

TL Kwan 關鼎樂
KW Tang 鄧國穎
KKT Pak 白錦姿
JYL Cheung 張毓靈

Screening for vestibular schwannoma by magnetic resonance imaging: analysis of 1821 patients

以磁力共振成像進行前庭神經鞘瘤普查：對1821名病人的分析

Objectives. To study the spectrum of diseases that can be detected by magnetic resonance imaging in patients suspected to have vestibular schwannoma (acoustic neuroma) presenting with sensorineural or mixed hearing loss, and to assess the extent of the problem of hearing loss in a screened population.

Design. Retrospective study.

Setting. Diagnostic radiology and imaging department of a regional hospital, Hong Kong.

Patients. A total of 1821 consecutive patients from September 1999 to February 2001 with sensorineural or mixed hearing loss were referred by otolaryngologists for magnetic resonance imaging of the internal auditory canal.

Main outcome measures. Vestibular schwannoma; other cerebellopontine angle masses and other diseases that could account for the patients' hearing loss.

Results. In all, 132 (7%) patients had positive findings that could explain their hearing loss. Fifty-four (41%) of the 132 patients had vestibular schwannoma; 39 (30%) had inflammation of the middle ear and mastoids; 17 (13%) had ischaemic foci in the brainstem; 10 (8%) had other cerebellopontine angle masses or tumours; four (3%) had inner ear dysplasia; seven (5%) had vascular loop compression; and one (1%) had chronic cryptococcal meningitis. The overall incidence of vestibular schwannoma detected in this screened population was about 3%.

Conclusions. This study indicates that magnetic resonance imaging is an effective tool to screen for vestibular schwannoma in patients with sensorineural or mixed hearing loss. It can also be used to assess a considerable number of different pathological conditions in patients with audiovestibular disorders.

目的：研究以磁力共振成像，為懷疑患有前庭神經鞘瘤（聽神經瘤）而病徵為感覺神經或混合性聽覺損失的病人可能測出的病譜；並評估聽覺損失問題在接受普查的人口中的嚴重程度。

設計：回顧性研究。

安排：地區醫院的診斷性放射及影像科，香港。

患者：1999年9月至2001年2月期間，連續1821名患有感覺神經或混合性聽覺損失的病人；他們均由耳鼻喉科專家轉介進行內耳道磁力共振成像。

主要結果測量：前庭神經鞘瘤、其他小腦橋腦角塊質，以及其他能解釋病人聽覺損失的疾病。

結果：共132名（7%）病人有可解釋其聽覺損失的發現。54人（41%）有前庭神經鞘瘤；39人（30%）的中耳或乳突發炎；17人（13%）的腦幹內有局部缺血灶；10人（8%）有其他小腦橋腦角物質或腫瘤；4人（3%）內耳發育不良；7人（5%）血管袢受壓；1人（1%）有慢性隱球菌腦膜炎。普查人口中測出前庭神經鞘瘤的總發病率約為3%。

結論：研究顯示，磁力共振成像是為患有感覺神經或混合性聽覺損失病人測出前庭神經鞘瘤的有用工具。它亦能用於為聽覺及前庭失調的病人評估相當多的各種病理學情況。

Key words:

Hearing loss;

Magnetic resonance imaging;

Neuroma, acoustic

關鍵詞：

聽覺損失；

磁力共振成像；

神經瘤，聽覺

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Department of Radiology and Imaging,
Queen Elizabeth Hospital, 30 Gascoigne
Road, Kowloon, Hong Kong

TL Kwan, FRCR, FHKAM (Radiology)

KW Tang, FRCR, FHKAM (Radiology)

KKT Pak, FRCR, FHKAM (Radiology)

JYL Cheung, FRCR, FHKAM (Radiology)

Correspondence to: Dr TL Kwan
(e-mail: tlokkwan@hotmail.com)

Introduction

Although tumours of the internal auditory canal (IAC) and cerebellopontine angle (CPA)—for example, vestibular schwannoma (VS)—are rare, physicians specialising in otolaryngology commonly consider them when making diagnoses. Patients with VS can present with a wide range of symptoms, such as tinnitus, progressive hearing loss, sudden hearing loss, fluctuating deafness, and dizziness. Because some of these symptoms have potentially treatable causes, it is difficult to diagnose VS. Thus, methods for screening VS have been developed.

Auditory brainstem response (ABR) testing was first described by Selters and Brackmann¹ in 1977 for the detection of VS. By using this method, subsequent studies reported VS detection rates ranging from 93% to 98%.²⁻⁴ Nevertheless, studies have also shown that smaller VS tumours are less likely to be detected by ABR than larger tumours: false-negative rates are as high as 22% for tumours with an extrameatal size of less than 15 mm,⁵ and the sensitivity of ABR for tumours of 1 cm or less is 58%.⁶ Therefore, ABR is not a satisfactory screening tool for the diagnosis of small VS tumours.

Magnetic resonance imaging (MRI) is the current imaging modality of choice in diagnosing VS. Contrast-enhanced T1-weighted spin-echo MRI is the accepted criterion standard for the evaluation of VS. To improve the cost-effectiveness in MRI screening of VS, different non-contrast MRI techniques are being assessed. For example, high-resolution fast spin-echo T2-weighted MRI of the IAC-CPA has equal sensitivity to conventional contrast-enhanced T1-weighted MRI.⁷ A study by Stuckey et al⁸ of submillimetre-resolution MRI using T2*-weighted three-dimensional Fourier transformation-constructive interference in steady state (3DFT-CISS) on a 1.5-T system has 94% to 100% sensitivity and 94% to 98% specificity in the detection of VS. More recently, Held et al⁹ showed that 3DFT-CISS is a very sensitive method for screening patients for VS, because it can detect even small meatal and labyrinthine tumours. Held et al¹⁰ have recommended inclusion of 3DFT-CISS in an MRI protocol of the facial and vestibulocochlear nerves; they found that T2*-weighted 3DFT-CISS was significantly better than T2-weighted 3-D turbo spin-echo for nerve detectability in the CPA, and equally as good as 3-D turbo spin-echo in the IAC.¹⁰

Since 1999, the Queen Elizabeth Hospital has been performing MRI to screen for VS, using T2*-weighted 3DFT-CISS. In this article, we present data on a screened population from September 1999 to February 2001, focusing on the diseases detected that could have accounted for patients' hearing loss.

Methods

In this retrospective study, we assessed the radiology

Table 1. Results of positive findings accounting for hearing loss, using magnetic resonance imaging as the screening method

Imaging findings	No. of cases
Vestibular schwannoma	54
Middle ear and mastoid inflammation	
No history of NPC*	34
History of NPC	4
Newly diagnosed NPC	1
Ischaemic foci in brainstem	16
Cavernous haemangioma in pons	1
Other cerebellopontine angle masses	
Meningioma	3
Trigeminal neuroma	3
Arachnoid cyst	2
Epidermoid cyst	1
Ninth-to-11th nerve schwannoma complex	1
Inner ear dysplasia	4
Vascular loop with compression	7
Chronic cryptococcal meningitis	1

* NPC nasopharyngeal carcinoma

records of a total of 1821 patients who were screened MRI examinations for suspected VS between September 1999 and February 2001. All patients had been referred to the Queen Elizabeth Hospital by otolaryngologists and all had presented with either sensorineural or mixed hearing loss as one of their major symptoms. They had undergone the same MRI protocol conducted on a 1.0-T MRI system (Siemens, Erlange, Germany). The protocol used was a 5-mm thick transverse T1-weighted spin-echo for the brain: 560, 15, 2, and 4 min 14 sec (repetition time, effective echo time, excitations, and acquisition time); a 5-mm thick transverse T2-weighted turbo spin-echo for the brain: 4000, 90, 1, 3 min; and a 1.2-mm thick transverse T2*-weighted 3DFT-CISS for the IAC-CPA: 16.7, 8, 1, 4 min 6 sec. Some patients had also undergone 4-mm thick coronal imaging using T2-weighted turbo spin-echo for the brain (4500, 120, 1, 1 min 7 sec) if the on-site radiographer noted any abnormality. Some patients had undergone contrast studies after staff radiologists reviewed the images.

A neuroradiologist or a head and neck radiologist, who was blinded to the radiology reports, re-examined CISS images of patients and made retrospective diagnoses of VS on the basis of the characteristic appearance and location of the tumours. When the tumour was small, it presented with characteristic T2 hypo-intensity that stood out vividly in the bright background of cerebrospinal fluid in the CISS images. Cavernous haemangioma or facial nerve neuroma was excluded as a differential diagnosis if the patient did not have facial nerve palsy.¹¹ In excluding the presence of VS, the entire course and individual component of the seventh to eighth nerve complex should have been clearly visible without interruption. The clinical management of patients with VS was also assessed.

From the radiology reports, we also encountered patients with findings other than VS that could explain their symptoms. These cases were also reviewed by a neuro-

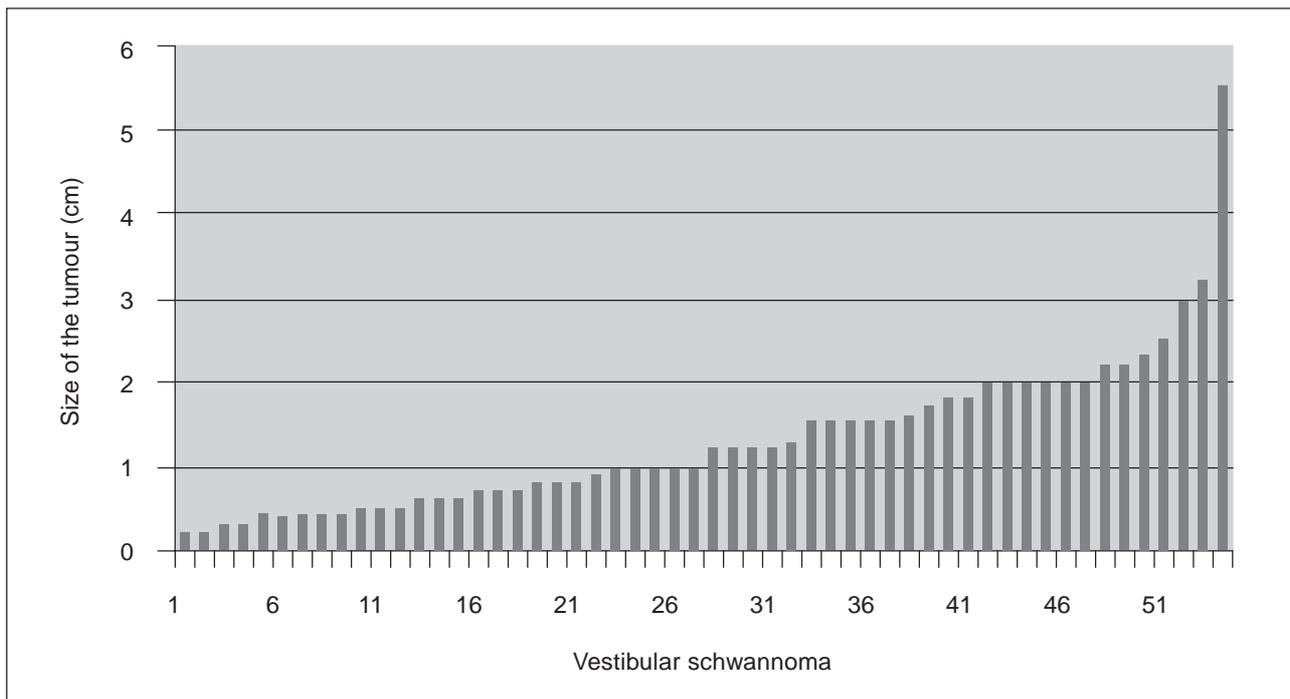


Fig 1. Graphical representation of the size of all 54 vestibular schwannomas

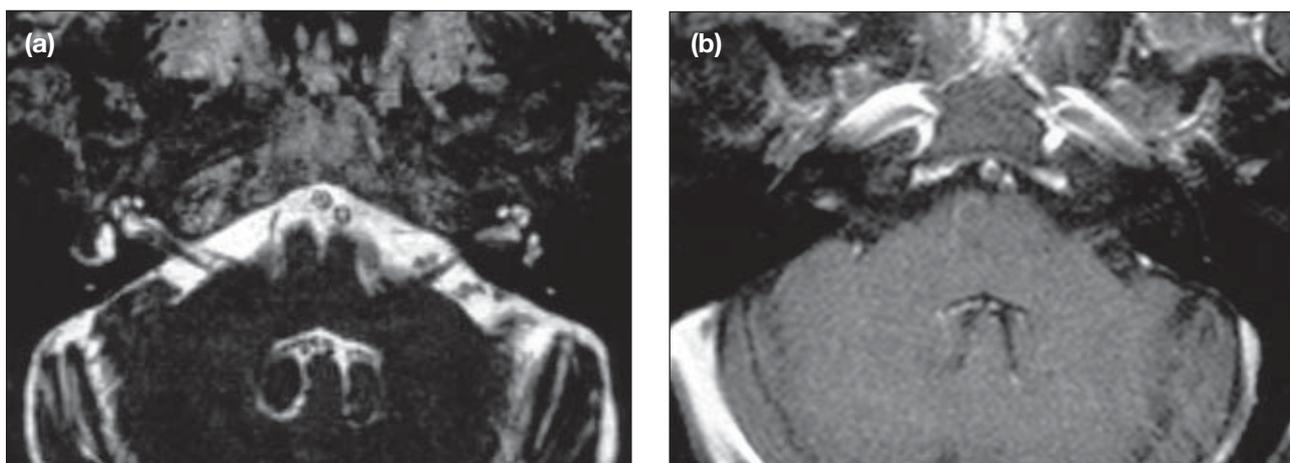


Fig 2. Right intracanalicular vestibular schwannoma

(a) Constructive interference in steady state image showing a small hypo-intense mass along the intracanalicular course of the right vestibular nerve; (b) contrast T1-weighted image showing enhancement of the small right intracanalicular tumour

radiologist or head and neck radiologist, and the confirmed cases were included in our results. Other findings that could not possibly explain the hearing loss were not included.

Results

A total of 132 patients had positive findings that could explain their hearing loss, making an overall positive rate of 7% (Table 1). Among the patients, 54 had VS, 39 had middle ear and mastoid inflammation (five of whom previously or currently had nasopharyngeal carcinoma), and 17 had ischaemic foci in the brainstem (one of whom was suspected to have cavernous haemangioma in the pons). Ten patients had other CPA masses and four had inner

ear dysplasia (one with bilateral enlarged vestibular aqueducts). A further seven patients had prominent basilar or vertebral arteries impinging on the root exit zone of cranial nerve VIII. One patient was found to have hydrocephalus and nodular enhancement of the meninges, later proven to be chronic cryptococcal meningitis.

There was no discrepancy in the number of cases of VS detected originally and the number detected in our retrospective review. The overall incidence of VS detected in the screened population was 3%. The size of tumour along its greatest dimension ranged from 0.2 cm to 5.5 cm (Fig 1). Tumours either were purely intracanalicular in location (Fig 2) or had an extracanalicular component with an ice-

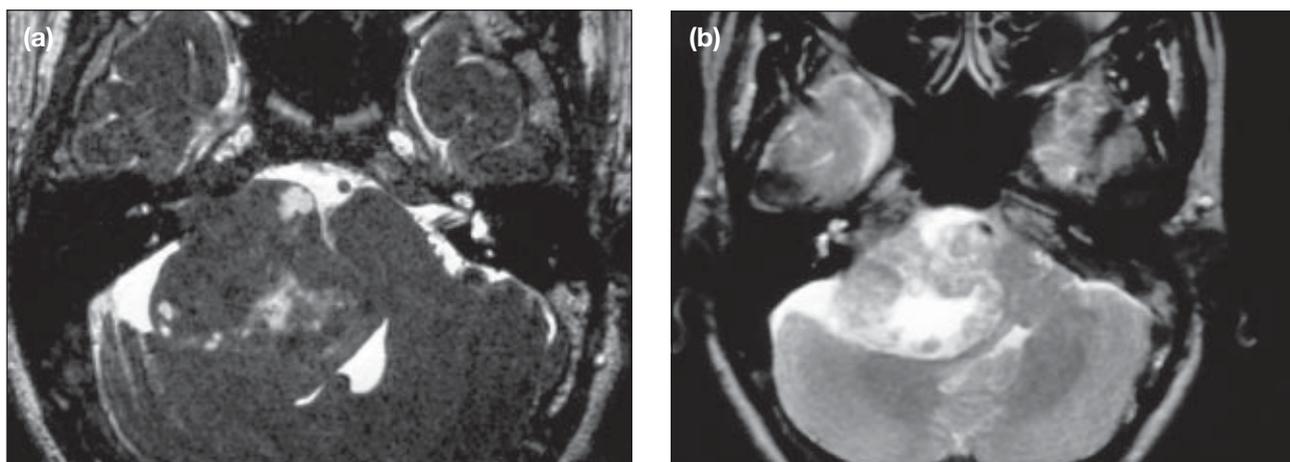


Fig 3. Right vestibular schwannoma with a large extracanalicular component

(a) Constructive interference in steady state image showing a large right cerebellopontine angle tumour with extension into the right internal auditory canal and compression on the brainstem by the tumour; (b) T2-weighted image showing the characteristic ice-cream cone appearance of the vestibular schwannoma with a large extracanalicular component, and high-signal areas suggestive of cystic areas of the tumour

cream cone appearance (Fig 3). As shown in Table 2, the larger the size of the tumour (>1.0 cm), the more likely it had such an extracanalicular extension. Twenty-two tumours were smaller than 1.0 cm, and only one (0.8 cm) had an extracanalicular component; 15 were 1.0 cm to 1.5 cm in size, 12 of which had an extracanalicular component. All 17 tumours that were larger than 1.5 cm had both intracanalicular and extracanalicular components. Only five tumours were surgically removed: two measured 2.0 cm, one 2.5 cm, one 3.0 cm, and one 5.5 cm; these tumours had extracanalicular components that indented or compressed the brainstem. Twenty-seven patients received gamma-knife radiosurgery; their tumours ranged from 0.7 cm to 3.0 cm. Sixteen cases of VS, most of which were smaller than 1.0 cm, were managed conservatively with regular follow-up. Two patients in this group had large tumours (1.7 cm and 2 cm) because they did not consent to treatment. A further six patients were lost to follow-up after diagnosis, and all had small intracanalicular tumours of less than 1.0 cm in size.

In the initial diagnoses of VS, 45 patients were called back for contrast studies, all of which showed characteristic contrast enhancement. Of the nine patients who did not have contrast studies performed, most had tumours larger than 1.0 cm. These nine patients except one attended for contrast MRI studies before gamma-knife radiosurgery or conservative treatment showed contrast enhancement of their tumours. The one exception was a patient with 0.6-cm VS that was detected by CISS, who refused to return for contrast study and defaulted from follow-up.

Table 2. Size and location of vestibular schwannomas

Size of tumour (cm)	Intracanalicular location only	With extracanalicular portion
<1.0	21	1
1.0-1.5	3	12
>1.5	0	17

Discussion

With the advance of MRI techniques and resolution capabilities, there is no doubt that MRI should replace ABR as the screening method for patients who are suspected to have VS. Compared with ABR, MRI can detect smaller tumours, and a specific diagnosis can usually be established from the lesion's location, morphology, signal characteristics, and enhancement pattern. Furthermore, diseases other than tumours can also be detected by MRI.

We identified 54 cases of VS, which accounted for 41% of positive findings. Our study showed that small tumours tend to be intracanalicular in location with no extracanalicular extension. The smallest size of tumour with an extracanalicular component was 0.8 cm. Twenty-two tumours were smaller than 1.0 cm; half of these would not have been undetected with ABR as the screening method, according to Schmidt et al.⁶

As we have many referrals from otolaryngologists for VS screening (about 25 patients a week), we have to perform the screening after hours, including on Sundays. A protocol that can adequately detect VS without the need for contrast studies will improve the cost-effectiveness and eliminate the need for the radiologist to stay behind for the examination. In our department, T2*-weighted 3DFT-CISS was used. Although we could not achieve a satisfactory signal in a submillimetre 3DFT-CISS study using a 1.0-T MRI system, images that were 1.2-mm thick were sufficient to render individual nerves identifiable. All cases of 54 VS (one was not confirmed by contrast study) were detected by this method. We presumed that the remaining patients did not have VS, as reviewed by a qualified neuro-radiologist or head and neck radiologist. Call back for contrast study just to confirm the diagnosis was unnecessary; this would have increased the burden on the radiographer. Furthermore, these patients would have had contrast

MRI studies performed (in the presence of a radiologist) prior to any intervention or follow-up. However, we did not discourage radiologists from calling back patients for contrast studies if they felt that a diagnosis of VS warranted a contrast study. Contrast studies were also done if other diagnoses such as meningioma were possible.

As for the management of VS, there is still no evidence-based approach, and different groups supporting the use of surgery, radiosurgery, and radiological surveillance are of low quality evidence (type III and IV evidence).¹² In our series, five cases of VS were managed surgically, 27 by gamma-knife radiosurgery, and 16 by conservative management. The surgically removed tumours were relatively large in size and compressed the brainstem (Fig 3). No operation was done for tumours smaller than 2.0 cm, although the general belief is that a small tumour in a patient who has good residual hearing has a fair chance of being removed while preserving hearing.¹³ These tumours were treated conservatively or by gamma-knife radiosurgery, thus reflecting the trend towards non-surgical approaches among our surgeons. A detailed discussion on the uses of gamma-knife radiosurgery or conservative management for VS is outside the scope of this study. Nevertheless, cases treated by these methods will require frequent neuroimaging follow-up, which we expect would further overwhelming the MRI system.^{14,15}

The disadvantage of performing an MRI study is the high cost. By using 3DFT-CISS in our protocol, we eliminated unnecessary contrast-enhanced studies, thus improving the cost-effectiveness of the screening test. Our protocol, however, was not able to demonstrate inflammatory (enhancing) lesions. Because hearing loss from inflammation of the inner ear or vestibulocochlear nerve (most commonly viral in origin) usually occurs acutely, the diagnosis and treatment of this condition does not depend on MRI findings. Documenting the cause of hearing loss by demonstrating enhancement of the inner ear or cochlear nerve is reassuring to the patient and physician, but it is of questionable significance and cost-effectiveness when it does not effectively alter the course of treatment. Furthermore, the average waiting time of the MRI screening in our hospital was approximately 1 to 2 months. By the time the imaging was performed, any inflammatory enhancement would have subsided, making the use of contrast agent unnecessary.

Using T2-weighted turbo spin-echo MRI, 39 (30%) of the 132 patients had high-signal intensity material in the middle ear and mastoids on the same side of hearing loss. We included these patients in our study because they were reported as having mixed hearing loss; the presence of inflammatory change in the middle ear and mastoid might have been a contributory cause, at least to the conductive component of hearing loss. Because nasopharyngeal carcinoma is common in our locality, we should look out for this cause of mastoiditis. In fact, four of the patients in

our series had a history of nasopharyngeal carcinoma that was treated with radiotherapy, and one patient was a new case of nasopharyngeal carcinoma. Furthermore, radiation damage to the inner ear is a recognised cause of sensorineural hearing loss, although we may not be able to demonstrate it.¹⁶

Using T2-weighted turbo spin-echo MRI for the entire brain also allowed us to detect in 17 (13%) of 132 patients hyperintense foci in the brainstem that were thought to be cases of vascular ischaemia. Three patients had the lesions at the cochlear nuclei in the inferior cerebellar peduncle on the side of the hearing loss. One patient was suspected to have cavernous haemangioma in the pons. The other patients had lesions at the pontine tegmentum—the site of major decussation of the acoustic pathway. There were also some patients with ischaemic infarcts in the cerebral cortex and other areas of the brainstem, but who were not included in the study because the infarcts might not have accounted for these patients' hearing loss.

Ten other CPA masses were identified. The term CPA refers to the space-bounded anteromedially by the lower midbrain, pons and medulla; posteromedially by the cerebellum; laterally by the posterior aspect of the petrous temporal bone; and superiorly by the tentorium. The cranial nerves within the CPA passing from cephalad to caudad are the trigeminal nerve; facial and vestibulocochlear nerve; and glossopharyngeal, vagus, and accessory nerves. The 10 CPA masses other than VS included three meningiomas, three trigeminal neuromas, two arachnoid cysts, one schwannoma of the ninth to 11th nerve complex, and one epidermoid cyst. Thus, in our study the incidence of CPA masses were 84% VS, 5% trigeminal neuroma, 5% meningioma, 5% arachnoid and epidermoid cyst, and 2% ninth-to-11th nerve complex neuroma. The results, however, may not reflect the true incidence of the CPA masses. For example, meningiomas, arachnoid cysts, and epidermoids are listed in other studies as comprising 10% to 15%, 5% to 10%, and 5% to 9% of CPA masses, respectively.^{17,18} One reason for the relatively low incidence of these non-acoustic tumours detected in our study is that the patients in our series more commonly presented to neurologists rather than to otolaryngologists, because of symptoms other than sensorineural hearing loss.

Seven patients had vertebrobasilar artery dolicho-ectasia with compression onto the root entry zone of the seventh-to-eighth nerve complex, which was considered a possible explanation for their hearing loss in the absence of other positive findings.¹⁹ These patients usually had symptoms of tinnitus as well.

Patients with inner-ear dysplasia were younger in age. Four patients—three with Mondini deformity and one with a bilateral large vestibular aqueduct—were aged 8, 13, 49, and 50 years, respectively. The two older patients actually gave a long history (more than 20 years) of hearing loss.

The patient with cryptococcal meningitis presented with left sensorineural hearing loss. Owing to abnormal dilation of the ventricles and to periventricular oedema in the temporal lobe, contrast injection was given to this patient. Multiple abnormal areas of nodular parenchymal and meningeal enhancement suggested infective meningo-encephalitis or metastatic deposits. Analysis of the biochemistry of the cerebrospinal fluid of this patient and a high blood titre of cryptococcal antigen confirmed the diagnosis of cryptococcal meningitis.

Conclusions

Hearing loss is a major health hazard encountered in the practice of otolaryngology. We presented our techniques and results of MRI screening for VS. The overall incidence of VS in our screened population was about 3%. We conclude that MRI provides an accurate and non-invasive method to evaluate patients who have hearing loss and who are suspected of having VS. Our protocol included a high resolution T2*-weighted sequence for imaging of the IAC-CPA. The 3DFT-CISS protocol served this purpose well. The whole brain was also assessed to look for causes other than VS. In our study, these other causes accounted for about 59% of positive findings, which included inflammation of the middle ear and mastoids, ischaemic foci in the brainstem, other CPA masses, inner ear dysplasia, vascular compression onto exit root of the eighth nerve, and chronic meningitis. The short scanning time and non-contrast technique used in our protocol improved the cost-effectiveness of MRI as a screening tool. Although non-contrast MRI cannot demonstrate enhancement consistent with inflammation, this information does not alter patients' treatment.

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