

## Introduction

Ganglioneuromas are slow growing, benign tumors derived from neural crest origin. They typically arise in the posterior mediastinum and retroperitoneum. Occasionally these tumors present as intracranial lesions; with very rare reports of tumors located in the internal auditory canal (IAC). We report a rare case of ganglioneuroma of the IAC. We also review the current literature on ganglioneuroma of the skull base.

## Methods

Case report and literature review.

## Results

A 33 year-old male presents with progressively worsening left-sided hearing loss over the course of 3 years. MRI showed an enhancing soft tissue mass within the left internal auditory canal measuring 10.5 mm by 5 mm in dimension, with slight protrusion of the mass into the cerebellopontine angle (Figure 1).

A left retrosigmoid transtemporal skull base approach was utilized for resection of the intracanalicular posterior fossa tumor. The internal auditory canal was skeletonized to expose the underlying tumor and cranial nerves. A well-encapsulated neoplasm was seen arising from the superior vestibular branch. The tumor was easily excised from the underlying nerve.

## Results, continued

Histologically, the tumor consisted of numerous ganglion cells scattered among a spindle cell background. The ganglion cells expressed Neu-N, while the spindle cell component expressed variable positivity for S-100 protein. The morphology and immunophenotype of this neoplasm was consistent with the diagnosis of ganglioneuroma.

## Conclusions

Intracranial ganglioglioma have been found to occur in the cranial skull base including the temporal bone and sphenoid wing; as well as, associated with the trigeminal nerve. Only two previous reports of ganglioglioma of the ICA are known in the literature<sup>1,2</sup>. These tumors typically are not treated with chemotherapy or radiation. Gross total resection is recommended as malignant transformation is possible. Prognosis is favorable with complete surgical excision. We suggest ganglioneuromas be considered in the differential for intracanalicular ICA lesions.

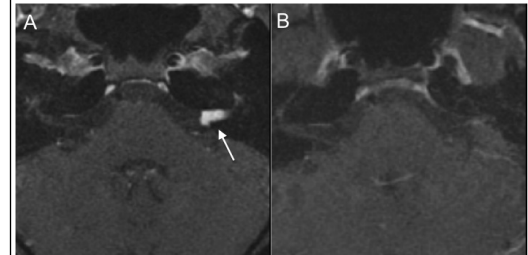
## Learning Objectives

1. Identify the clinical presentation, radiographic features, and intraoperative appearance of ganglioneuroma of the internal auditory canal.
2. Review the histopathological features of ganglioneuroma.
3. Review the management and treatment of intracranial ganglioneuroma.

## References

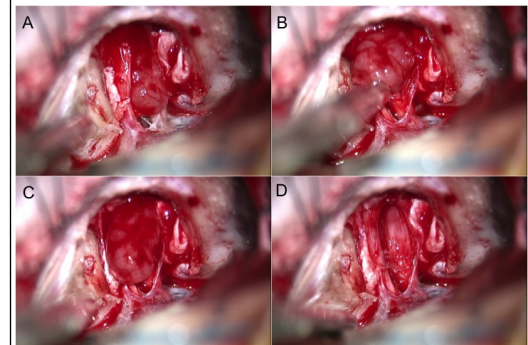
1. Ozluoglu LN, Yilmaz I, Cagici CA, Bal N, Erdogan B. Ganglioneuroma of the internal auditory canal: a case report. *Audiol Neurootol*. 2007; 12: 160-164.
2. Bekelis K, Meiklejohn DA, Missios S, Harris B, Saunders J, Erkmen K. Ganglioneuroma of the Internal Auditory Canal Presenting as a Vestibular Schwannoma. *Skull Base Rep*. 2011; 1: 89-94.

**Figure 1**



(A) Pre-operative axial T1-post-gadolinium image demonstrating an enhancing tumor in the internal auditory canal consistent with an acoustic neuroma. The tumor was removed via a left retrosigmoid transmeatal approach. Pathological examination revealed a ganglioneuroma. (B) Post-operative fat-suppressed axial T1-post-gadolinium MRI demonstrates complete resection of the tumor.

**Figure 2**



(A-C) Intraoperative photographs demonstrating dissection of the tumor away from the facial and vestibulocochlear nerves via a retrosigmoid transmeatal approach. (D) Final view of the facial and vestibulocochlear nerves after complete tumor removal.