Clinical Capsule Report

Endoscope-assisted Partial Cochlectomy for Intracochlear Schwannoma With Simultaneous Cochlear Implantation: A Case Report

AQ1 *†Andrew K. Ma and *†‡Nirmal Patel

AQ2 *Sydney Endoscopic Ear Surgery (SEES) Research Group; †Kolling Deafness Research Centre, Royal North Shore Hospital, University of Sydney; and ‡Department of Otolaryngology – Head and Neck Surgery, Faculty of Medicine and Health Sciences, Macquarie University, Sydney, Australia

Introduction: Intralabyrinthine schwannomas are a small subset of vestibular schwannomas which originate within the labyrinthine structures. Management typically consists of watch-and-wait strategies given that surgical intervention will sacrifice hearing. Endoscopic resection of primary intracochlear schwannoma with simultaneous cochlear implantation for a patient with progressive hearing loss and debilitating tinnitus is described.
Patient: A 56-year-old male presenting with asymmetric left sensorineural hearing loss (SNHL) was diagnosed with intracochlear schwannoma on MRI.
Intervention: Surgery was indicated due to tumor growth on serial imaging, worsening SNHL, and severe tinnitus. Partial cochlectomy was performed via transcanal endoscopic approach. Cochlear implantation via mastoidectomy and posterior tympanotomy was simultaneously performed with a CI512 Contour Advanced implant (Cochlear, Sydney, Australia).
Main Outcome Measures: Post partial cochlectomy speech performance.
Results: Preoperative audiometry showed left profound SNHL with 20% speech recognition score despite maximal amplification. Speech perception testing 5 months postoperatively demonstrated good unilateral discrimination when testing the implanted ear alone (BKB sentences 66%, CUNY sentences 79%), open-set comprehension, and excellent binaural performance.
Conclusion: The endoscope offers an additional viable approach to the otic capsule for the removal of intracochlear schwannoma and good audiologic outcomes can be achieved with simultaneous cochlear implantation even after partial cochlectomy. **Key Words:** Cochlear implant—Cochlectomy—Endoscopic ear—Hearing rehabilitation—Intracochlear schwannoma.

Otol Neurotol **40:**xxx–xxx, 2019.

In comparison to the classical vestibular schwannoma which arises within the internal auditory canal and/or cerebellopontine angle, intralabyrinthine schwannomas (ILS) represent a small subset that originate within the labyrinthine structures from either the distal cochlear or vestibular nerves. Few case reports of surgical management of ILS exist in the literature (1–3). Generally, ILS are managed conservatively as surgical extirpation involves sacrifice of remaining hearing function. Herein, a case of intracochlear schwannoma (ICS) managed via endoscopic resection with simultaneous cochlear implantation is presented.

CASE REPORT

A 56-year-old male was first seen in 2014 in consultation for asymmetric mild left-sided SNHL. This was investigated with temporal bone CT and MRI which demonstrated a suspected left intracochlear lesion. The patient was monitored with serial imaging. MRI demonstrated progressive tumor enlargement within the left cochlea (Fig. 1). There was no evidence of vestibular, transmodiolar, or transotic extension. Clinically, the patient continued to have hearing deterioration of the left ear to profound SNHL with increasingly distressing tinnitus. Preoperative audiometry showed poor speech recognition (20% SRS) despite maximal amplification. BKB and CUNY sentence testing was not performed preoperatively. The right ear was audiometrically normal. The patient denied any vestibular symptoms. Given the progressive ICS growth with profound SNHL and tinnitus, the patient provided informed consent to undergo surgical removal. Simultaneous cochlear implant was offered for hearing rehabilitation.

Address correspondence and reprint requests to Andrew K. Ma, M.D., AQ3 ~~F.R.C.S.C.,~~ Kolling Deafness Research Centre, Department of Otolaryngology, Royal North Shore Hospital, Pacific Hwy, St Leonards NSW 2065, Australia; E-mail: andrew.k.ma@gmail.com
AQ4 The authors disclose no conflicts of interest.
DOI: 10.1097/MAO.0000000000002539

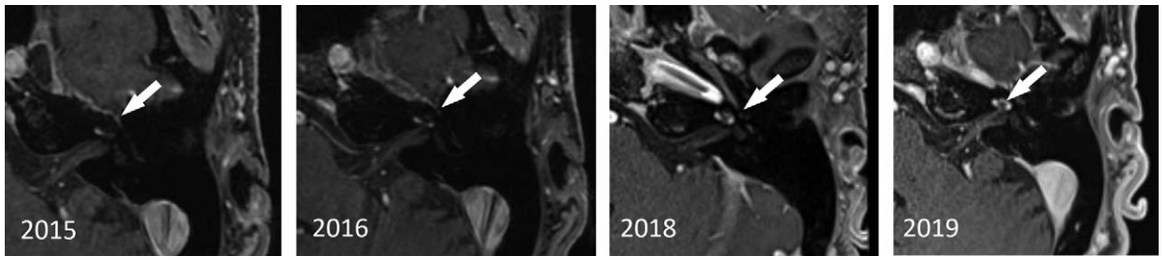

FIG. 1. Axial-plane T1-weighted with gadolinium contrast MRI of the internal auditory canals demonstrates progressive enhancement of intracochlear schwanomma (white *arrow*), originating in the basal turn and extending to involve the middle turn. Note there is no involvement of the IAC fundus or the vestibule.

Operative Details

A transcanal endoscopic approach of the left ear was first performed, with routine elevation of a tympanomeatal flap. The handle of the malleus was removed to improve exposure to the promontory. The bony promontory was then drilled with a standard otologic drill. The lateral basal and second turns of the cochlea were exposed. Endoscopic magnification of the intracochlear contents demonstrated schwannoma throughout the basal and second turn of the cochlea (Fig. 2A). A deliberate effort was made to preserve the modiolus.

Tumor extirpation was performed with suction and gentle dissection, taking care to exert minimal trauma on the modiolus. The anterior, lateral, and posterior segments of the cochlear turns were cleared of tumor. Under endoscopic magnification, a CI depth gauge was guided through the remaining round window groove and passed through the basal turn, with resultant medial tumor pushed through from the middle turn into the middle ear space for removal (Fig. 2B). The depth gauge was gently used in a ''pipe cleaner'' fashion to fully clear the cochlea of tumor. Angled 30° and 45° endoscopes were used to confirm macroscopic clearance of disease.

Once tumor was cleared, mastoidectomy was performed in routine fashion for cochlear implantation. Posterior tympanostomy was developed in the usual fashion. The device used was a CI512 Contour Advance electrode (Cochlear, Sydney, Australia) to allow maximal perimodiolar placement. The electrode was placed through the cochlear remnant and good perimodiolar approximation was directly visualized (Fig. 3). The cochlectomy defect was reconstructed with tragal composite cartilage graft (Fig. 4) and Tisseel, and the tympanomeatal flap and postauricular wound closed in the usual fashion. Intraoperative neural response telemetry demonstrated appropriate impedance in 18 of 21 electrodes, with the distal 3 exposed electrodes demonstrating high impedance.

Postoperative Details

The patient underwent objective speech reception testing 5 months after surgery. With masking of the normal ear and reliance on the implanted ear only, the patient scored 66% on BKB sentences and 79% on CUNY sentences. Testing of binaural speech recognition against competing background noise demonstrated

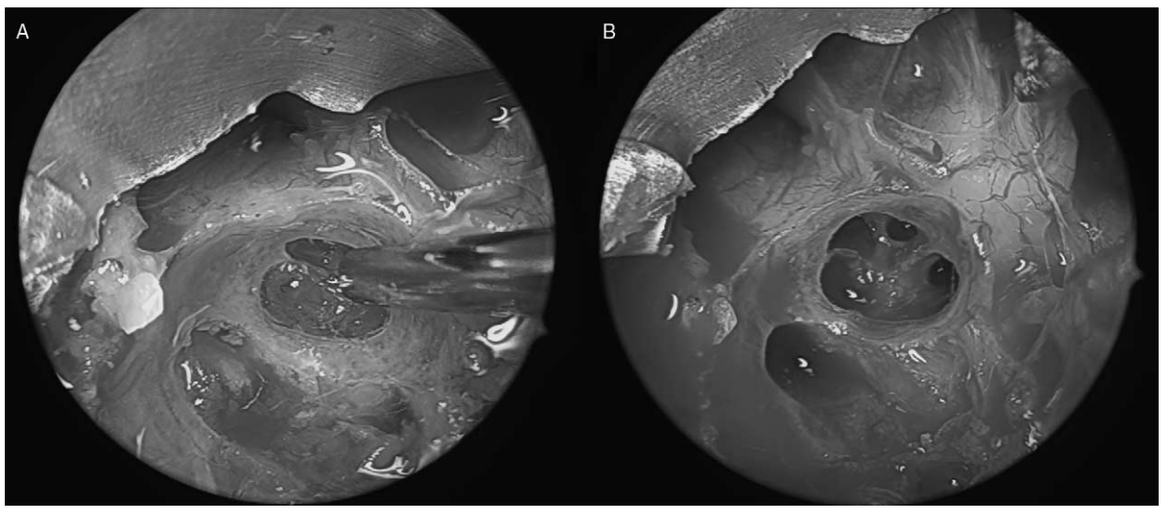

FIG. 2. *A*, Endoscopic view of the left ear after partial cochlectomy demonstrates soft tissue mass seen within the basal and mid-turns of the cochlea. Malleus handle has been removed to improve visualization. *B*, Endoscopic appearance of the left cochlea after complete extirpation of the intracochlear schwannoma. The osseous spiral lamina and modiolus can be seen.

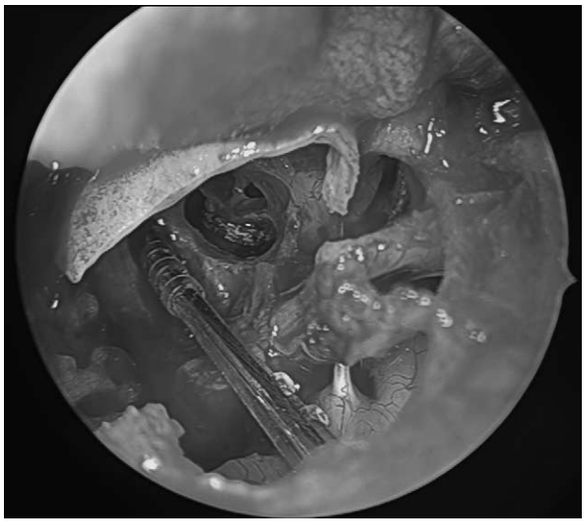

FIG. 3. Insertion of CI512 Contour Advance (Cochlear, Sydney, Australia) into left cochlear remnant, demonstrating good perimodiolar approximation of the electrode.

excellent performance with SNR-50% (signal-to-noise ratio where the patient scores 50% speech recognition) measured at −2.5 dB SNR (sentences presented 2.5 dB softer than competing noise), a result comparable to normal-hearing ears. Binaural performance was excellent, with 91% on BKB sentences at 0 dB SNR. Furthermore, the Tinnitus Reaction Questionnaire (TRQ) was scored at 0 five months postoperatively, compared to 20 preoperatively (>17 suggests clinically disturbing tinnitus).

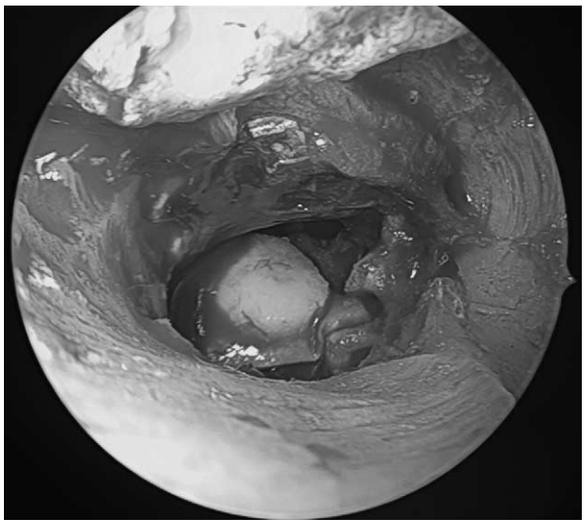

FIG. 4. Repair of cochlectomy defect with tragal composite cartilage graft, with perichodrium packing of the exposed scala deep to the cartilage graft. The repair was then reinforced with Tisseel.

DISCUSSION

ILS are a rare entity on the spectrum of vestibular schwannoma. Management of primary ICS have been described in few case reports and series (1,2,4,5). Most otologists agree that these tumors can be adequately managed via active surveillance with serial MRI. Surgery is indicated in progressive tumor growth, worsening hearing loss, tinnitus, intractable vertigo, or imminent otic capsule ''escape'' into the internal auditory canal. Depending on the location and burden of disease, various surgical approaches are available, from endoscopic/microscopic transcanal transpromontorial, to microscopic translabyrinthine transotic with middle ear obliteration and blind sac closure of the external auditory canal (6–9).

In this patient, a combined approach endoscopic transcanal removal of intracochlear schwannoma with traditional postauricular mastoidectomy and posterior tympanotomy access for simultaneous cochlear implantation was chosen. Endoscopic cochlear implant techniques without mastoidectomy have been described previously (10). However, we elected a traditional postauricular approach for several reasons. Angulation for electrode insertion could be an issue as described in a radiologic study by Tarabichi (11). This is particularly important in partial cochlectomy as the lateral round window niche and wall are removed, which serves as a lateral strut on which the basal electrode rests against. A transcanal insertion approach would present a more perpendicular entry to the angle of the basal turn in a situation where maximal perimodiolar approximation is desired, and better achieved with a posterior tympanotomy approach. Second, the transmastoid approach obviates any potential risk of electrode extrusion through the canal skin.

With respect to tumor extirpation, the visualization of the promontory via endoscope and microscope is not drastically different as the promontory is in direct line of sight via transcanal view. However, one advantage could be the use of angled endoscopes and significant endoscopic magnification of the cochlear scala after partial cochlectomy. It is postulated the improved visualization can reduce the degree of cochlectomy required for complete tumor removal, which could improve the overall hearing outcome as the risk of modiolar injury is reduced with less dissection. Furthermore, as the modiolus and medial cochlea were preserved, the ~~IAC~~, was not entered AQ5 and no significant perilymph or cerebrospinal fluid leakage was noted intraoperatively. The defect was repaired with composite cartilage graft and Tisseel, with closure of tympanic membrane and preservation of the EAC. By avoiding blind-sac closure, the risk of iatrogenic entrapment cholesteatoma is reduced and cosmetic outcome improved.

The completeness of tumor resection can be debated. Endoscopic partial cochlectomy provides excellent visualization into the lateral, anterior, and posterior scala of the cochlea. However, short of sacrificing the modiolus

or exposing the vestibule, the anteromedial aspect of the scala cannot be fully visualized and thus present possible sites of residual tumor. Furthermore, the risk of microscopic intramodiolar tumor extension cannot be ruled out. The surgeon must weigh the benefits of modiolus preservation versus completeness of tumor resection, as there is increasing risk of tumor recurrence with greater burden of residual disease (12). Given the slow growth rate of vestibular schwannoma in general, we favor modiolus preservation and accept a small risk of residual tumor to maximize hearing rehabilitation. In fact, leaving gross residual tumor in situ with insertion of a CI through tumor has been described (2). Another consideration is the ability to repeat MRI surveillance long-term. MRI follow-up has been described even with CI insertion (13), but if MRI quality is significantly reduced by the presence of the CI, magnet removal or even CI explantation may be required to make a diagnosis of recurrent schwannoma.

The audiologic outcomes following partial cochlectomy raise interesting points regarding well-established techniques of ''soft surgery'' of the cochlea. ''Soft surgery'' techniques, first described in 1993 by Lehnhardt, are employed to preserve residual low-frequency hearing in CI candidates (14,15). Full discussion of all soft surgery techniques is beyond the scope of this article, but in particular, speed of electrode insertion as well as use of perioperative steroids have been shown to have significant effect on hearing preservation in CI (16). The goal of these maneuvers is to reduce mechanical trauma as the resultant histopathologic cochlear changes are deleterious—including fibrosis, neo-osteogenesis, hair cell apoptosis, and nonspecific inflammatory responses (17–20). In contrast, this case report highlights a situation of significant cochlear trauma via partial cochlectomy. In spite of significant cochlear trauma, intraoperative neural response telemetry could be seen. Furthermore, subsequent speech threshold testing showed a high degree of accuracy in open-set sentences even when relying on the implanted ear alone. When challenged in the presence of competing background noise, the patient performed exceptionally well, with an SNR-50% of −2.5 dB SNR, considered comparable to the performance of a person with normal binaural hearing. His sentence performance was also strong in the presence of competing background noise. In Plontke's series of 12 ILS patients, CI was performed in 5 patients (2). Of these, two were intracochlear, with one achieving 90% word recognition score at 65 dB, and 0% in the second patient who underwent subtotal cochlectomy (modiolus stump remaining only). At present, CI outcomes following resection of ILS are not completely clear. In light of promising audiologic outcomes despite significant cochlear trauma, it seems appropriate to preserve as much cochlear architecture as possible while facilitating removal of tumor. In the event CI is to be done simultaneously, it is important to preserve the spiral ganglion cells in the modiolus, as well as leave some cochlear framework on which the array would be adequately supported. A perimodiolar array is superior in this setting given partial removal of the lateral cochlear wall. This case highlights that cochlear function could be surprisingly resilient as long as key elements are preserved. Long-term research is lacking as to whether there is a risk of delayed CI failure after cochlectomy.

CONCLUSION

Primary surgical extirpation of ICS via transcanal endoscopic approach is a viable approach. Good audiologic outcomes are attainable with cochlear implantation, even after partial cochlectomy. Preservation of the modiolus and spiral ganglion cells is likely central to CI success. More research is needed to determine the long-term outcomes of CI following cochlectomy.

Acknowledgments: The authors acknowledge James Leask, audiologist at Northside Audiology (Sydney, Australia), who prepared audiometric and speech testing results.

REFERENCES

1. Bento RF, Gebrim EMMS, Maglhaes AT, et al. Minimally invasive surgery for intracochlear schwannoma removal and simultaneous cochlear implantation. *Int Arch Otorhinolaryngol* 2016;20:271–4.
2. Plontke SK, Kösling S, Rahne T. Cochlear implantation after partial or subtotal cochleoectomy for intracochlear schwannoma removal—a technical report. *Otol Neurotol* 2018;39:365–71.
3. Park HS, Choi JW. Combined surgical approach to intralabyrinthine schwannoma. *Eur Ann Otorhinolaryngol Head Neck Dis* 2019. [Epub ahead of print].
4. Plontke SK, Rahne T, Pfister M, et al. Intralabyrinthine schwannomasIntralabyrinthäre Schwannome. *HNO* 2017;65:136–48.
5. Plontke SK, Kösling S, Pazaitis N, Rahne T. Intracochlear schwannomaIntracochleäres Schwannom. *HNO* 2017;65:158–62.
6. Mazzoni A, Zanoletti E, Faccioli C, Martini A. Acoustic schwannoma with intracochlear extension and primary intracochlear schwannoma: Removal through translabyrinthine approach with facial bridge cochleostomy and transcanal approach. *Eur Arch Otorhinolaryngol* 2017;274:2149–54.
7. Xia Y, Zhang W, Li Y, et al. The transotic approach for vestibular schwannoma: Indications and results. *Eur Arch Otorhinolaryngol* 2017;274:3041–7.
8. Alicandri-Ciufelli M, Federici G, Anschuetz L, et al. Transcanal surgery for vestibular schwannomas: A pictorial review of radiological findings, surgical anatomy and comparison to the traditional translabyrinthine approach. *Eur Arch Otorhinolaryngol* 2017;274:3295–302.
9. Marchioni D, Carner M, Soloperto D, et al. Expanded transcanal transpromontorial approach: A novel surgical technique for cerebellopontine angle vestibular schwannoma removal. *Otolaryngol Head Neck Surg* 2018;158:710–5.
10. Dia A, Nogueira JF, O'Grady KM, Redleaf M. Report of endoscopic cochlear implantation. *Otol Neurotol* 2014;35:1755–8.
11. Tarabichi M, Nazhat O, Kassouma J, Najmi M. Endoscopic cochlear implantation: Call for caution. *Laryngoscope* 2016;126:689–92.
12. Nakatomi H, Jacob JT, Carlson ML, et al. Long-term risk of recurrence and regrowth after gross-total and subtotal resection of sporadic vestibular schwannoma. *J Neurosurg* 2017;1–7. AQ6
13. Carlson ML, Neff BA, Sladen DP, Link MJ, Driscoll CL. Cochlear implantation in patients with intracochlear and intralabyrinthine schwannomas. *Otol Neurotol* 2016;37:647–53.
14. Friedland DR, Runge-Samuelson C. Soft cochlear implantation: Rationale for the surgical approach. *Trends Amplif* 2009;13:124–38.

15. Gantz BJ, Turner C, Gfeller KE, Lowder MW. Preservation of hearing in cochlear implant surgery: Advantages of combined electrical and acoustical speech processing. *Laryngoscope* 2005;115:796–802.
16. Nguyen S, Cloutier F, Philippon D, et al. Outcomes review of modern hearing preservation technique in cochlear implant. *Auris Nasus Larynx* 2016;43:485–8.
17. Nadol JB, Shiao JY, Burgess BJ, et al. Histopathology of cochlear implants in humans. *Ann Otol Rhinol Laryngol* 2001.
18. Shepherd RK, Clark GM, Xu SA, Pyman BC. Cochlear pathology following reimplantation of a multichannel scala tympani electrode array in the macaque. *Am J Otol* 1995;16:186–99.
19. Marsh MA, Jenkins HA, Coker NJ. Histopathology of the temporal bone following multichannel cochlear implantation. *Arch Otolaryngol Neck Surg* 1992;118:1257–65.
20. Rizer FM, Arkis PN, Lippy WH, Schuring AG. A postoperative audiometric evaluation of cochlear implant patients. *Otolaryngol Neck Surg* 1988;98:203–6.

AQ7

MAO

Manuscript No. ON-19-760

Otology & Neurotology

Dear Author,

During the preparation of your manuscript for typesetting, some queries have arisen. These are listed below. Please check your typeset proof carefully and mark any corrections in the margin as neatly as possible or compile them as a separate list. This form should then be returned with your marked proof/ list of corrections to the Production Editor.

QUERIES: to be answered by AUTHOR

QUERY NO.	QUERY DETAILS	RESPONSE
<AQ1>	Please confirm whether surnames/family names (red) have been identified correctly in the author byline.	
<AQ2>	Please check the affiliation for correctness.	
<AQ3>	Please check the correspondence address for correctness.	
<AQ4>	Please check the accuracy of the conflicts of interest statement.	
<AQ5>	Please spell out ‘‘IAC.’’	
<AQ6>	Please provide volume in ref. (12).	
<AQ7>	Please provide volume and page range in ref. (17).	