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Clinical Note

Cochlear implantation in irradiated tympanojugular paraganglioma

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Abstract

Objective: Indications for cochlear implantation have extended progressively over the years both from an audiological and clinical point of view, including the introduction of cochlear implants (CI) for hearing rehabilitation following skull base surgeries and neurotological procedures. In the past, cochlear implantation has been used for hearing rehabilitation in radiation-induced sensorineural hearing loss in nasopharyngeal and tonsillar carcinoma with successful outcomes. Here we describe a similar outcome following total deafness after tympanojugular paraganglioma irradiation, which is also the first such report in literature. *Design:* The irradiated ear of this patient was implanted with a CI through standard posterior tympanotomy. *Study sample:* A 26 year old male with multiple paragangliomas with bilateral deafness, secondary to surgery on one ear and radiotherapy for the opposite ear. *Results:* After a follow-up of 48 months, no local complications have occurred and the hearing results have remained stable with 100% sentence recognition. *Conclusions:* Bilateral and multiple paragangliomas are rare tumors. Despite the modality of treatment, the hearing is almost always compromised. In these patients, cochlear implantation offers a new perspective for hearing restoration. This report demonstrates that cochlear implantation can be effectively performed after tympanojugular paraganglioma irradiation with long-lasting, satisfactory results, even in the presence of residual tumor.

Key Words: Cochlear implant; radiotherapy; tympanojugular; paraganglioma

Tympanojugular paragangliomas (TJP) are histologically benign and slow growing tumors of the temporal bone. Multicentric paragangliomas are relatively common, found in 10%–20% of sporadic cases and up to 80% of familiar cases (Sanna et al, 2013). Total surgical extirpation is the mainstay of treatment for TJP (Jackson et al, 1996). However, radiotherapy (RT) plays a role in the decision-making process in some cases such as bilateral and multiple paragangliomas, in which avoiding disabling sequelae (i.e. bilateral lower cranial nerves (LCNs) palsy) is of utmost importance (Sanna et al, 2011).

Regardless of the modality of treatment chosen, each treatment has the potential to precipitate sensorineural hearing loss (SNHL). Thus, hypoacusia is an expected sequela of TJP, either as the natural course of the disease, as a consequence of surgical treatment, or as secondary damage to the cochlea and retro-cochlear auditory pathways by RT (Low et al, 2005).

Cochlear implants (CIs) are an established method for hearing rehabilitation in patients with profound hearing loss. Indications for cochlear implantation have extended progressively over the years both from an audiological and clinical point of view. Anatomical anomalies and malformations, cochlear ossification and chronic otitis media no longer pose a problem for CI placement. Moreover, combination with the subtotal petrosectomy procedure allows implantation in special situations (Free et al, 2013). In the last few years, CIs have also been introduced with promising results to restore hearing in combination with skull base surgeries and neurotological procedures (Vicenti et al, 2008).

Along with these increasing prevalent applications, CIs have been progressively introduced for hearing rehabilitation of head and neck cancer radiation-induced hearing loss. Cochlear implantation has been previously described in post-irradiation SNHL for nasopharyngeal carcinoma and tonsillar carcinoma with successful outcomes (Chua &Tan, 2007; Adunka & Buchman, 2007). However, it has not been previously described after TJP irradiation.

Case report

A 26-year-old male with a prior diagnosis of paraganglioma was referred to our center. He had undergone tympanoplasty elsewhere

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Abbreviations

CI	Cochlear implant
CN	Cranial nerve
HRCT	High-resolution computer tomography
LCN	Lower cranial nerve
MRI	Magnetic resonance imaging
RT	Radiotherapy
SNHL	Sensorineural hearing loss
T	Tesla
TJP	Tympanojugular paraganglioma

for presumed chronic otitis media with cholesteatoma. On admission, he gave a five-year history of right-sided pulsatile tinnitus and progressive hypoacusia. Otoscopic examination revealed an open cavity with chronic inflammatory haemorrhagic tissue. Neurological examination demonstrated no cranial nerve (CN) deficits. Pure-tone audiometry showed a severe, mixed hearing loss in the right ear and normal left-sided hearing. The imaging work-up with high-resolution computed tomography (HRCT), gadolinium-enhanced magnetic resonance imaging (MRI), and selective arteriography of the right carotid artery established the diagnosis of TJP class C3 (Fisch classification) (Figure 1). On the basis of extensive internal carotid involvement, a protective endocarotid stent was inserted preoperatively. Three months later, after superselective embolization, the tumor was resected by means of a type A infratemporal approach. As a consequence of this surgery, the patient suffered ipsilateral anacusia and VII, IX, and X CN palsy. At one-year follow-up, VII CN function was grade IV, and fiberoptic laryngoscope examination showed right vocal cord palsy with complete compensation from the left vocal cord.

Three years later, the patient reported a progressive hearing loss in the left side. Though follow up imaging studies showed no evidence of disease on the right side, there was however, a new lesion compatible with TJP class C2 on the left side (Figure 2). A carotid body tumor of 20×24 mm was also detected on the left carotid bifurcation. Pure-tone audiometry showed a right-sided anacusia and a moderate SNHL in the left ear.

In order to avoid bilateral LCN deficits, a decision was taken to irradiate the second TJP. But, after the full course of RT, the patient lost his hearing on the left side, becoming bilaterally deaf.

With the aim of restoring hearing, the possibility of placing a CI was considered one year later. Neuropsychological examination did not reveal disorders concerning intelligence and cognitive abilities. Moreover, the patient was realistic in his expectations and showed strong motivation and a normal psychosocial and family-related situation. HRCT scan showed no signs of recurrence on the right side and fibrotic tumor on the hypotympanum with integrity of the cochlea on the left side. Auditory brainstem responses could not be detected at the highest levels of stimulation on either side. In order to test cochlear nerve conduction after RT, an electrical promontory stimulation test was conducted on the left ear. The dynamic range at 50 Hz was 4.5 μA, with a threshold (T) level of 7.8 μA, and Comfort (C) level of 12.3 μA. At 100 Hz, the dynamic range was 11.6 μA, T level was 11.3 μA, and C level 22.9 μA. Finally, at 200 Hz, the dynamic range was 11.9 μA, and the T and C levels were 24.6 μA and 36.5 μA, respectively. Since the selection program did not reveal any contraindication and the results of the electrical promontory stimulation showed that the cochlear nerve conduction was optimal, we decided to implant the patient. Subsequently, the left ear was implanted with a Digisonic SP device (Neurelec MXM Corporation, Antibes, France) through standard posterior tympanotomy. Full electrode insertion into the scala tympani was achieved through the round window. Intraoperatively, it was necessary to coagulate the tumor with bipolar forceps over the round window to obtain access. Intraoperative electric auditory brainstem responses, stapedial reflex, and telemetry impedance confirmed proper implant function and effective stimulus delivery. The postoperative course underwent uneventful. On the second post-

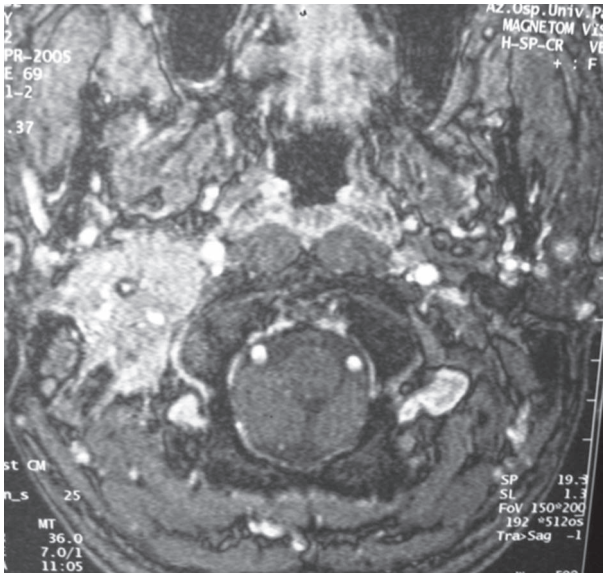

Figure 1. MRI examination of the first tumor. Cervical MRI (axial gadolinium-enhanced T1-weighted). A lesion with heterogeneous enhancement after gadolinium administration with the classic 'salt and pepper' pattern is seen. Note the latero-medial displacement of the cervical segment of the internal carotid artery.

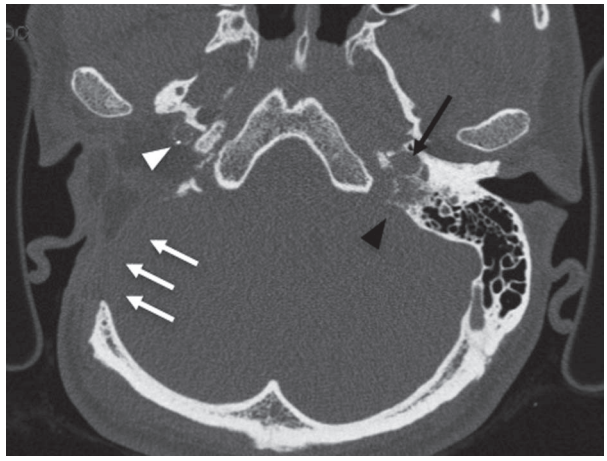

Figure 2. HRCT examination of the second tumor. HRCT, axial cut at the level of the jugular foramen. A characteristic irregular bony erosion (moth-eaten bone) of the jugular foramen and jugular spine are seen on the left side (black arrowhead) with vertical portion of the left internal carotid artery involvement (black arrow). Note the extensive craniotomy on the right side as a result of the previous infratemporal type A approach (white arrows). An endocarotid stent (white arrowhead) is seen in the right internal carotid artery.

operative day, a check radiogram confirmed the correct positioning of the intracochlear electrodes.

The first postoperative control was performed four weeks later. The wound healed well without any flap infection or necrosis. At this same control, the 20 electrodes were activated. He was fitted using the main peak interleaved sampling (MPIS) strategy programmed with eight channels of maxima and a stimulation frequency of 500 Hz. Nonauditory, facial, or pain sensations were absent.

Postoperative auditory performance was evaluated in the auditory-only condition in both closed-set (vowel identification) and open-set formats (bisyllabic word recognition, sentence recognition, common phrases comprehension) with monitored live voice through the sound field at a level of 70 dB sound pressure level. An Italian version of the Northwestern University phonetically balanced word list and the Central Institute for the Deaf everyday sentence list was used. The scores for the hearing test at activation, 6, 12, and 48 months after cochlear implantation are shown in Figure 3. The patient uses the telephone in his daily life.

To date, after a follow-up of 98 months since the surgery for the right-sided tumor, 63 months since the RT treatment for the left-sided tumor and 48 months since the CI, there have been no local complications and hearing results have remained stable. The patient is on annual radiological follow-up with HRCT and remains free from tumor regrowth on both sides. Last control with magnetic resonance imaging (MRI) of 1.5 Tesla (T) demonstrated stability of residual tumor. Despite the CI and the magnet being in position, the area of distortion did not hamper visualization of the jugular foramen (Figure 4).

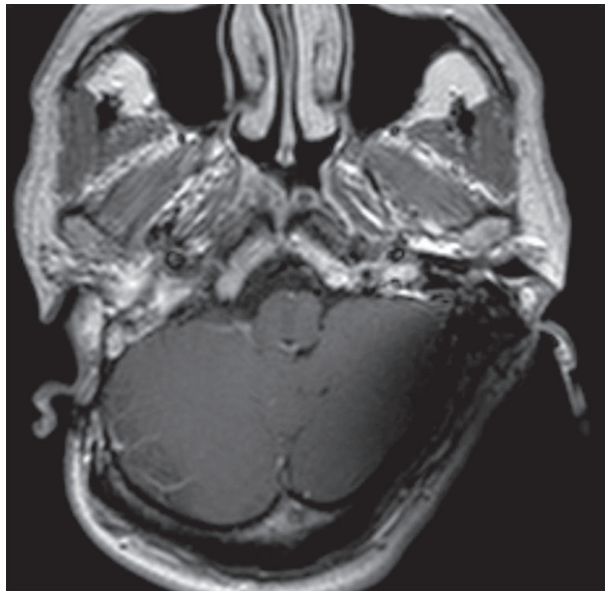

Figure 4. MRI imaging at last follow up, axial T1 weighted with Gadolinium. A paraganglioma centred on the left jugular foramen is seen. Note the artefact produced by the magnet of the CI does not impede visualization of these structures.

Discussion

There is still controversy regarding the proper management of TJP, options include surgical treatment, wait and scan policy, and RT. Most authors agree that surgery is the primary treatment option for this subset of tumors. We believe that RT is not an adequate primary treatment for this subset of tumors. Despite surgery being the preferable mode of treatment in the vast majority of these tumors, it is not always the most prudent choice (Sanna et al, 2004). In case of bilateral tumors, LCN function should be preserved at least unilaterally (Sanna et al, 2011). The possibility of multiple and contralateral lesions should also be considered, especially in the young patient or patients with family history. In these cases, screening for coexisting lesions is indicated, as the presence or absence of synchronous or methachronous tumors would condition any therapeutic intervention. MRI of the entire neck should be obtained, with investigation of the abdomen if there are any signs of catecholamine excess or family history. Octreotide and PET scanning can also be used to screen for multicentricity (Sanna et al, 2013).

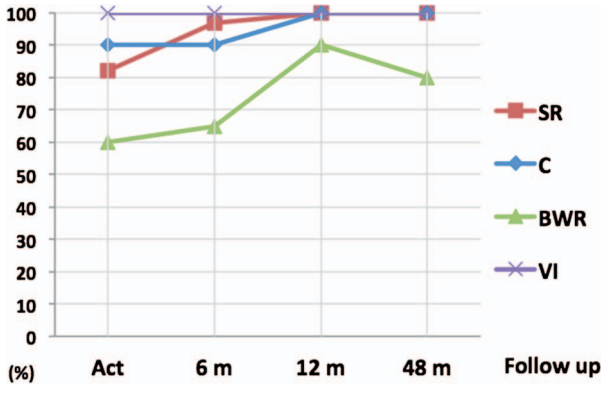

Figure 3. Speech audiometry test results at cochlear implant activation, 6, 12, and 48 months. Note that the results remain stable with time. SR: speech recognition; C: common phrases comprehension; BWR: bysillabic word recognition; VI: vowel identification; Act: activation; m: months.

In a young patient like ours, with a symptomatic, extensive tumor, and the presence of contralateral LCN deficits due to previous surgery, a wait and scan policy cannot be adopted. In such cases, irradiation therapy is preferred to avoid the surgical risks of bilateral LCN deficits.

Bilateral and multiple paragangliomas are rare tumors, for which management has a considerable and direct effect on the patient's life. Whichever modality of treatment is chosen, all of them have the potential to destroy the hearing. In these patients, cochlear implantation offers a new perspective for hearing restoration as long as the cochlea remains intact. As far as we know, there have not been previous reports on CI after irradiated TJP. There has been one previous report of a case of bilateral paraganglioma that received a CI on one side for hearing restoration (Selivanova et al, 2009). In this case, the patient underwent surgical tumor excision on both sides, with CI placement simultaneously at the time of the second surgery. Hearing results were satisfactory, but 18 months later, a revision surgery was performed for partial extrusion of the electrode array through the tympanic membrane. Eight months later the patient complained of progressive hearing loss. Imaging studies showed complete resorption of all cochlea turns. The CI was removed and the histological exam showed no evidence of tumor recurrence. The authors hypothesized that the bone resorption may have occurred as a result of long-silent infection in the cochlea or an unusual foreign body reaction.

This report demonstrates that CI can be safely performed after TJP irradiation with long-lasting, satisfactory results, even in the presence

of residual tumor. If this situation is encountered, it can be overcome by coagulating the tumor with bipolar forceps and subsequent excision to obtain access to the round window. If there is extensive residual tumor after RT treatment, it may be necessary to perform cochlear implantation by subtotal petrosectomy approach with blind sac closure of the external auditory canal, sealing of the eustachian tube and obliteration of the cavity with abdominal fat (Free et al, 2013). This technique allows better access to the round window and facilitates identification of anatomical structures in difficult situations with minimum morbidity. In this case, it was possible to obtain sufficient space through the posterior tympanotomy to coagulate the tumor and insert the CI; however, an initial approach by means of a subtotal petrosectomy would have been a reasonable option as well.

An important issue of paraganglioma patients, either irradiated or surgically treated, is the necessity of follow-up with imaging. We are in agreement with the observation that follow-up should be life-long (Jackson et al, 2001). Radiological follow-up can be performed with HRCT, which allows monitoring of soft tissue density masses at the level of the tympanic cavity and bony erosion of the jugular spine and jugular crest (Sanna et al, 2013). If there is any doubt on any radiological view, a MRI scan up to 1.5 T can be safely performed in patients with a CI with diagnostic efficacy (Crane et al, 2010). Another point to take into consideration is the distortion of the image obtained from the MRI scan in these patients. In the contralateral side of the cranium and remainder of the body there are no artefacts. In the side ipsilateral to the CI, there is an area of distortion usually located above and behind the internal auditory canal that does not hamper visualization of the jugular foramen area (Crane et al, 2010). Therefore, MRI scan up to 1.5 T can be safely obtained after CI procedure, providing adequate visualization of the key anatomical areas involved by TJP growth (Figure 4).

In certain cases, the presence of extensive tissue dissection and subsequent scar tissue make postoperative radiological assessment challenging. When postoperative scans are difficult to interpret, the option of performing an angiography should be considered five years after the treatment (Sanna et al, 2013). The presence of a CI does not represent any contraindication to perform an angiography (Jacobs, 1999).

Conclusion

Indications for cochlear implantation have extended progressively over the years both from an audiological and clinical point of view, with CIs introduced to restore hearing in combination with skull base surgeries and neurotological procedures in the past few years.

Bilateral and multiple paragangliomas are rare tumors. Whichever modality of treatment is chosen for their management, all of them have the potential to destroy the hearing. In these patients, cochlear implantation offers a new perspective for hearing restoration.

This report demonstrates that CI can be safely performed after TJP irradiation with long-lasting, satisfactory results, even in the presence of residual tumor.

The CI can be placed by standard approach through the facial recess, however, if there is extensive residual tumor after RT treatment, it may be necessary to associate a subtotal petrosectomy procedure.

The presence of a CI does not pose a problem for follow-up of paragangliomas. If necessary, both a MRI scan up to 1.5 T and angiography can be safely obtained with diagnostic efficacy in implanted patients.

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References

Adunka O.F. & Buchman C.A. 2007. Cochlear implantation in the irradiated temporal bone. *J Laryngol Otol*, 121(1), 83–6.

Chua D. & Tan H. 2007. Successful rehabilitation with cochlear implant in post-irradiation induced hearing loss in nasopharyngeal carcinoma patient. *Ann Acad Med Singapore*, 36, 74–7.

Crane B.T., Gottschalk B., Kraut M., Aygun N. & Niparko J.K. 2010. Magnetic resonance imaging at 1.5 T after cochlear implantation. *Otol Neurotol*, 31(8), 1215–20.

Free R.H., Falcioni M., Di Trapani G., Giannuzzi A.L., Russo A. et al. 2013. The role of subtotal petrosectomy in cochlear implant surgery: A report of 32 cases and review on indications. *Otol Neurotol*, 34(6), 1033–40.

Jackson C.G., Haynes D.S., Walker P.A., Glasscock M.E. 3rd, Storper I.S. et al. 1996. Hearing conservation in surgery for glomus jugulare tumors. *Am J Otol*, 17, 425–437.

Jackson C.G., McGrew B.M., Forest J.A., Netterville J.L., Hampf C.F. et al. 2001. Lateral skull base surgery for glomus tumors: Long-term control. *Otol Neurotol*, 22, 377–82.

Jacobs J.M. 1999. Diagnostic neuroangiography. Basic techniques. In: Osborn A.G. (ed.), *Diagnostic Cerebral Angiography: Second edition*. Philadelphia: Lippincott Williams & Wilkins, 421–473.

Low W.K., Burgess R., Fong K.W. & Wang D.Y. 2005. Effect of radiotherapy on retro-cochlear auditory pathways. *Laryngoscope*, 115(10), 1823–6.

Sanna M., Jain Y., De Donato G., Rohit Lauda L. et al. 2004. Management of jugular paragangliomas: The Gruppo Otologico experience. *Otol Neurotol*, 25, 797–804.

Sanna M., Piazza P. & Shin S. (eds.). 2013. *Microsurgery of Skull Base Paragangliomas*. New York: Thieme.

Sanna M., Shin S.H., De Donato G., Sivalingam S., Lauda L. et al. 2011. Management of complex tympanojugular paragangliomas including endovascular intervention. *Laryngoscope*, 121(7), 1372–82.

Selivanova O., Gouveris H. & Mann W. 2009. Cochlear implantation after surgery for bilateral jugular foramen paragangliomas: A case report. *Eur Arch Otorhinolaryngol*, 266, 463–464.

Vincenti V., Pasanisi E., Guida M., Di Trapani G. & Sanna M. 2008. Hearing rehabilitation in neurofibromatosis type 2 patients: Cochlear versus auditory brainstem implantation. *Audiol Neurotol*, 13, 273–280.