Bilateral sudden sensorineural hearing loss and chronic venous cerebrospinal insufficiency : a case report.

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Bilateral sudden sensorineural hearing loss and chronic venous cerebrospinal insufficiency : a case report

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Abstract

Objectives. We report a case of bilateral sudden sensorineural hearing loss in a patient suffering from chronic venous cerebrospinal insufficiency. **Methods.** Audiometric testing confirmed bilateral sensorineural hearing loss with hypoexcitability to caloric stimulation on the left side and echo-colour Doppler examination showed abnormal cerebral venous deficiency. **Results.** The patient's condition improved after fifteen days following medical treatment. **Conclusions.** Chronic venous cerebrospinal insufficiency may explain the anatomical background which provides a predisposing factor for sudden sensorineural hearing loss although further studies are needed to verify whether this observation is casual or coincidental.

Introduction

Sudden sensorineural hearing loss (SSHL) is defined as a sensorineural hearing loss of 30dB or greater over at least three contiguous audiometric frequencies occurring within a 72 hour period.[1] Various etiologies have been proposed to explain SSHL including vascular mechanisms (microembolism, hemorrhage, thrombosis), inflammation, meningitis, syphilis, HIV infection, autoimmune inner ear disease, ototoxic drugs, tumors, traumatic and idiopathic mechanisms. [2] SSHL has also been associated with vertebro basilar insufficency (VBI).[3-10] However, for the majority of patients an etiologic factor is not identified.

Chronic venous cerebrospinal insufficiency (CCSVI) was initially described as being strongly associated with multiple sclerosis (MS).[11] It is a syndrome characterized by stenosis of the internal jugular veins (IJVs) and/or azygous vein (AZ) with opening of collaterals and insufficient drainage. [12-14]

We report a case of bilateral SSHL with vertigo, showing evidence of the CCSVI pattern but not associated with MS. To the best of the authors' knowledge, this kind of association has never been reported.

Case Report

In 2010, a 56-year-old female came to our observation for a sudden sensorineural hearing loss (SSHL) in the right ear associated with aspecific dizziness and lightheadedness. In 2003, without any previous history of hearing difficulties, the patient had experienced an episode of SSHL in the left ear. In both episodes, there was no prior history of temporal bone fracture, meningitis, autoimmune disease or exposure to ototoxic drugs. General and neurological evaluations were normal; extensive blood tests including liver function tests, complete blood count, urea nitrogen, creatinine, erythrocyte sedimentation rate, circulating immune complex, and serum electrolytes all

gave normal results. Antinuclear antibody, rheumatoid factor, C reactive protein, HIV and VDRL tests were unremarkable as were clotting tests. A transthoracic echocardiogram and an electrocardiogram showed no abnormalities.

Audiometric testing, performed on admission, showed sensorineural hearing loss with a pure-tone threshold (PTA) at 500- 1000-2000 Hz at 55 dB with 90% speech discrimination in the left ear and PTA 45 dB with 95% speech discrimination in the right ear. Stapedial reflex thresholds at multiple frequencies were normal on both sides.

Normal waveform responses were evoked bilaterally on brain stem auditory evoked potentials. Electronystagmography showed hypoexcitability (more than 22% asymmetry in our laboratory) to caloric stimulation on the left side. There were no abnormalities in saccades, pursuit, or optokinetic nystagmus. Magnetic resonance imaging of the brain, including diffusion images, was normal for a subject of this age.

The echo-colour Doppler for assessing cerebral venous return, according to the protocol of Zamboni P et al., (15) showed an abnormal cerebral venous deficiency. A type B pattern characterized by significant stenoses of both IJVs and of the proximal azygous was observed. Abnormal reflux of the vertebral veins and bilateral stenosis of the brachiocefalic junction were also detected.

The problem we observed concerning the IJVs appeared to consist mainly of a malfunction of the valve leaflets that usually allow the correct outflow of the venous bloodstream and impede back stream. In both cases the leaflets appeared to have a reduced motility and, de facto, caused a stenosis of the vessel lumen. Secondly, the reflux of the vertebral vein system was quite evident, especially when the examination was performed in the upright position, since there was a poor outflow into the azygos venous system.

The patient was treated with systemic steroid, hyperbaric oxygen and platelet aggregation inhibitor (16) and an improvement of 35 dB of PTA was obtained in the right ear after fifteen days. No modifications were observed in the left ear.

Discussion

In 2007, Shim et al.[17] presented a case of unilateral profound sensorineural hearing loss due to a venous malformation of the internal auditory canal. As expected, no malformations of the internal auditory canals were detected in our patient and therefore we had to focus on the possible effects of CCSVI on the outflow of the inner ear, that is completely drained by the vein of the cochlear aqueduct.[18] In fact, the common modiolar vein enters the bony channel immediately adjacent to the aqueduct to become the vein of the cochlear aqueduct which in turn drains via the inferior petrous sinus into the IJVs. Injury or occlusion of this vessel would be particularly significant since it is widely believed to provide virtually the entire venous drainage of the cochlea. [13, 19] The cochlear aqueduct and the internal auditory canal communicate with the subarachnoidal space; in the guinea pig model, an occlusion of the veins of the cochlear aqueduct results in an increase of perilympatic endolymphatic pressure, a decrease of cochlear blood flow and endolymphatic potential. [20]

Furthermore, since many of the venous vessels in the scala tympani have little or no bony covering and are essentially exposed to the perilymphatic space, the venous system is a route of entry for the cells participating in the inner ear inflammatory process. [21]

Another interesting point is that the blood leaves the brain by using the back propulsion of the residual arterial pressure (vis a tergo), complemented by anterograde respiratory mechanisms (vis a fronte). [1, 2, 4 – 7] The latter consist of the thoracic pump increased venous outflow during inspiration: the increase of negative thoracic pressure improves the aspiration of blood toward the right atrium. In addition to vis a tergo and vis a fronte, postural mechanisms play a fundamental role in ensuring a correct cerebral venous return. Several ultrasound studies of healthy volunteers have demonstrated that the pattern of cerebral venous drainage changes, even under physiological conditions,

depending on the body position. [1, 2, 4 – 7] In the prone or supine position, the outflow through the IJVs is favoured, whereas passing to the upright position transfers most of the encephalic drainage to the vertebral veins. [1, 4 – 7]

On the basis of these considerations, it would be interesting to study the CCSVI in certain cases of bilateral SSHL following general anesthesia for non-otologic surgery involving the use of cardiopolmunary bypass of unknown origin [22] since the CCSVI might explain the anatomical background which provides a predisposing factor for SSHL. Further studies are needed to verify whether this observation is causal or coincidental.

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