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VERTEBROBASILAR DOLICHOECTASIA AS A CAUSE OF SUDDEN DEAFNESS: A CASE REPORT

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ABSTRACT The sudden and unexplained hearing loss may occur at any age group and is characterized, according to the US National Institute for Deafness and Communication Disorders (NIDCD), in all idiopathic hearing loss of at least 30 dB in at least 3 connected frequencies, occurring within a period of 72 hours. The etiology often is uncertain probably viral, autoimmune or microvascular. We present a case report about a patient who developed sudden hearing loss due to a vertebrobasilar dolichoectasia, a rare cause of microvascular etiology.

KEYWORDS : sudden, sensorineural, hearing loss, dolichoectasia

INTRODUCTION

The emergence of sudden and unexplained hearing loss is a major cause of ENT emergency. The incidence ranges from 2 to 20 cases per 100,000 individuals annually [1]. It may occur at any age group and generally affects patients ranging from 43 to 53 years [2]. It is commonly unilateral and must occur within 72h.

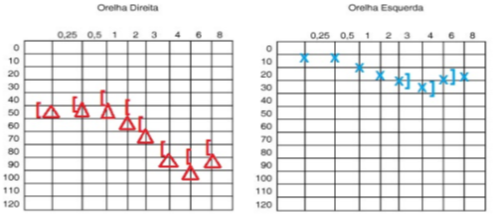
The etiology of sudden deafness in the majority of patients is uncertain, probably viral, autoimmune or microvascular. However, other causes of unilateral hearing loss, such as acoustic neuroma, perilymphatic fistula, Meniere's disease, vascular insufficiency, multiple sclerosis, or other conditions involving the central nervous system can only be detected by magnetic resonance imaging.

Among the causes of central involvement, those related to vascular alterations are highlighted. Vertebrobasilar dolichoectasia is a rare cause of sudden deafness and asymmetric sensorineural hearing loss [3]. This symptom occurs due to compression of the 8th cranial nerve, resulting from vascular ectasia [4].

CASE REPORT

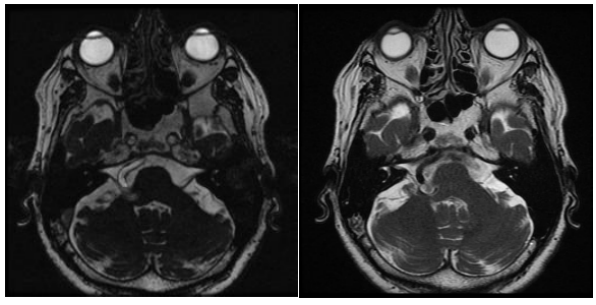
VBR, male, aged 68 years, with a history of sudden onset of hypoacusis and tinnitus in the right ear at 24 years of age. Tinnitus was acute and constant with sounds similar to a cicada,. The patient denied any triggering event that preceded hearing loss or any progressive worsening of the condition. There were no factors of improvement or worsening. He has been using a hearing aid for 2 years without improvement of hypoacusis or tinnitus. He denies any symptoms in the left ear. Personal history: DVT of the lower limb, neurocysticercosis, former smoker. ENT examination with otoscopy, anterior rhinoscopy and oroscopy was normal. Cranial nerves showed no alterations.

A calibrated diagnostic audiometer was useed for the diagnostic. A pure tone audiometry was normal in the left ear while there was a moderate sensorineural hearing loss on the right ear (Image 1).



(Image 1)

Magnetic resonance imaging was ordered. It showed preserved cranial nerve complex VI-VIII on the left and deviated posteriorly on the right, due to a tortuous vertebrobasilar artery, which determined nerve compression in the topography of the right cerebellopontine angle cisterna (Images 2 and 3).



(Image 2) **(Image 3)**

Based on the findings, the diagnosis of sudden deafness secondary to 8th cranial nerve compression due to dolichoectasia of the basilar artery. Evaluation by the neurosurgical team was requested. Clinical treatment with symptomatic medication for tinnitus was recommended. The patient's hearing aid was maintained, although it did not improve hypoacusis or tinnitus. Due to the high risk of morbidity and mortality, surgical treatment was contraindicated.

DISCUSSION

Sudden deafness is defined as sudden hearing loss. Audiometry reveals sensorineural hypoacusis without an identifiable factor during history-taking or physical examination. According to the US National Institute for Deafness and Communication Disorders (NIDCD), sudden deafness is considered all idiopathic hearing loss of at least 30 dB in at least 3 connected frequencies, occurring within a period of 72 hours [5]. However, this definition has not been completely accepted by all authors [6].

Patients typically report sudden or rapidly progressive hearing loss, or notice hearing loss when waking up. In a study conducted with 56 patients, 28 had sudden hearing loss, 27 noticed hypoacusis on awakening, and 1 had rapidly progressive hearing loss [11]. In more than 90% of the patients, hearing loss is unilateral. Bilateral loss is rare (3%). The patient commonly reports a sensation of fullness in the ear

hours or days before hearing loss occurs. Tinnitus appears in over 90% of the patients. Vertigo is reported in 20-60% of the patients [12].

Physical examination was normal, without any signs of otitis or obstruction of the external ear. Tuning fork tests are indicated to evaluate if hearing loss is conductive or sensorineural. If during Weber's test, the tone does not lateralize or lateralizes to the opposite side of hearing loss, the patient has sensorineural hearing loss and must be urgently evaluated.

Patients should be questioned about history of trauma, otalgia or ear discharge, fever, focal neurologic symptoms, headache, diplopia, recent eye pain or redness and previous history of hearing loss.

A neurologic examination is necessary, to exclude cerebrovascular accident in the anterior inferior cerebelar artery territory, as the cause of sudden deafness.

Regarding etiology, the majority of cases is unclear [7]. The most common triggers include: viral neuritis, microvascular alteration [8] and autoimune disorders [9,10]. Type 1 Herpes virus (HSV-1) is considered a significant etiologic fator. There are a variety of other causes, ranging from snake venom to oral contraceptive use: (1) Infectious causes: meningitis, mumps, German measles, syphilis, herpes virus, HIV, mononucleosis, mycoplasm, toxoplasmosis and cytomegalovirus; (2) Toxic: snake venom and ototoxic drugs; (3) Immune: Wegener's granulomatosis, Cogan's Syndrome, polyarteritis nodosa, lupus; (4) Other causes: Meniere's disease, internal cranial hyperostosis, pseudoacusis; (5) Malignancy: Acoustic neuroma, meningioma, lymphoma, meningeal carcinomatosis; (6) Neurologic: multiple sclerosis, neurosarcoidosis; (7) Circulatory: cerebrovascular accident, labyrinthine hemorrhage, vertebrobasilar insufficiency or compression and (8) Trauma: temporal bone fracture, acoustic trauma, barotrauma, perilymphatic fistula and history of previous ear surgery. [6]

Vertebrobasilar dolichoectasia is defined as dilatation of the vertebrobasilar artery, in which the intralumimal diameter is >4.5 mm and the basilar artery lies lateral to the margin of the clivus, dorsum sellae or bifurcation above the suprasellar cystern [15]. Its incidence ranges from 0.06 to 5.8%. Due to its topography, vertebrobasilar dolichoectasia may lead to compression of the cranial nerves, most commonly the 5th and 7th nerves and less commonly the 4th, 6th and 3rd nerves. In the literature, there is no description of compression of the 8th cranial nerve. Furthermore, a database review did not find 8th cranial nerve compression due to vertebrobasilar dolichoectasia that resulted in sudden deafness.

Diagnosis of the patient relied on magnetic resonance imaging. Clinical treatment was chosen, maintaining use of the patient's hearing aid. Due to the location of the dolichoectasia, surgery could have resulted in a cerebrovascular accident.

Therefore, all patients with sudden deafness should undergo audiometric testing. The majority of patients must undergo gadolinium-enhanced magnetic resonance imaging, unless the etiology is absolutely clear [13. 14]. During MRI, we can identify other causes of unilateral hearing loss, such as acoustic neuroma, perilymphatic fistula, Meniere's disease, vascular insufficiency, multiple sclerosis, or other conditions with central nervous system involvement. In some rare cases such as ours, the cause of sudden deafness and aymmetric hearing loss is dolichoectasia of the basilar artery.

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