Management and Outcomes of Transotic Schwannoma: A 3 Case Series

Sophie Jabban, BA[1]; Michael Bartellas MD, MSc[1]; Mari Hagiwara, MD[2]; J. Thomas Roland Jr. MD[1]; John Golfinos, MD[3]; Donato Pacione, MD[3]; David Friedmann MD, MSc[1]

[1]NYU Langone Department of Otolaryngology
[2]NYU Langone Department of Radiology
[3]NYU Langone Department of Neurosurgery

Department of Otolaryngology — Head & Neck Surgery
Department of Neurosurgery

ABSTRACT

We present a case series of three adult patients with radiographically diagnosed transotic schwannomas (TOS), highlighting diagnostic and surgical considerations in this rare pathology. Patients presented with a spectrum of symptoms including sensorineural hearing loss, aural fullness, vertigo, tinnitus, and facial nerve dysfunction. Management strategies included microsurgical resection, stereotactic radiosurgery, and active surveillance, selected based on tumor extent, growth behavior, and baseline hearing status. This series highlights the unique diagnostic and therapeutic challenges posed by TOS due to their ability to mimic middle ear pathology, underscoring the importance of intentional surgical approach selection, multidisciplinary decision-making, and long-term follow-up to optimize outcomes.

INTRODUCTION

Transotic schwannomas (TOS) are a rare subtype of inner ear schwannoma (IES) defined by their involvement of the internal auditory canal, inner ear, and middle ear. While incidence rates are not well-defined, the incidence of IES is approximately 1 in 100,000, with one study identifying TOS in 2 of 45 IES cases. Management of IES is complex and centers on precise tumor location and patient symptomatology. Given this tumor's rarity and diverse treatment modalities, our study aims to outline the management and outcomes of 3 patients with TOS.

METHODS AND MATERIALS

We performed a single-institution retrospective review and identified three patients diagnosed with TOS between 2013 and 2025. Pre-op symptoms, imaging, and management decisions and rationale were documented for each patient. Pre- and post-op House-Brackmann (HB) scores and audiogram assessments, post-op complications, and post-op symptom resolution were collected. We report a summary of presentations and outcomes in TOS treated at our institution.

RESULTS

Case 1. A 59-year-old presented with ipsilateral facial spasms, vertigo, and SNHL for 6 to 8 weeks. The patient first experienced hearing loss 18 years prior to initial presentation. MRI demonstrated an enhancing mass in the right IAC extending to the cerebellopontine angle (CPA), cochlea, vestibule, semicircular canals (SCC), and middle ear (**Figure 1**). After an initial period of active surveillance, Gamma Knife radiosurgery (GKRS) was performed for interval tumor growth at five years, after which the patient experienced complete resolution of facial weakness without any complications. No tumor progression was noted seven years following treatment.

Case 2. A 63-year-old presented with tinnitus, vertigo, and SNHL. MRI demonstrated an enhancing mass in the left IAC, extending to the CPA, vestibule, cochlea, and middle ear (**Figure 2**). After two years of active surveillance, the patient underwent microsurgical resection via a left retrosigmoid approach at an outside institution due to interval tumor growth. Pathology demonstrated a Grade I schwannoma with a Ki-67 index of 5%. Schwannoma recurrence was noted two years after initial resection, leading to re-resection at an outside institution. The patient experienced two additional recurrences, two and five years later, both managed with GKRS. MRI from the time of the second recurrence demonstrated a 4 by 5mm enhancing soft tissue mass in the distal left IAC, while the MRI from the third recurrence demonstrated growth of the tumor with extension along the facial nerve to the geniculate as well as invasion of the petrous bone and middle ear. After each session of GKRS, they developed House-Brackmann grade II then IV facial weakness with partial improvement following facial physical therapy.

Case 3. A 28-year-old presented with progressive ear fullness, vertigo, tinnitus, ipsilateral facial-twitch, and ipsilateral profound SNHL from childhood. Otoscopic exam revealed a mass behind the intact right tympanic membrane raising prior concern for a paraganglioma (**Figure 3**). MRI demonstrated an enhancing mass within the right IAC with extension to the CPA, basal and middle turns of the cochlea, vestibule, proximal SCC, and middle ear (**Figure 4**). Following multidisciplinary review, the surgical team planned for a transcochlear approach given intracochlear tumor involvement. Pathology demonstrated a Grade I schwannoma with a Ki-67 index of 10%. Postoperatively, the patient had normal facial function. At one week, they developed a pseudomeningocele and underwent lumbar drain placement with resolution. They developed delayed ipsilateral facial weakness that resolved completely after one month. The patient exhibited no sign of recurrence on MRI at the last follow-up.

FIGURES

Figure 1. Case 1 Pre-op

Figure 2. Case 2 Pre-op

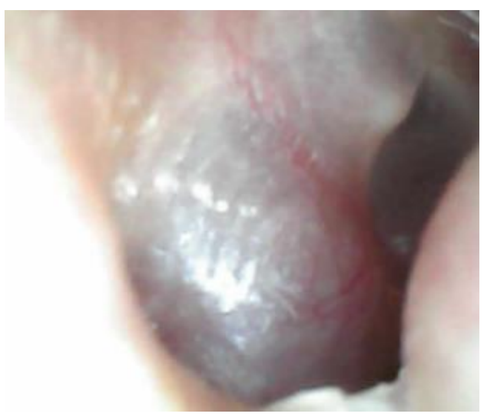

Figure 3. Case 3 Otoscopic Exam

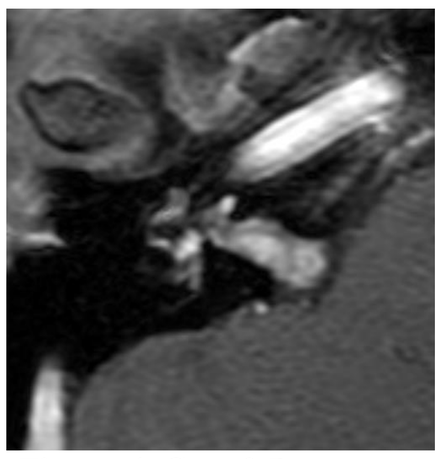

Figure 4. Case 3 Pre-op

DISCUSSION

These cases underscore diagnostic complexity and the importance of maintaining a broad differential in patients with progressive vestibular symptoms. Key to diagnosing these cases include multimodal imaging. These diagnostic tests must be considered in conjunction with history and physical examination to establish the diagnosis of this rare pathology.

These cases also emphasize the importance of surgical approach selection in TOS management, as limited exposure of the middle ear and intralabyrinthine components may increase the risk of residual disease. The retrosigmoid and translabrynthine approaches do not afford access to the middle ear components. In contrast, the transcochlear approach combined with blind sac closure of the ear canal provides full direct access to the middle ear space and the cochlea. Our institutional experience with this rare pathology supports consideration of a transcochlear approach in patients with TOS, especially if long-term tumor control is the primary objective.

REFERENCES

1. Salzman KL, Childs AM, Davidson HC, Kennedy RJ, Shelton C, Harnsberger HR. Intralabyrinthine Schwannomas: Imaging Diagnosis and Classification. *AJNR Am J Neuroradiol*. 2012;33(1):104-109. doi:10.3174/ajnr.A2712
2. Gibbon FL, Lindner RJ, Vial ADM, et al. Translabyrinthine versus Retrosigmoid Approach for Vestibular Schwannoma: A Systematic Review and An Updated Meta-Analysis. *Otolaryngol--Head Neck Surg Off J Am Acad Otolaryngol-Head Neck Surg*. 2025;172(3):774-786. doi:10.1002/ohn.1031
3. Ahmad RARL, Sivalingam S, Topsakal V, Russo A, Taibah A, Sanna M. Rate of recurrent vestibular schwannoma after total removal via different surgical approaches. *Ann Otol Rhinol Laryngol*. 2012;121(3):156-161. doi:10.1177/000348941212100303
4. Obaid S, Nikolaidis I, Alzahrani M, Moumdjian R, Saliba I. Morbidity Rate of the Retrosigmoid versus Translabyrinthine Approach for Vestibular Schwannoma Resection. *J Audiol Otol*. 2018;22(4):236-243. doi:10.7874/jao.2018.00164

Corresponding Author: Michael Bartellas, MD, MSc

Email: Michael.Bartellas@nyulangone.org;