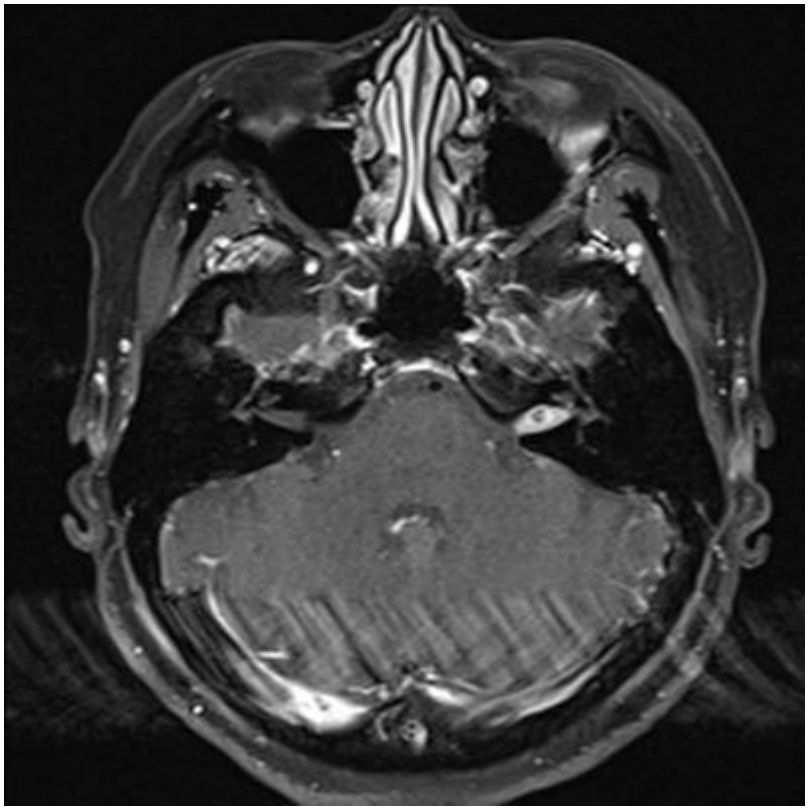
Intracanalicular vestibular schwannoma

The current practitioner is more often managing intracanalicular vestibular schwannomas than in the past, as improved imaging and heightened awareness leads to earlier diagnosis of these tumors.



Case courtesy of Dr Ian Bickle, Radiopaedia.org. From the case rID: 48853

Differential diagnosis

Intracanalicular meningioma

Pantopaque (iophendylate) is an oily contrast medium historically used during spine imaging. Due to its persistence in the subarachnoid space and the potential to lead to severe arachnoiditis, it is no longer used today. Deep et al., present a 40-year-old male with new-onset headaches, imbalance, and vertigo. Brain magnetic resonance imaging revealed a 2-mm T1 -hyperintense intracanalicular lesion. Numerous hyperdense foci were scattered throughout the subarachnoid space on computed tomography. Further history revealed the patient received Pantopaque 30 years prior, after sustaining spinal trauma. Remnant Pantopaque contrast is an important differential when evaluating a patient with a suspected intracranial tumor in order to avoid unwarranted surgical intervention [1)].

A 46-year-old man with venous compression of the vestibulocochlear nerve inside the internal auditory canal (IAC). The patient presented with a 2-year history of recurrent attacks of disabling

vertigo and intermittent high-frequency tinnitus on the right side. Magnetic resonance images showed a small, contrast-enhancing lesion in the fundus of the right IAC, which was suspicious for vestibular schwannoma. During surgical exploration, a large venous loop was found extending into the IAC and compressing the vestibulocochlear nerve. The vessel was mobilized and rerouted out of the IAC. The presumed vestibular schwannoma at the cochlear fossa was left in situ. The patient's symptoms resolved immediately after surgery. Hearing was unchanged postoperatively. On follow-up, there has been no growth of the contrast-enhancing lesion in the IAC for 3 years so far.Disabling vertigo can also be caused by venous microvascular compression of the vestibulocochlear nerve inside the IAC and may be treated successfully by microvascular decompression. A sensitive, conservative approach to lesions in the fundus may be justified in the presence of an additional, more prominent pathology that causes compression of the vestibulocochlear nerve [2)].

Treatment

Intracanalicular Vestibular Schwannoma Treatment.

Case series

In a study, Corrivetti et al. described the surgical treatment of 3 cases of intracanalicular vestibular schwannomas (ICVSs) with an endoscopic assisted retrosigmoid approach (EARSA), highlighting the advantages and limitations of flexible endoscopy in accomplishing a safe radical resection with hearing preservation. Three patients with an ICVS underwent surgery via a flexible endoscopic-assisted microneurosurgical retrosigmoid approach. Flexible endoscopic assistance allowed the identification of residual tumor located in the most lateral portion of the fundus of the internal auditory canal in all cases. Endoscopic controls and further microsurgical resection were attempted, and complete surgical resection was achieved in all cases without the occurrence of postoperative facial or auditory nerve dysfunction. Flexible endoscopy appears to be particularly useful and safe in the surgical management of ICVS by microneurosurgery via an EARSA [3)].

2016

A retrospective study was done in 14 patients who underwent MFA for vestibular schwannoma in Asan Medical Center.

The median age at diagnosis was 46.3 years. At initial presentation, 57% of the patients had vertigo, 43% hearing disturbance, and 64% tinnitus. The mean tumor size was 9.7 mm. The tumors were completely resected in 86% of the patients. Hearing was post-operatively preserved in 12 patients and two patients lost their hearing following surgery. Facial nerve function post-operatively remained unchanged in 12 patients (86%) [4)].

A retrospective analysis of 19 patients with intracanalicular VS and disabling vestibular dysfunction as the main or only symptom (Group A). All of the patients reported having had disabling vertigo attacks. Subjective evaluation of the impairment of patients was performed before surgery, 3 weeks after

surgery, 3 months after surgery, and 1 year after surgery, using the Dizziness Handicap Inventory (DHI). The main outcome measures were improvement in quality of life as measured using the DHI, and general and functional outcomes, in particular facial function and hearing. Patient age, preoperative tumor size, preoperative DHI score, and preservation of the nontumorous vestibular nerve were tested using a multivariate regression analysis to determine factors affecting the postoperative DHI score. The Mann-Whitney U-test was used to compare the postoperative DHI score at 3 weeks, 3 months, and 1 year after surgery with a control group of 19 randomly selected patients with intracanalicular VSs, who presented without vestibular symptoms (Group B). The occurrence of early postoperative discrete vertigo attacks was also compared between groups. RESULTS The preoperative DHI score was ≥ 54 in all patients. All patients reported having had disabling rotational vertigo before surgery. The only significant factor to affect the DHI outcome 3 weeks and 3 months after surgery was the preoperative DHI score. The DHI outcome after 1 year was not affected by the preoperative DHI score. Compared with the control group, the DHI score at 3 weeks and 3 months after surgery was significantly worse. There was no significant difference between the groups after 1 year. Vertigo was improved in all patients and completely resolved after 1 year in 17 patients.

Disabling vestibular dysfunction that affects quality of life should be considered an indication for surgery, even in otherwise asymptomatic patients with intracanalicular VS. Surgical removal of the tumor is safe and very effective in regard to symptom relief. All patients had excellent facial nerve function within 1 year after surgery, with a very good chance of hearing preservation [5].

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156 patients diagnosed with an intracanalicular VS managed conservatively.

After a follow-up of 9.5 years, tumor growth had occurred in 37% and growth into the cerebellopontine angle had occurred in 23% of patients. Conservative treatment failed in 15%. The pure tone average had increased from 51- to 72-dB hearing level, and the speech discrimination score (SDS) had decreased from 60% to 34%. The number of patients with good hearing (SDS > 70%) was reduced from 52% to 22%, and the number of patients with American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS) class A hearing was reduced from 19% to 3%. Hearing was preserved better in patients with 100% SDS at diagnosis than in patients with even a small loss of SDS. Serviceable hearing was preserved in 34% according to AAO-HNS (class A-B) and in 58% according to the word recognition score (class I-II). Rate of hearing loss was higher in patients with growing tumors.

Tumor growth occurred in only a minority of patients diagnosed with an intracanalicular VS during 10 years of observation. The risk of hearing loss is small in patients with normal discrimination at diagnosis. Serviceable hearing is preserved spontaneously in 34% according to AAO-HNS and in 58% according to the word recognition score [6].

2014

31 patients who were followed up for more than 1 year among patients diagnosed as having VS limited to the internal auditory canal. The median follow-up period was 31 months (range, 12-84 mo). We analyzed the patients' clinical features, clinical courses, and audiologic changes.

The most frequent initial presenting symptom in patients with ICVS was hearing loss, and one-half of the patients (8 of 16) had a history of sudden hearing loss. Seven patients (22.5%) showed tumor growth during the follow-up period. When we considered the initial tumor size in ICVS, the patients

larger in size than the median showed a significantly higher rate of tumor growth. In terms of the initial hearing levels of ICVS according to the Consensus Meeting Guidelines, five patients were classified as Class A (normal hearing) and six patients were classified as Class B. Only one patient among patients with useful hearing (Classes A and B) showed tumor growth. The follow-up hearing levels of all Class A patients were preserved; however, all Class B patients deteriorated to Class C.

Patients with ICVS showed favorable results with conservative management. Among them, patients with small tumors and normal hearing showed a good prognosis [7].

2011

47 patients with a unilateral intracanalicular vestibular schwannoma. Evaluation of growth was monitored by repeat MRI scanning. Repeated pure-tone and speech audiometry results were evaluated for subgroups of patients showing growth or no growth and by subsite location of tumor in the internal auditory canal.

Patients had a mean follow-up of 3.6 years. Over the entire population, the pure-tone average thresholds at 0.5, 1, 2, and 3 kHz and the word recognition scores both significantly deteriorated from 38 to 51 dB HL, and from 66% to 55%, respectively. Overall, 74% of subjects with good hearing, according to the 50/50 rule, maintained hearing above this rule. There were no significant differences in hearing loss by subsite in the internal auditory canal (porus, fundus, central) or by growth status (stable, growing, shrinking). Only 6 patients showed a large hearing change. This happened early during follow-up, with relatively stable hearing after this.

Hearing will deteriorate in some intracanalicular vestibular schwannomas, regardless of tumor growth. Hearing deterioration, if on a large scale, most likely occurs early in follow-up. The present results using conservative management in these tumors appear similar to those reported for stereotactic radiotherapy or microsurgery [8].

2008

Forty-seven patients (22 men and 25 women) harboring an intracanalicular vestibular schwannoma were followed prospectively. Mean age at the time of inclusion was 54.4 (20-71) years. The mean follow-up period was 43.8 months (+/-40 months) ranging from 9 to 222 months. Failure was defined as significant tumor growth and/or hearing deterioration that required a microsurgical or radiosurgical treatment. Failure was observed in 35 cases while a conservative treatment is still ongoing in 12 patients. Ten patients kept an unchanged tumor size (21.3%), while 36 patients experienced a tumor growth (76.6%), and 1 patient experienced a mild decreased tumor size (2.1%). Among the 40 patients who where available for hearing level study, 24 patients (60%) did not change their Gardner and Robertson hearing class. Fifteen patients (37.5%) experienced a >10-dB hearing loss and 2 of them became deaf. One patient (2.5%) improved her hearing level from 56.3 to 43.8 dB over a 39.5-month follow-up period. These data suggest that the wait and see policy exposes the patient to degradation of hearing and tumor growth. Both events may occur in an independent way in the middle-term period. This information has to be given to the patient, and a careful sequential follow-up may be adopted when the wait and see strategy is chosen [9].

Between 1987 and 2003, 96 patients (65 men and 31 women) underwent gamma knife stereotactic radiosurgery (SRS) for intracanalicular tumors. The median patient age was 54 years (range, 22-80 years). Hearing was graded using the Gardner-Robertson (GR) and the American Academy of Otolaryngology-Head and Neck Surgery classifications. Dose planning was performed on intraoperative stereotactic images using multiple 4-mm isocenters. The median tumor volume was 0.112 mm3 (range, 0.05-0.447 mm3), and the median margin dose was 13 Gy (range, 10-18 Gy).

The mean and median audiologic follow-up periods were 42 months and 28 months (range, 12-144 months), respectively. Serviceable hearing was preserved in 31 of 40 (77.5%) patients with initial American Academy of Otolaryngology-Head and Neck Surgery Class A hearing. Serviceable hearing was preserved in 40 of 79 (64.5%) patients with GR Grade I or II pre-SRS hearing. Ninety-two patients had GR Grade I, II, or III hearing before SRS, and GR Grade I, II, or III hearing was maintained in 78 patients (85%). Hearing grades improved in 7 patients. Facial and trigeminal nerve function was preserved in all patients. The tumor control rate (freedom from additional intervention) was 99.0% (95 of 96) at a median follow-up of 28 months (range, 12-144 months). One patient underwent tumor resection 18 months after radiosurgery.

SRS is a minimally invasive first-line management option for patients with intracanalicular tumors and provides high rates of hearing preservation with minimal morbidity [10)].

2000

40 patients with 40 unilateral VS in the period 1973 to 1996 (mean 3.6 years). Twenty-seven tumours (67.5%) revealed growth and 13 tumours (32%) had no measurable growth. Four growth patterns were observed: (i) 15 tumours (37.5%) exhibited constant growth; (ii) 13 tumours (32.5%) had no measurable growth; (iii) 8 tumours (20%) revealed growth subsequent to a no-growth period; and (iv) 4 tumours (10%) manifested different growth patterns during the observation period. The mean diameter growth per year was 3.2 mm. The findings of the present study, especially those achieved in groups B (the non-growing tumours) and C (tumour growth subsequent to a silent period), question the reliability of the results achieved by radiosurgery, as no tumour growth may occur with no intervention [11)].

Case reports

2008

A unique case of unilateral widening of the internal auditory canal (IAC) with no significant contact with an ipsilateral intracanalicular vestibular schwannoma (VS), raising the issue of the cause(s) of this IAC widening.

The medical record and radiologic data were reviewed of a patient presenting an enlarged unilateral IAC, which led to the diagnosis of an intracanalicular VS that could not account for the dilation.

The patient had a unilateral dilation of the IAC that did not match the ipsilateral VS he had. As a result, this case motivated discussion of whether such dilation of the IAC was congenitally asymmetrical or the result of the mechanisms involved in the widening of the IAC.

Although asymmetry of IAC is a current notion, this case demonstrates a contrario that increased pressure exerted on the walls of the IAC cannot be the only mechanism in such widening [12].

2004

The first reported case of hemifacial spasm responsive to gamma knife radiosurgery in a patient with an intracanalicular vestibular schwannoma. Both the resolution of the spasm as well as tumor growth control were achieved with a single session of gamma knife radiosurgery. We report a 49-year-old male patient with a 6-month history of right-sided hearing loss and hemifacial spasm. MR examination revealed an intracanalicular vestibular schwannoma. The patient was treated with radiosurgery and received 13 Gy to the 50 % isodose line. Tumor growth control was achieved and no change in the tumor volume was present at the last follow-up at 22 months. The hemifacial spasm completely resolved after one year. Surgical removal of the presumably causative mass lesion has been reported to be the sole treatment in secondary hemifacial spasm. This case report indicates that it may be responsive to gamma knife radiosurgery. Whether or not this might be a treatment option in selected refractory cases of hemifacial spasm remains to be defined [13].

2002

A 68-year-old man with complete deafness of the left ear since childhood, who developed sudden, profound sensorineural hearing loss in the right ear. Magnetic resonance imaging revealed a small right-sided intracanalicular tumor. Treatment with high-dose corticosteroids produced only minimal improvement in hearing. Subsequent emergency decompression and resection of a VS resulted in rapid improvement and restoration of hearing, with facial nerve preservation. Although most neurotologic lesions in patients with hearing in only one ear are managed nonsurgically, resection of small tumors in the setting of sudden hearing loss should be considered in selected cases. This finding indicates that a therapeutic window may exist during which sudden hearing loss caused by intracanalicular tumors is reversible [14].

1998

An unusual case in which they recognized an additional branch arising from the jugular bulb. Three-dimensional computed tomography (3-D CT) revealed this anomaly beforehand, enabling us to avert excessive bleeding upon resection of the tumour. The abnormal vein was thought to be a remnant of the petrosquamosal sinus in the embryonic stage [15].

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