Radiology Case Reports

Volume 5, Issue 2, 2010

Asymmetric sensorineural hearing loss caused by vestibular schwannoma: Characteristic imaging features before and after treatment with stereotactic radiosurgery

Nicholas Krause, MD; Kathleen Tozer Fink, MD; and James R. Fink, MD

We report the case of a 71-year-old man who presented with a 2-year history of progressive left-sided hearing loss caused by a cerebellopontine angle mass lesion with classic imaging characteristics of a vestibular schwannoma. Vestibular schwannomas are typically diagnosed on dedicated MRI of the internal auditory canals obtained for asymmetric sensorineural hearing loss, as in this case. We review the characteristic imaging features of vestibular schwannomas that enable their differentiation from other mass lesions of the cerebellopontine angle cistern, allowing for treatment with stereotactic radiosurgery in this case.

Introduction

Mass lesions in the cerepellopontine angle (CPA) are not easily differentiated based on clinical symptoms and physical examination. Asymmetric sensorineural hearing loss is most commonly associated with vestibular schwannomas (VS), as they arise directly from the sheath of the vestibular nerve in the internal auditory canal (IAC). However, meningiomas and other mass lesions that affect the vestibulocochlear nerve along its cisternal or intracannalicular course may present in a similar manner to VS. Differentiation of these lesions is important because it alters clinical decision-making and treatment planning.

Magnetic resonance imaging (MRI) provides an essential means for characterizing and distinguishing CPA mass lesions. Knowledge of classic imaging features such as lesion shape and the presence or absence of adjacent hyperostosis, calcification, or a dural tail can increase diagnostic specificity and confidence. Our case illustrates how asymmetric sensorineural hearing loss resulting from a CPA mass lesion with classic imaging features for VS may be directly treated with stereotactic radiosurgery.

Case report

A 71-year-old man presented with a two-year history of progressive left-sided hearing loss. Though hearing loss had been gradual, he was unable at presentation to use a phone with his left ear or hear people talking to him if they were standing to his left. He denied any history of head trauma, otologic surgery, or occupational noise exposure. He also denied tinnitus or vertigo. An MRI performed for evaluation of asymmetric sensorineural hearing loss revealed an extra-axial ice-cream-cone-shaped mass in the left CPA, centered at the porus acousticus and extending into the fundus of the IAC. The mass was isointense to brain on T1- and T2-weighted images. Postcontrast images demonstrated uniform enhancement of the mass without evidence of a dural tail. The imaging appearance was classic for a VS (Fig. 1, A-D).

Treatment options for VS were discussed. Due to the patient's age, the relatively small size of the tumor, and an estimated less-than-50% chance of preserving hearing on the affected side from skull-base microsurgery, the patient

Citation: Krause N, Fink KT, Fink JR. Asymmetric sensorineural hearing loss caused by vestibular schwannoma: Characteristic imaging features before and after treatment with stereotactic radiosurgery. *Radiology Case Reports.* [Online] 2010;5:437.

The authors are all in the Department of Radiology, University of Washington, Seattle WA. Contact Dr. Fink at jrfink@uw.edu.

Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v5i2.437

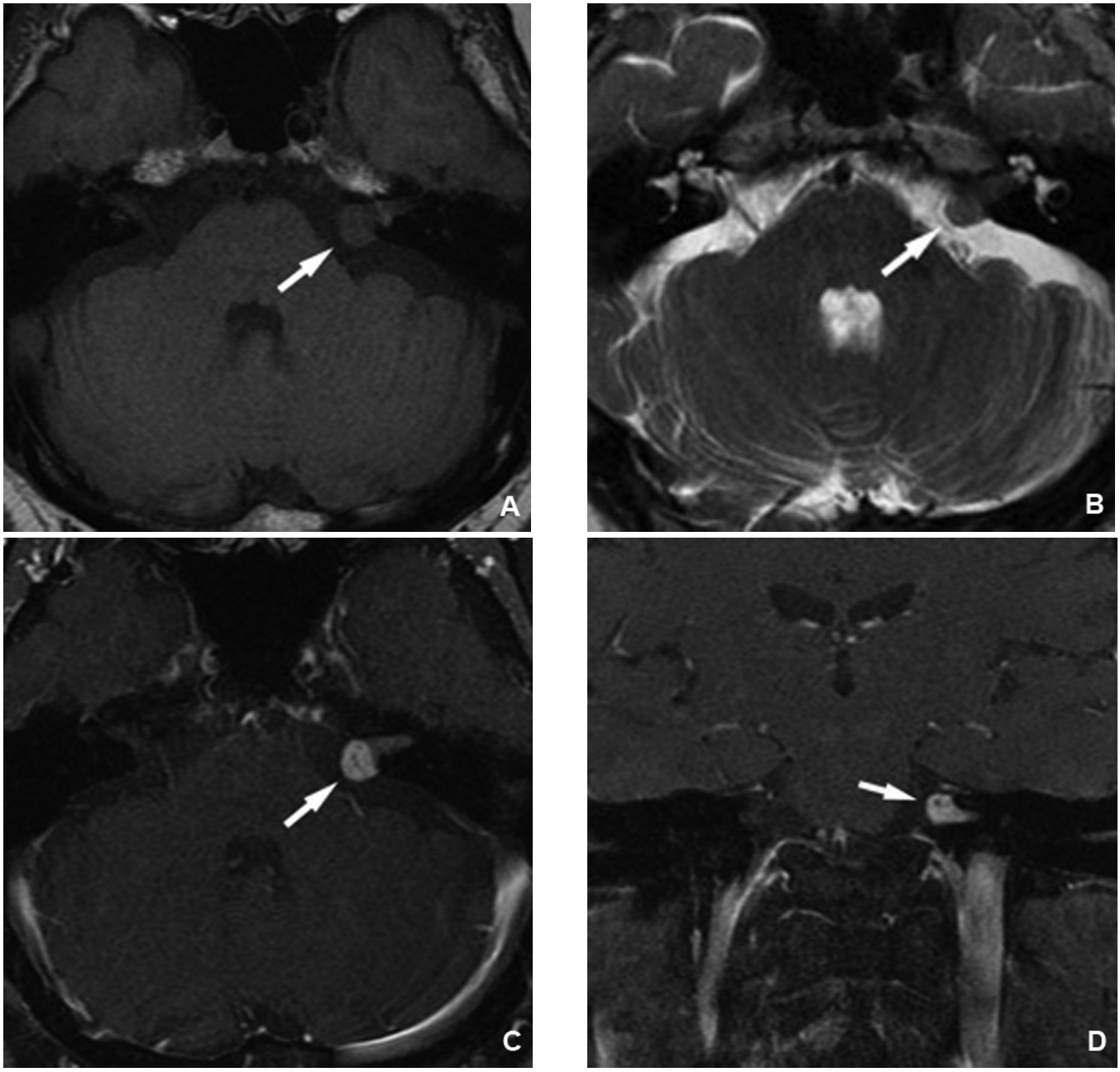

Figure 1. 71-year-old man with asymmetric sensorineural hearing loss. Contrast-enhanced MRI of the brain and IACs. Axial precontrast T1- (A) and T2-weighted (B) images show an ice-cream-cone-shaped mass centered in the left porus acousticus that is isointense to brain parenchyma. Axial (C) and coronal (D) postcontrast fat-suppressed T1-weighted images show avid enhancement of the lesion, without evidence of a dural tail. The findings are characteristic of a vestibular schwannoma (white arrows).

elected to undergo treatment with stereotactic radiosurgery. The lesion was treated by gamma knife with 12 Gray delivered to the 50% isodose surface. On followup MRI three months after radiotherapy, the lesion appeared slightly larger with new areas of nonenhancement centrally, consistent with necrosis (Fig. 2A). Followup MRI approximately three years after treatment showed slight interval decrease in size of the tumor as compared to the pretreatment baseline MRI (Fig. 2B).

Discussion

Vestibular schwannomas typically arise from Schwann cells lining the inferior vestibular nerve. Formerly called acoustic neuromas, these benign tumors are the most

common mass lesions in the CPA, accounting for 70% to 80% of all masses in this region and 8% to 10% of intracranial tumors overall (1, 2). The tumor most often presents in the fifth and sixth decades of life (3). Postmortem examination reveals an incidence of 2.7%, though the actual incidence in clinical practice is much lower, suggesting that most VS are completely asymptomatic (4). Symptomatic patients typically present with unilateral sensorineural hearing loss, tinnitus, and/or disequilibrium. VS are commonly seen in patients with neurofibromatosis type 2 (NF 2), a phakomatosis related to a mutation on chromosome 22 in which patients have multiple schwannomas, meningiomas, and ependymomas. Bilateral VS are diagnostic of NF 2.

Like the other common lesions in the CPA region, such as meningiomas and epidermoid cysts, VS are extra-axial masses. While the classic ice-cream-cone-shaped VS involves the IAC (cone) with extension into the CPA cistern (ice-cream scoop), isolated intracanalicular and purely intracisternal lesions can also occur. Purely intracisternal schwannomas are typically much larger at presentation, likely because they have more room to grow before becoming symptomatic. Large, purely intracisternal schwannomas can cause mass effect on the brainstem, fourth ventricle, and cerebellum, leading to presenting symptoms other than hearing loss (5).

Contrast-enhanced MRI is the method of choice for imaging tumors of the CPA, including VS. According to the schema of Bonneville et al. (6, 7), enhancing lesions are divided by anatomic compartment: intra-axial, extra-axial, and skull-base. Extra-axial contrast-enhancing lesions account for 80% to 95% of CPA masses. Of these, VS is by far the most common CPA tumor, accounting for over two-thirds of all such cases.

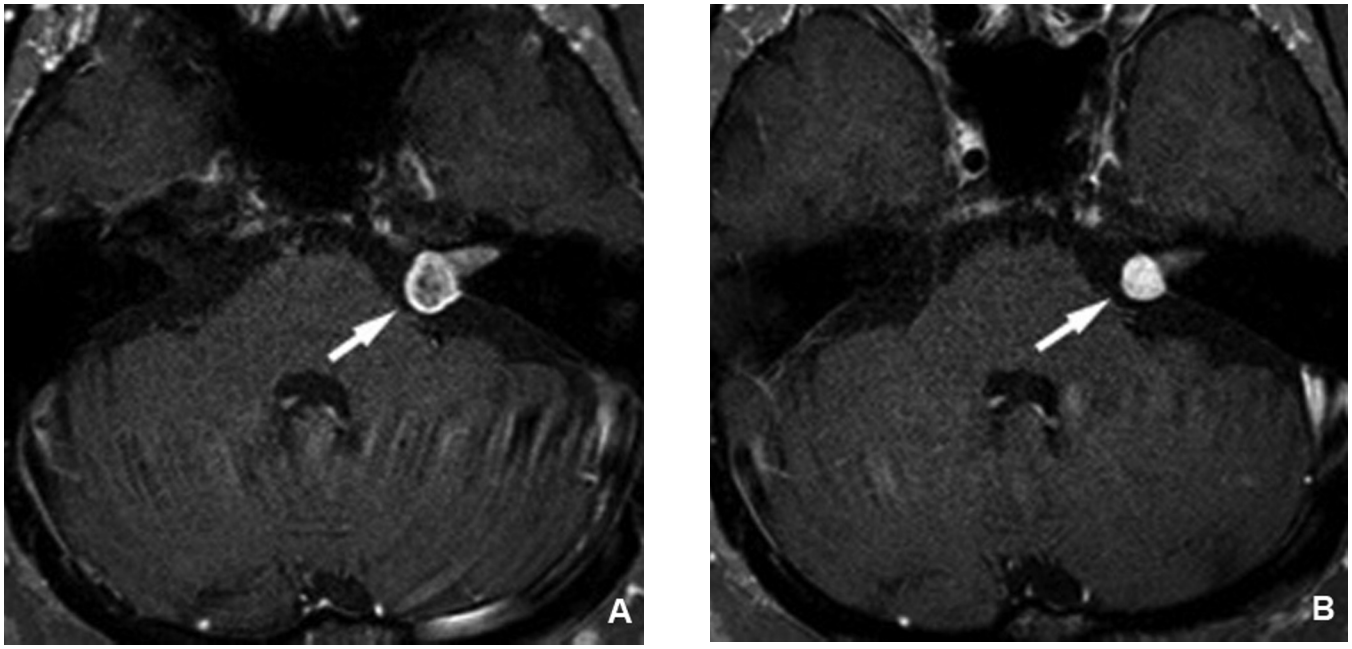

Figure 2. 71-year-old man with asymmetric sensorineural hearing loss. Serial followup contrast-enhanced, T1-weighted, fat-suppressed axial MRI sequences obtained after gamma knife radiosurgery. The left-sided vestibular schwannoma (white arrows) shows slight interval enlargement with central nonenhancement of the cisternal component at three months post-treatment (A), consistent with early post-treatment changes expected after stereotactic radiosurgery. Three years after gamma-knife treatment (B), the lesion has not increased in size; the size of the cisternal component is slightly decreased compared to the pretreatment baseline scan (Fig. 1C, above).

Dedicated MR imaging protocols for evaluation of CPA masses (or other causes of retrocochlear pathology in the setting of asymmetric sensorineural hearing loss) generally consist of precontrast T1- and T2-weighted images, postcontrast T1 with fat suppression, and thin-section T2-weighted MR cisternography (8, 9). Typical imaging characteristics for VS include a soft-tissue mass in the CPA/IAC that is isointense to brain on precontrast T1 and hyperintense on standard T2, though hypointense on MR cisternography. Schwannomas typically enhance strongly, though more heterogeneous enhancement and even cystic components are often seen with larger lesions (10, 11). On CT, VS appear isodense to brain parenchyma on noncontrast images, with enhancement on postcontrast images. VS involving the IAC can remodel the porus acusticus, particularly if large.

Differentiating VS from other common CPA lesions, such as meningiomas, is important because the treatment and operative approach can differ (12). Clinical signs and symptoms are unreliable, as the clinical triad of sensorineural hearing loss, tinnitus, and disequilibrium has only 10% specificity for VS , with most CPA meningiomas also presenting with hearing loss (13). Imaging plays a key role in differentiating these two entities. CT can be helpful, as 25% to 36% of meningiomas have intratumoral calcification, a rare finding in schwannomas. Meningiomas can cause hyperostosis of the adjacent bone, whereas VS erode or widen the IAC. On MRI, a "dural tail sign," indicating enhancement of thickened peritumoral dura, is most characteristic of meningiomas (12).

Additional MRI sequences may provide further information that can increase diagnostic specificity and confidence in difficult cases. On T2*-weighted GRE images, intralesional microhemorrhages are highly specific for VS (14). Diffusion-weighted MR imaging is unreliable in differentiating the two; while mean ADC values tend to be higher in VS than meningiomas, there is significant overlap (15). In challenging cases, more advanced imaging techniques such as perfusion-weighted MR imaging and MR spectroscopy can play a role. Using dynamic susceptibility-weighted perfusion-MR imaging, the relative cerebral blood volume (rCBV) ratio of VS (3.23 +/- 0.81) is significantly lower than meningiomas (8.02 +/- 3.89), though some overlap can exist (16). With MR spectroscopy, meningiomas characteristically show the presence of an alanine peak at 1.5 ppm, while schwannomas lack the alanine peak and instead show the presence of myo-inositol at 3.55 ppm (17).

Once the diagnosis of VS is established, imaging plays a key role in both treatment planning and followup. Treatment strategies include watchful waiting, radiotherapy, and surgery, and multiple clinical variables factor into the initial mode of treatment, including the age of the patient, coexisting conditions, hearing status in both ears, patient and physician preference, and NF 2 status (18). The primary goal of treatment is to control tumor growth, though secondary aims include minimizing symptoms and complications. Evidence-based guidelines comparing the different treatment modalities are lacking, and both patient preference and physician bias often play a strong role in treatment decision-making (19). While VS growth rate tends to be slow, the actual growth rate among individual cases is variable. Furthermore, tumor size does not always correlate with symptoms. A greater likelihood of future growth is seen in larger lesions, younger patients, an extracanalicular location, and documented prior growth (20, 21).

Watchful waiting is often a reasonable initial strategy for older individuals, for patients with comorbidities that make them poor surgical candidates, and for small tumors. Typically, these patients will have audiovestibular symptoms monitored regularly as well as serial MR examinations (often once per year) to assess for growth (22). While followup examinations are traditionally performed with pre- and post-contrast images, a recent study suggests that noncontrast MR cisternography or similar thin-slice T2 MRI may be sufficient for following lesion size (23). While lesion size is generally assessed in two dimensions, recent evidence suggests that volume quantification may be more accurate for followup (24).

Imaging plays a key role in planning for radiotherapy or surgery, and can help with creating radiotherapy treatment fields or determining the surgical approach. Relevant variables include tumor size, relation to the facial nerve, and intralabyrinthine signal intensity. While nonoperative approaches are often used for small tumors, surgery is indicated for lesions greater than 3 centimeters (19). MR cisternography and postcontrast sequences can be helpful to delineate the facial nerve relative to the schwannomas (25). Demonstration of low signal on T2 within the the labyrinth contents is also important for treatment planning, as it predicts a poor prognosis for hearing following treatment (26). Imaging also plays a role with followup after treatment. While the success rate of gamma-knife therapy for controlling tumor growth is in the 90% range (27), there will be patients whose disease continues to worsen after treatment. In our reported case, the patient was treated with gamma-knife therapy, and has no evidence of tumor growth after 3 years.

VS are among the most common intracranial tumors, and imaging plays a key role in diagnosis, treatment planning, and followup. Classic imaging features of VS allow them to be distinguished from other common CPA masses in many cases. Treatment planning decisions for vestibular schwannomas may therefore hinge on imaging findings together with clinical information, as in the case presented here. VS treated with stereotactic radiosurgery require post-treatment assessment with serial MRI scans, which may show characteristic changes on early post-treatment scans followed by either lack of growth or decreased size in most cases. In cases where CPA mass-lesion imaging findings are less definitive, additional MRI imaging methods including T2*-weighted sequences, MR spectroscopy, and MR perfusion may be helpful to increase diagnostic specificity.

References

1. Moffat DA, Ballagh RH. Rare tumours of the cerebellopontine angle. *Clin Oncol (R Coll Radiol)*. 1995;7(1):28-41. [PubMed]
2. Kartush JM, Brackmann DE. Acoustic neuroma update. *Otolaryngol Clin North Am*. 1996 Jun;29(3):377-92. [PubMed]
3. Lanser MJ, Sussman SA, Frazer K. Epidemiology, pathogenesis, and genetics of acoustic tumors. *Otolaryngol Clin North Am*. 1992 Jun;25(3):499-520. [PubMed]
4. Yoshimoto Y. Systematic review of the natural history of vestibular schwannoma. *J Neurosurg*. 2005 Jul;103(1):59-63. [PubMed]
5. Tos M, Drozdziewicz D, Thomsen J. Medial acoustic neuromas. A new clinical entity. *Arch Otolaryngol Head Neck Surg* 1992;118:127-133. [PubMed]
6. Bonneville F, Savatovsky J, Chiras J. Imaging of cerebellopontine angle lesion: an update. Part 1: enhancing

extra-axial lesions. *Eur Radiol* 2007; 17:2472-2482. [PubMed]
7. Bonneville F, Savatovsky J, Chiras J. Imaging of cerebellopontine angle lesion: an update. Part 2: intra-axial lesions, skull base lesions that may invade the CPA region, and non-enhancing extra-axial lesions. *Eur Radiol* 2007; 17:2908-2920. [PubMed]
8. Casselman JW, Kuhweide R, Ampe W, Meeus L, Steyaert L. Pathology of the membranous labyrinth: comparison of T1- and T2-weighted and gadolinium-enhanced spin-echo and 3DFT-CISS imaging. *AJNR Am J Neuroradiol*. 1993 Jan-Feb;14(1):59-69. [PubMed]
9. Casselman JW, Kuhweide R, Deimling M, Ampe W, Dehaene I, Meeus L. Constructive interference in steady state-3DFT MR imaging of the inner ear and cerebellopontine angle. *AJNR Am J Neuroradiol*. 1993 Jan-Feb;14(1):47-57. [PubMed]
10. Charabi S, Tos M, Thomsen J, Rygaard J, Fundova P, Charabi B. Cystic vestibular schwannoma--clinical and experimental studies. *Acta Otolaryngol Suppl*. 2000;543:11-3. [PubMed]
11. Gomez-Brouchet A, Delisle MB, Cognard C, Bonafe A, Charlet JP, Deguine O, Fraysse B. Vestibular schwannomas: correlations between magnetic resonance imaging and histopathologic appearance. *Otol Neurotol*. 2001 Jan;22(1):79-86. [PubMed]
12. Voss NF, Vrionis FD, Heilman CB, Robertson JH. Meningiomas of the cerebellopontine angle. *Surg Neurol*. 2000 May;53(5):439-46; discussion 446-7. [PubMed]
13. Nassif PS, Shelton C, Arriaga M. Hearing preservation following surgical removal of meningiomas affecting the temporal bone. *Laryngoscope*. 1992 Dec;102(12 Pt 1):1357-62. [PubMed]
14. Thamburaj K, Radhakrishnan VV, Thomas B, Nair S, Menon G. Intratumoral microhemorrhages on T2*-weighted gradient-echo imaging helps differentiate vestibular schwannoma from meningioma. *AJNR Am J Neuroradiol*. 2008 Mar;29(3):552-7. Epub 2007 Dec 13. [PubMed]
15. Yamasaki F, Kurisu K, Satoh K, Arita K, Sugiyama K, Ohtaki M, Takaba J, Tominaga A, Hanaya R, Yoshioka H, Hama S, Ito Y, Kajiwara Y, Yahara K, Saito T, Thohar MA. Apparent diffusion coefficient of human brain tumors at MR imaging. *Radiology*. 2005 Jun;235(3):985-91. Epub 2005 Apr 15. [PubMed]
16. Hakyemez B, Erdogan C, Bolca N, Yildirim N, Gokalp G, Parlak M. Evaluation of different cerebral mass lesions by perfusion-weighted MR imaging. *J Magn Reson Imaging*. 2006 Oct;24(4):817-24. [PubMed]
17. Cho YD, Choi GH, Lee SP, Kim JK. (1)H-MRS metabolic patterns for distinguishing between meningiomas and other brain tumors. *Magn Reson Imaging*. 2003 Jul;21(6):663-72. [PubMed]
18. Telian SA. Management of the small acoustic neuroma: a decision analysis. *Am J Otol*. 1994 May;15(3):358-65. [PubMed]
19. Nikolopoulos TP, O'Donoghue GM. Acoustic neuroma management: an evidence-based medicine approach. *Otol Neurotol*. 2002 Jul;23(4):534-41. [PubMed]
20. Battaglia A, Mastrodimos B, Cueva R. Comparison of growth patterns of acoustic neuromas with and without radiosurgery. *Otol Neurotol*. 2006 Aug;27(5):705-12. [PubMed]
21. Hajioff D, Raut VV, Walsh RM, Bath AP, Bance ML, Guha A, Tator CH, Rutka JA. Conservative management of vestibular schwannomas: third review of a 10-year prospective study. *Clin Otolaryngol*. 2008 Jun;33(3):255-9. [PubMed]
22. Stangerup SE, Caye-Thomasen P, Tos M, Thomsen J. The natural history of vestibular schwannoma. *Otol Neurotol*. 2006 Jun;27(4):547-52. [PubMed]
23. Ozgen B, Oguz B, Dolgun A. Diagnostic accuracy of the constructive interference in steady state sequence alone for follow-up imaging of vestibular schwannomas. *AJNR Am J Neuroradiol*. 2009 May;30(5):985-91. Epub 2009 Feb 4. [PubMed]
24. van de Langenberg R, de Bondt BJ, Nelemans PJ, Baumert BG, Stokroos RJ. Follow-up assessment of vestibular schwannomas: volume quantification versus two-dimensional measurements. *Neuroradiology*. 2009 Aug;51(8):517-24. Epub 2009 May 6. [PubMed]
25. Sartoretti-Schefer S, Kollias S, Valavanis A. Spatial relationship between vestibular schwannoma and facial nerve on three-dimensional T2-weighted fast spin-echo MR images. *AJNR Am J Neuroradiol*. 2000 May;21(5):810-6. [PubMed]
26. Somers T, Casselman J, de Ceulaer G, Govaerts P, Offeciers E. Prognostic value of magnetic resonance imaging findings in hearing preservation surgery for vestibular schwannoma. *Otol Neurotol*. 2001 Jan;22(1):87-94. [PubMed]
27. Kaylie DM, Horgan MJ, Delashaw JB, McMenomey SO. A meta-analysis comparing outcomes of microsurgery and gamma knife radiosurgery. *Laryngoscope*. 2000 Nov;110(11):1850-6. [PubMed]