits in the VBA territory.

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