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Contralateral progressive hearing loss in a patient with unilateral vestibular schwannoma

Perda auditiva progressiva contralateral em paciente com schwannoma vestibular unilateral

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PALAVRAS-CHAVE: Schwannoma vestibular. Perda auditiva. Otorrinolaringologia.

INTRODUCTION

Vestibular schwannoma, also called acoustic neuroma, is a Schwann cell tumor that typically originates from the nerve sheath of a vestibular branch of the 8th cranial nerve, with most tumors being sporadic and unilateral. Bilateral alterations correspond to less than 5% of cases and are usually related to neurofibromatosis type 2.[1,2]

As these tumors grow, they extend to the cerebellopontine angle, where they account for 80% of the tumors there. The incidence varies from 1:10,000 for unilateral cases to 1:30,000 for bilateral cases.[1]

Vestibular schwannoma typically presents with ipsilateral sensorineural hearing loss and tinnitus in 60% of patients, and more rarely may present with hydrocephalus, paresthesia, headache, and vertigo. Although it is a benign tumor with insidious growth, it is a condition with a risk of injury to various intracranial structures due to its mass effect.[2]

Treatment options include conservative treatment, surgical removal of the tumor, and radiation therapy. Despite surgical removal, hearing improvement is uncommon, even if anatomical preservation of the 8th. cranial nerve is reached and the resection leads to nerve decompression.

Pathophysiological mechanisms of vestibular schwannoma in course of sensorineural hearing loss are still incompletely understood. There is a lack of correlation between audiometric tests and the size and extent of the tumor to the inner auditory canal, as well as changes in auditory thresholds seen in cases without tumor growth detected in imaging studies. These clinical observations may suggest additional explanations for the pathophysiology beyond just the mechanical compression of adjacent structures.[1]

Recent studies have revealed new insights into the ability of factors secreted by sporadic tumor, such as tumor necrosis factor-alpha, and human extracellular vesicles that can cause cochlear damage. Secretions produced by tumors may have the potential to reach the contralateral ear by traveling through cerebrospinal fluid (CSF) or blood. To date, there are few studies that have investigated possible effects of vestibular schwannoma on hearing in the contralateral ear.[1]

Hearing loss in the contralateral ear is a very rare complication that has been reported in several patients in the postoperative period. Several theories have been proposed, including perforation noise, vascular compromise, meningitis, ototoxicity, intratympanic pressure, autoimmunity, allergy, general anesthesia, and hemodialysis. In addition, CSF loss during surgery is transmitted to the perilymph through the cochlear aqueduct, causing a decrease in its pressure and, consequently, compensatory expansion of the endolymph, mimicking endolymphatic hydrops. Thus, the mechanism behind this phenomenon remains unclear, and the optimal treatment is also uncertain.[3]

CASE REPORT

A 56-year-old man, previously healthy, presented with a history of hypocusis and unilateral tinnitus on the right with insidious and progressive evolution since 2014. At the time, the patient had audiometry showing anacusis on the right and moderate to severe sensorineural hearing loss on the left. Investigation was carried (Figures 1 to 4) out and a diagnosis of vestibular schwannoma on the right was made with a neurosurgery team, with indication of surgical approach via occipital craniotomy. The tumor was partially resected in 2016 and was followed up by the assistant team. After the operation, he noticed progressive hearing loss also on the left and used an individual sound amplification device on the left with

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a good response for about 1 year, but with regression of improvement after that, he chose to cease its use. She performed a test with a speech therapy team with more powerful individual sound amplification, but without response. He also complained of continuous tinnitus without laterality, which began soon after the operation. The patient is being followed up with annual magnetic resonance imaging with no evidence of tumor growth, with a residual nodular lesion with intermediate signal intensity on T1- and T2-weighted sequences, hyposignal on all sequences, and restriction on diffusion and peripheral contrast uptake at the right cerebellopontine angle of 14x12x13 mm (lower than in the previous study of 18 mm in its longest axis). Audiometry from 2020 already showed bilateral anacusis.

Patient denies other head and neck operations, family history of hearing loss, previous treatments with radiotherapy, chemotherapy, long-standing use of antibiotics or atypical infections. He had no alterations on otoscopy.

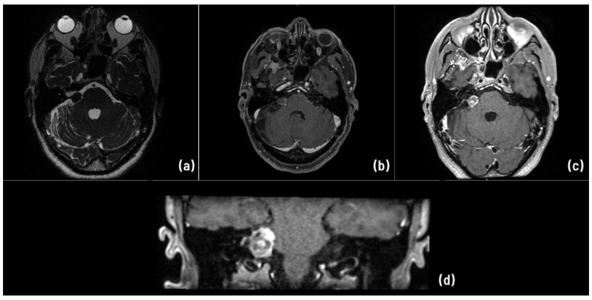

FIGURE 1 — Preoperative computed tomography: Images (a) and (b) of axial T2-weighted sequence, (c) of axial T1-weighted sequence, and coronal T1-weighted image (d) showing an intermediate-signal residual nodular lesion on T1- and T2-weighted sequences, hyposignal on all sequences, and restriction on diffusion and peripheral contrast uptake at the right cerebellopontine angle.

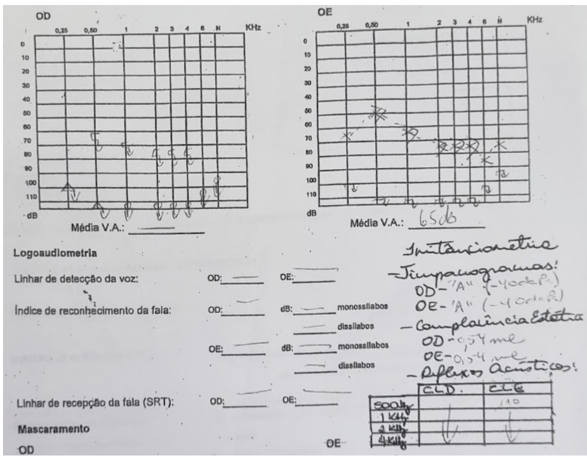

FIGURE 2 — Audiometry from 2014 showing anacusis in the right ear and moderate to severe sensorineural hearing loss in the left ear. Tympanometry with bilateral "A" curve and bilateral absent reflexes.

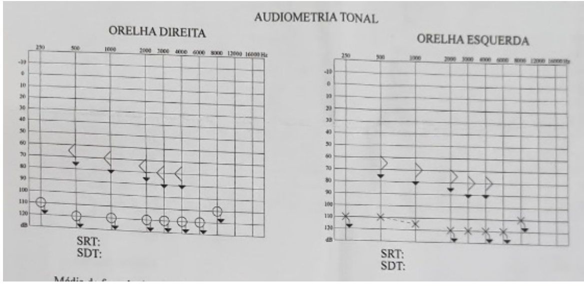

FIGURE 3 — 2020 pure tone audiometry showing ankacusis on the right and progression of sensorineural hearing loss, profound on the left

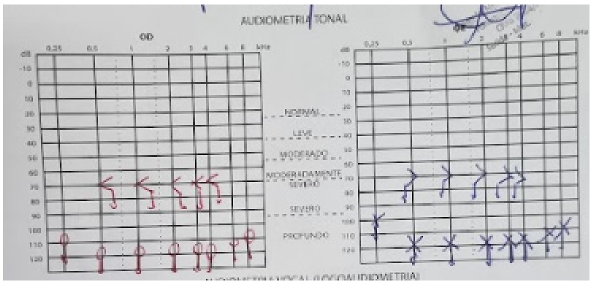

FIGURE 4 — 2022 pure tone audiometry showing bilateral hearing loss

DISCUSSION

To date, no drugs have been approved to treat vestibular schwannoma, limiting treatment to expectant management, surgery, and/or radiation therapy. Both surgery and radiation therapy are associated with significant risks, including hearing loss and facial nerve palsy, with up to 28% of patients experiencing at least one of these complications.[1]

Hearing preservation after vestibular schwannoma operation is, in itself, a challenge, and complications such as the development of contralateral deafness are extremely rare. By 2016, 14 cases of contralateral hearing loss had been reported after surgery.[4]

The mechanism responsible for the induction of sensorineural deafness by vestibular schwannoma is not fully elucidated. The main hypothesis is mechanical compression to the adjacent auditory nerve, but recent studies have not correlated an increase in tumor dimensions with alterations by audiometric criteria. Several theories have been proposed, including meningitis, ototoxicity, perforation noise, vascular compromise, autoimmunity, general anesthesia, and compensatory endolymphatic dropsy caused by CSF loss.[1,5] The patient in question did not present signs of infection, alterations on otoscopy, nor did he use ototoxic drugs.

Among the plausible causes of contralateral sensorineural deafness is also autoimmune injury. Even surgical manipulation can release sequestered antigens into the systemic circulation, leading to the autoimmune reaction that can harm the contralateral ear.[5] The patient presented progressive hypoacusis on the left with onset soon after the operation, as well as tinnitus immediately after the postoperative period. This places autoimmunity as an unlikely cause, due to the short window of time.

The most widely accepted theory for contralateral hearing loss is compensatory endolymphatic hydrops caused by CSF loss. Under normal conditions, the pressure of the cerebrospinal fluid, perilymph and endolymph are equal. The loss of this fluid results in decreased pressure, which is transmitted to the perilymph by the cochlear aqueduct. As consequence, there is expansion of the endolymphatic component and endolymphatic hydrops.[5,6]

Walsted et al.[7] conducted a prospective study involving 60 patients undergoing surgical treatment for vestibular schwannoma resection and reported the presence of contralateral sensorineural hearing loss in 40 of them. The changes were greater in the low frequencies after the operation, however, after 1 week, the treble was also impaired. A similar phenomenon of decreased hearing attributed to CSF loss has been described following spinal anesthesia, spontaneous intracranial hypotension, and CSF shunt operation.[6]

Yet, in another study by Walsted et al.[8] On the same subject, an association was found between the number of affected patients, the mean threshold, the number of frequencies affected, and the amount of CSF lost. In the literature, hearing loss attributed to this cause tends to normalize after 3 months, although there are cases of permanent deafness associated with CSF loss.[6]

Because the etiology is uncertain, there are no guidelines for treatment. Steroid therapy has been used in some patients5, but there is no scientific evidence to support the adoption of this treatment, and the results are often insignificant.[3] Even so, there is a recent report in which the administration of high doses of steroids associated with hyperbaric oxygen therapy was recommended.[9] Other potential options for treatment are Bevacizumab, Everolimus, and Lapatinib.[2]

Authors' contributions

Conceptualization: Debora Emi Shibukawa
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