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REVIEW ARTICLE

Idiopathic sudden sensorineural hearing loss: etiopathogenic aspects

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Summary

Several factors have been postulated to elicit the etiology of idiopathic sudden sensorineural hearing loss. Through a bibliographic review, we made a critical analysis of the different etiopathogenic aspects of its clinical manifestation. The most recent studies concerning the possible causes of sudden hearing loss suggest vascular disorders, rupture of the inner ear membrane and autoimmune diseases; however, viral infections have received a great deal of attention in recent years. Little is known about the mechanism of sudden hearing loss. Viruses can cause sudden hearing loss in an acute infection, however the latent form, and its possible reactivation have also been considered as explanations of the cochlear injury mechanism. Even though hearing loss can be explained by a blood viscosity change, experimental and clinical studies do not show any evidence of labyrinthine fibrosis and new bone formation, or labyrinthine membrane breaks. These findings are not in agreement with vascular and rupture membrane factors, respectively. The eventual presence of antibodies against the inner ear suggests that sudden hearing loss pathogenesis may be of autoimmune nature, but the difficulty in establishing the correlation of its morphological and clinical aspects to the hearing loss also do not help to support this statement. Sudden hearing loss is still a controversial and obscure subject in several aspects.

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INTRODUCTION

Sudden hearing loss (SHL) means a sensorineural hearing loss of sudden onset or one that happens in minutes, hours or even in a few days. The hearing impairment varies as far as intensity and sound frequency are concerned, and in general, it is unilateral (98-99% of the cases)[1,2].

It may be missed in children, specially when it is unilateral. Tinnitus frequently follow the symptoms of idiopathic sudden hearing loss (ISHL) (70% of the cases) and diziness is sometimes present (up to 40% of the cases), thus completing the triad of Ménière. About 10% of dizziness cases may be incapacitating and associated to nausea and vomit. There may also be ear fullness, headache and viral infection symptoms of the upper airways[1-3].

One of the few emergencies in otology, the ISHL affects mostly those in their fourth decade of life, involving both the right and the left ears in equal proportions, and the same trend is seen genderwise[4].

It affects 5 to 20 persons for each 100 thousand individuals. It is estimated that there are approximately 4 thousand new cases per year in the United States, and 15 thousand all over the world[7].

In Brazil we lack good epidemiological references as to the true incidence of SHL, due to the difficulty in safely assessing its incidence. Poor cultural and socioeconomical conditions, together with the possibility of spontaneous recovery before seeking medical help or the very disregard towards light symptoms are responsible for this epidemiological picture[3].

Although no seasonal influence has been proven for ISHL, it is known that patients with such disorder are mainly affected after an upper airway infection or herpes labial infection[8].

The hearing loss (HL) caused by ISHL may become permanent or regress either totally or partially. Many authors report that spontaneous recovery happens in 45 to 60% of the patients[4,6,9,10]. In a more detailed fashion, it is estimated that about 25% of patients have total spontaneous recovery; 50%, partial and 25% present no recovery at all[2].

Since ISHL physiopathology is still unclear, there are disagreements as to its true cause[11,12]. Such fact becomes even more evident when it happens as an isolate clinical situation, that is, symptoms restricted to the inner ear only. Thus, there are many theories as to the origin of this disease, including viral infections, blood disorders, immune disorders and perilymphatic fistulas.

Having stated that, our goal is to assess ISHL as to its main and possible etiology and physiopathology.

BACKGROUND

Prosper Ménière was probably the first author to clearly describe a case of SHL without a definitive cause[4,5,6,13], in 1861; however, it was DeKlein, in 1944, the first to report a clinical study with 21 patients with SHL, listing causes of hearing loss among inner ear hemorrhage, acute and chronic inflammations, traumatic fractures, multiple sclerosis, brain tumors, ototoxic drugs, emboli, hypercoagulability, radiation, pregnancy and herpes zoster as potential predisposing causes.

Later, Rasmussen in 1949, apud Fowler (1950) and Hallberg (1956), report cases of ISHL and discuss the disease origin, proposing the vascular origin and the neuritis of the VIII nerve as possible causes. Following that, Lindsay & Zuidema (1950) also studied patients with SHL and discuss possible causes. According to these authors, vascular diseases such as thrombosis could justify the hearing loss, however the age range did not match; hemorrhage would be another possibility, provided the patients had previous history of coagulation problems or head injury. They then consider, although hypothetic, the possibility of vasospasms, besides infection as the possible cause for hearing loss (HL), what could then justify this variable pattern of auditory and vestibular involvement.

Back in 1950, Fowler present some SHL cases and stresses that most of the patients had a psychosomatic disorder associated. He reports not only the importance of the supratentorial disorder, but also the change in blood viscosity, as possible causes for SHL. It was also in 1950 that Opheim, apud Hallberg (1956), reported cases of SHL, highlighting the possibility of an acute increase in labyrinth pressure as being responsible for such happening.

Later, in 1952, Moulonguet and Bouche, apud Hallberg (1956), reported the case of a young physician with SHL after having used an intravenous medication, and also considered that the arteriolar spasm caused by this drug caused the clinical signs and symptoms in this patient.

Disorders of the labyrinthine circulation such as the occlusion of microvessels in the inner ear and cochlear hemorrhage have been considered as possible causes of ISHL, specially because they are of sudden onset, notwithstanding, there are a number of non-compatible aspects, such as age range, risk group and prognosis[2,9,14].

It may be that an alteration in blood viscosity caused by a prior disease be consider a predisposing factor, however, not as a primary cause of sudden hearing loss[13].

In 1957, Van Dishoeck & Bierman[15] influenced by previous studies and seeing that flue-like symptoms almost always precede ISHL, are among the first to report a viral infection as a possible cause for such disorder, specially when they compared cochlear lesions found in temporal bones of individuals with ISHL with those with viral labyrinthitis.

Heller & Lindenberg16 in 1955, and Lindsay & Zuidema (1950) had also published a study in which they identified some patients with hearing loss possibly