Simultaneous cochlear implantation as a therapeutic option in vestibular schwannoma surgery: case report

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Hearing loss is the most common symptom of vestibular schwannomas (VSs). The management of these lesions includes observation, radiosurgery, and microsurgical resection. Hearing preservation and rehabilitation are the major challenges after the tumor treatment. A 43-year-old man with previous left-sided profound hearing loss and tinnitus presented with a 2-mm left-sided intracanalicular VS. The decision was made to perform a simultaneous cochlear implantation (CI) and microsurgical resection of the tumor. The patient did well postoperatively, with significant improvement of tinnitus, sound localization, and speech recognition in noise. Previous reports of simultaneous CI and VS resection in patients with neurofibromatosis type 2 and sporadic VS in the only hearing ear have been described. The role of CI in patients with VS and normal contralateral hearing has been recently described, showing positive outcomes due to the binaural benefits. Tinnitus also can be treated by the implantation of the cochlear device. The simultaneous microsurgical removal of VS and implantation of a cochlear device is a feasible approach in patients with unilateral hearing loss and severe tinnitus.

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VESTIBULAR schwannomas (VSs) are benign and slow-growing tumors that correspond to approximately 6% of all intracranial neoplasms. These lesions may lead to a varied clinical profile, including hearing loss, tinnitus, imbalance, vertigo, facial paralysis, sensory changes associated with the trigeminal nerve, and, not infrequently, hydrocephalus and compression of the brainstem, especially in large lesions.^{4,16}

Hearing loss is the leading presenting symptom in the clear majority of patients and is frequently associated with tinnitus resulting from cochlear nerve compression and cochlear vascularization disorders. Current management includes observation, a variety of radiation modalities, and microsurgical resection. Observation is preferred in asymptomatic patients with small lesions, in elderly patients, and in patients at a high surgical risk, thus requiring periodic imaging evaluation and audiometric tests.²⁶ Radiation has expanded the management options for VS, demonstrating good outcomes, and typically is reserved for tumors smaller than 3 cm.^{10,15,20} Microsurgical treat-

ment is suitable for any type of VS, and is mandatory for life-threatening tumors (e.g., acute hydrocephalus, brainstem compression). It is also recommended in cases of failed previous radiosurgery.⁷ When hearing preservation is not possible, hearing rehabilitation is an important step to afford quality of life to the patient. Cochlear implantation (CI) and auditory brainstem implants can be used.²² In this paper, we report a case of a small intracanalicular VS treated simultaneously with microsurgical resection and CI. A review of the literature on this topic and medical options will be discussed.

Case Report

History

This 43-year-old man presented with a 30-year-history of unilateral left-sided deafness attributed to mumps infection and tinnitus. He was treated by the otolaryngology team for tinnitus, and an attempt was made to recover his hearing. The patient did not adapt to a bone-anchored

ABBREVIATIONS CI = cochlear implantation; VS = vestibular schwannoma.

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hearing aid. In a routine MRI study we found a 2-mm intracanalicular VS (Fig. 1). Management options including observation with serial imaging, radiation, and resection were discussed. A cochlear implant was suggested, and a simultaneous tumor resection could be performed.

Examination

Audiometric testing demonstrated good hearing in the right ear. However, the left ear presented profound sensorineural hearing loss.

Operation

We routinely perform a conventional retrosigmoid approach for most VSs; however, because the patient already presented with complete hearing loss and the surgical goal was also the placement of a cochlear implant, we adopted a transmastoid translabyrinthine approach. Figure 2 illustrates the procedure stepwise. The surgical procedure was performed under electroneurophysiological monitoring. The cochlear implant was manufactured in Sydney, Australia (Freedom Implant CI24RE—Contour Advance; Cochlear Corp.).

Impedance and neuronal response telemetry was performed after the cochlear implant placement, indicating correct position of the cochlear implant and integrity of the cochlear nerve (Software Custom Sound EP—4.4; Cochlear Corp.).

Postoperative Course

The patient did well, with fitting of the sound processor 40 days after surgery with normal findings on impedance and neuronal response telemetry. The mapping strategy was to obtain progressive gain, which facilitates the patient's adaptation. The CI mapping was conducted 30 days after activation and showed normal values for impedance and neuronal response measurements. The patient had a significant improvement in tinnitus, sound localization, and sound discrimination.

The speech recognition tests before implantation were all negative (0%). After implantation, the accuracy average in the Ling Sound Test for detection and discrimination increased to 54.2%. The identification of vocabulary extension was 50%. The patient will continue the hearing rehabilitation with speech therapy every 4 months until he achieves total adaptation. He also will receive vestibular rehabilitation, which is necessary after the translabyrinthine approach.

Discussion

The Role of CI

In the 1960s, several attempts to stimulate the cochlear nerve to treat deafness were performed.²³ Years later, a multichannel implant formed by electrodes stimulating the cochlea allowed the perception of different frequencies. The present cochlear implant is an evolution of this device.¹⁷ Composed of a microphone, signal processor, transmission system, and multiple electrodes, the sound is captured, processed, and transmitted, causing electrical stimulation at the cochlear nerve.^{5,14}

Simultaneous CI in the management of VS was first

FIG. 1. Axial T1-weighted postcontrast MRI study depicting a left-sided intracanalicular tumor (arrow).

described in 1995 by Arriaga and Marks.² Since then, they have been considered in a few cases in which neurofibromatosis type 2 and sporadic VS were found in the only hearing ear. Under these conditions, the risk of complete hearing loss, either due to the treatment or from the natural course of the disease, requires more effort to provide a satisfactory result.¹³ Previous studies show a positive outcome in patients with sporadic VS in the only or better hearing ear, with similar prognosis to that in patients without VS who were treated with CI.^{1,6,12,13,27} Cochlear implants after radiosurgery in patients with neurofibromatosis type 2-associated VS have been described, with significant restoration of hearing.¹⁰

However, the lack of knowledge about the benefits of CI in patients with normal contralateral hearing still exists. The first study showing the results of a series of patients with this condition who underwent simultaneous CI and tumor resection was published in 2016, with 13 patients. Sanna et al. evaluated the cochlear implant in monaural conditions, but also assessed the binaural benefit perceived by the patient.²¹ Binaural benefits are an important aspect to the patient's quality of life that should be considered in addition to the auditory gain.

The audiological impact of sporadic VS in patients with normal contralateral hearing is mainly the loss of binaural hearing. This comprises a diminished summation effect (identical signal arriving at both ears), a reduction of the squelch effect (ability of the brain to separate noise and

FIG. 2. Intraoperative photographs. The mastoid process is carefully drilled out and the semicircular canals are opened (A); the posterior wall of the auditory canal is exposed (B) and opened (C), and the tumor is identified. Following tumor resection (D), the cochlea is exposed (E) and the electrode is placed (F).

speech coming from different locations), and the head shadow effect (speech discrimination when the head is between the source of the sound and the hearing ear). CI presented as a good option to recover these deficits, providing benefits in conditions like squelch effect and sound localization.^{11,25}

Hassepass et al. demonstrated in their 11-patient series a similar outcome regarding benefits associated with binaural hearing, and they suggested another advantage of CI after translabyrinthine VS surgery: tinnitus suppression.^{8,18} However, the implantation was performed 1 year after tumor resection. The late implantation may be controversial due to the induced postsurgical ossification of the cochlea in the translabyrinthine approach, recommending an intracochlear placeholder to allow later cochlear implant placement. Some studies observed total cochlear ossification, with subtotal degeneration of spiral ganglion cells and cochlear nerve.^{3,9} The main advantage of the simultaneous procedure is to avoid these anatomical obstacles.

Hearing preservation is the major challenge of surgical treatment because complete hearing loss and hypoacusis affect approximately 90% of patients.¹⁹ This fact justifies the concern about hearing preservation and rehabilitation. The remarkable success of cochlear implants had been a

game changer in the rehabilitation of these patients and emerges as a therapeutic option in cases in which there is a low chance of hearing preservation, even with normal hearing in the contralateral ear.²⁴ So, the preservation of the cochlear nerve can allow hearing restoration. However, we need more studies demonstrating the results of this approach.

Conclusions

The simultaneous microsurgical removal of VS and CI is a feasible approach to patients with unilateral hearing loss and severe tinnitus. In this situation, the translabyrinthine approach is preferred instead of the retrosigmoid approach because it is easier to implant the cochlear device. In patients with serviceable preoperative hearing we advocate the retrosigmoid approach, with the intention of preserving hearing. When it is not possible to achieve hearing preservation, a second-stage procedure can be done to implant the cochlear device.

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Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Borba, dos Santos Neto. Acquisition of data: Borba, dos Santos Neto, Zamponi, Hamerschmidt, Wiemes. Analysis and interpretation of data: Borba, dos Santos Neto, Zamponi, Hamerschmidt, Wiemes. Drafting the article: dos Santos Neto. Critically revising the article: Rassi. Reviewed submitted version of manuscript: Rassi. Approved the final version of the manuscript on behalf of all authors: Borba. Study supervision: Borba.

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