Intracanalicular Venous Malformation of the Internal Auditory Canal

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ABSTRACT

Introduction: Venous malformations within the internal auditory canal are exceedingly rare. They may clinically and radiographically mimic the presentation of vestibular schwannomas.

Study Design: Case report.

Discussion: We report a 54 year old male who presented to our clinic with persistent debilitating vertigo, right-sided tinnitus, rapidly progressing right-sided hearing loss, and facial spasm for approximately 2 years. Physical exam was largely unremarkable with exception of lateralization to the left on Weber test. Audiogram demonstrated profound unilateral sensorineural hearing loss (SNHL) on the right and on MRI, an 8x4mm lesion with patchy, partial contrast enhancement within the right IAC was identified.

The patient underwent a translabrynthine approach to resection of the lesion. The fundus appeared dilated on intraoperative inspection. The lesion appeared vascular, adherent, and soft. The resection was uncomplicated. Post-operatively the patient had resolution of his tinnitus, vertigo, and facial spasms. He has mild facial weakness (HB 2/6) that is currently being observed. Histopathologic evaluation of the surgical specimen demonstrated fibrous tissue containing large vascular spaces, consistent with venous malformation.

Conclusions: Venous malformations of the IAC are rare and can present similarly to vestibular schwannomas. Though these lesions are typically small, patients commonly present with facial nerve abnormalities and/or worse than would be expected SNHL. On MRI post-contrast imaging, enhancement is patchy and partial in contrast to avid and homogenous (in non-cystic areas) enhancement of VS.

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INTRODUCTION

Nearly 90% of cerebellopontine angle (CPA) and internal auditory canal (IAC) tumors are vestibular schwannomas (VS) [1,2]. Vascular malformations of the IAC and CPA are rare lesions that may clinically and radiographically mimic VS. Vascular malformations are typically categorized as arteriovenous malformations, cavernous malformations, venous malformations (VMs), and telangiectasias.[3] They are hamartomas that can cause tumor-like, compressive local effects when they enlarge from hemorrhage.[3]

Patients with vascular malformations may have identical symptoms to those presenting with VS however these patients tend to have increased likelihood of facial nerve abnormalities and more severe than would be expected hearing loss.[1,4]

We present a case of a 54 year old male with a venous malformation of the distal right IAC.

CASE PRESENTATION

History

A 54 year old male presented to our clinic with 2 years of persistent episodic vertigo, right-sided tinnitus, and rapidly progressing right-sided hearing loss. He also endorsed periodic facial twitching over the right forehead/ brow region that was infrequent. Audiogram performed at initial onset of symptoms demonstrated mild right-sided SNHL however audiogram performed nearly a year later demonstrated progression to profound right-sided SNHL. The episodic vertigo was particularly distressing to the patient as it was interfering with his occupation.

Patient denied other symptoms including fever, headache, otalgia, and otorrhea. Patient's medical history was unremarkable. No history of trauma.

Clinical Exam

On physical exam , no facial spasm or asymmetry was appreciated. There was no nystagmus. The ear canals and tympanic membranes were normal appearing bilaterally. Lateralization to the left was appreciated on the Weber test. Rinne test was unremarkable. Audiogram demonstrated profound right-sided SNHL across all frequencies.

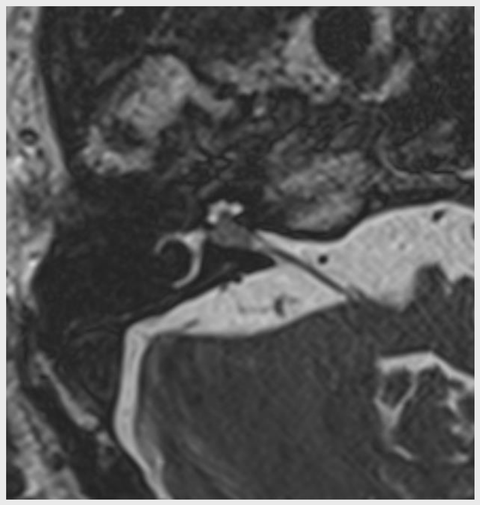
Magnetic Resonance Imaging (MRI)

T2-weighted images demonstrated a 0.8 x 0.4cm filling defect in the distal right internal auditory canal **(Figure 1**), extending to the fundus and filling the cochlear aperture. No evidence of extension into the inner ear structures was apparent. On post-contrast imaging, patchy and partial enhancement of the lesion was appreciated, greatest laterally at the fundus. Progressive increased central enhancement within the lesion was seen on sequential imaging (**Figure 2a / 2b)**.

Management

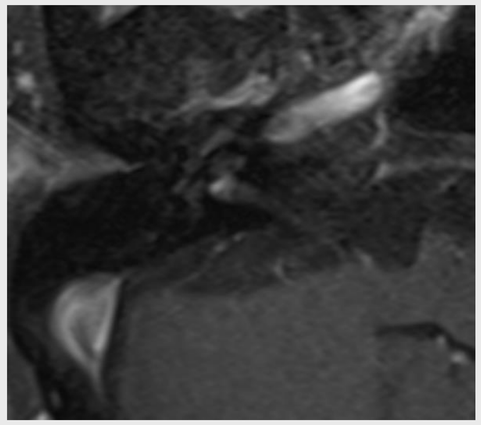
Because of the patient's debilitating vertigo and lack of serviceable hearing, a translabrynthine resection of the intracanalicular lesion was performed. The lesion appeared to be very adherent to the superior vestibular nerve and facial nerve near the fundus and was vascular appearing. The lesion was excised and noted to be soft and compressible.

Figure 1.

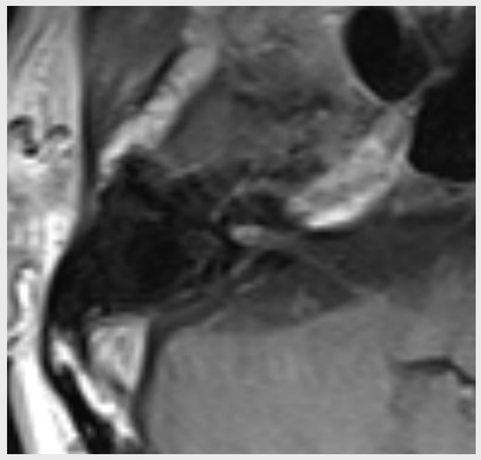


Axial orientation, MRI T2-weighted CISS pulse sequence (constructive interference in steady state).

Figure 2.



***a.** Axial orientation, MRI T1-weighted post-contrast with fat saturation. 3mm focus of enhancement.*



***b.** Axial orientation, MRI T1-weighted post-contrast with fat saturation, image captured 5 minutes after above image Figure 2a. 5mm focus of enhancement.*

Post-Operative Course

The patient's vertigo and tinnitus resolved postoperatively. He has not experienced any further facial twitching however he has a mild facial weakness (HB II) after 6 months of follow-up which is currently being observed. The histopathologic examination of the lesion demonstrated large vascular spaces lined with thin endothelium, surrounded by fibrous tissue. These findings are consistent with VM.

CONCLUSIONS

VMs are a type of vascular malformation. Their occurrence in the IAC is exceedingly rare with only a few cases reported. They are congenital in origin and when arising in the IAC, are thought to develop from the vascular plexus of Scarpa's ganglion.[5-7] They are low-flow, low-resistance lesions that have minimal risk of hemorrhage, estimated to be about 0.22% per year.[1] Pre-operative diagnosis is challenging as imaging and clinical characteristics typically overlap with those of VS.

Despite their typically diminutive size, patients commonly present with rapidly progressing unilateral hearing loss and/or facial nerve abnormalities (i.e. spasm or paresis), depending on the location of the lesion.[1,4,5] This is in contrast to VS of a similar size, where facial nerve abnormalities would be unlikely and hearing loss would be expected to progress at a slower rate.

MRI imaging characteristics may help to discern the diagnosis of VM. Following gadolinium administration, enhancement of the lesion is typically partial or heterogeneous whereas it is typically homogeneous and avid (in non-cystic areas) in VS.[3,7,8] On sequential post-contrast imaging, progressive central enhancement within the lesion may be identified.

Given the low risk of hemorrhage, many advocate for close observation.[8] However if progressive neuropathies are evident, microsurgical resection of these lesions may provide relief.

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