Clinical and radiological analysis of cerebellopontine angle pathology: a retrospective study

Rhiannon Oliver, MBBS; Shadaba Ahmed, FRCS

Rhiannon Oliver was a medical student at Lancaster University and is now a foundation doctor at Cumberland Infirmary, Carlisle.

Pathology arising in the cerebellopontine angle (CPA) presents in a number of different ways. In this retrospective study, the presenting symptoms and audiometric tests of patients with lesions in the CPA will be analysed to look for features that could be included in a screening proforma to identify patients at risk and prompt further investigation with magnetic resonance imaging (MRI).

The CPAs are located on either side of the cerebellum, between the cerebellar hemispheres and brain stem. The lateral wall of each CPA consists of the temporal bone, and the tentorium forms the roof. Cerebrospinal fluid fills the space. The CPA contains several important neurovascular structures, including cranial nerves V to XII and the anterior inferior cerebellar artery (a branch of the basilar artery).

The commonest lesion found in the CPA is vestibular schwannoma (VS), also known as acoustic neuroma – a benign, slow-growing tumour. Other pathology found in this region includes meningioma, epidermoid cyst, cholesterol granuloma, arachnoid cyst and posterior cerebellar artery aneurysm.

MRI scans of the pathology encountered in this study are shown in figures 1 to 3.

Lesions in the CPA give rise to a range of symptoms. High-frequency hearing loss commonly occurs in VS. Table 1 describes the effects of compression of each cranial nerve in more detail. Headache, nausea and vomiting, loss of balance and ataxia are also reported.⁽¹⁾ MRI is the primary investigation used in the diagnosis of patients presenting with audiovestibular symptoms.⁽²⁾

Cranial nerve	Name	Features of compression
V	Trigeminal	Trigeminal neuralgia
VI	Abducens	Double vision on side gaze
VII	Facial	Facial palsy
VIII	Vestibulocochlear	Deafness
IX	Glossopharyngeal	Laryngeal incompetence: difficulty swallowing
X	Vagus	Difficulty swallowing, weak voice
XI	Accessory	Shoulder drop
XII	Hypoglossal	Tongue paralysis

Table 1 Cranial nerves in the CPA and symptoms associated with local pathology

Current guidelines recommend that patients presenting with unexplained or asymmetrical auditory symptoms comprising hearing loss or tinnitus should be investigated for the presence of VS. Impaired facial sensation and imbalance also warrant further investigation.⁽³⁾

Several studies have endeavoured to describe clinical features that suggest the presence of masses in the CPA.^(2,4-8) Hearing loss and tinnitus are the commonest presenting symptoms of CPA lesions,^(2,6) but there is no combination of symptoms that can be used to make a formal diagnosis.

A study in 1997 looked at 25 patients with unilateral meningioma in the CPA and found that hearing loss was the commonest presenting symptom (affecting 84% of study participants), followed by imbalance in 76%.⁽⁴⁾ Similarly, all the CPA meningioma patients in a 2009 study presented with dizziness, but tinnitus and hearing loss were less frequent, both reported by 60% of the study sample.⁽⁵⁾ These studies suggest that dizziness and imbalance occur more commonly in meningiomas than in VS; in which the incidence of dizziness varies from 8.3% to 18%.^(2,6,9)

MRI is a limited and costly resource, so a number of researchers have defined criteria that should be met before

scanning patients.⁽⁹⁻¹⁴⁾ Many of these protocols are highly sensitive but not very specific, therefore large numbers of normal patients are scanned unnecessarily. For example, the Charing Cross Protocol had a sensitivity of 97%, but a specificity of only 49%.⁽¹²⁾

Most of the protocols require a combination of specific hearing thresholds on audiometry and presenting symptoms such as tinnitus or vertigo.^(9,10,12,13) One of the best screening protocols had a sensitivity of >90% and a low screening rate of 24%.⁽⁹⁾ It recommended screening patients with either (i) ≥ 20 dB asymmetry at two neighbouring frequencies, or unilateral tinnitus, or (ii) ≥ 15 dB asymmetry at two frequencies from 2-8kHz.

Of the currently published series, several had small sample sizes. The majority of the studies were retrospective analyses, which may have measurement bias if a protocol for recording data was not used at the time patients presented. It is also likely that the studies using data spanning a long time period, such as the 2010 study analysing patients diagnosed between 1973 and 2008,⁽⁹⁾ do not account for changes in clinical practice.

METHODS

A retrospective analysis of the presenting symptoms and audiograms in patients with CPA pathology was undertaken. Suitable patients were found by examining all scans requested by one ENT consultant in the period May 2007 to June 2010.

Two factors were investigated in this study: the presenting symptoms; and the patient's audiogram. Information about how the patients presented was gathered from clinic letters obtained from the hospital records, and audiograms for each patient were also obtained; this information was recorded on a spread sheet, with hearing levels for frequencies of 250, 500, 1,000, 2,000, 4,000 and 8,000Hz recorded in decibels. The level of hearing impairment for each ear was recorded as normal, mild, moderate, severe or profound using the parameters defined by the Royal National Institute for Deaf People

An MRI scan of the internal auditory meati is indicated in:

- Unilateral hearing loss
- Difference in bone conduction threshold of >15dB in two adjacent frequencies

Or

- Bilateral hearing loss (better ear has mean threshold ≥ 30 dB)
- Difference in bone conduction threshold of >20dB in two adjacent frequencies

Or

- Unilateral tinnitus
- Only if >10dB difference in 50% speech discrimination score

Or

- Non VIII nerve symptoms/signs

Figure 4 The Charing Cross Protocol⁽¹²⁾

Level of hearing impairment	Threshold (decibels, dB)
Normal	0-20
Mild	25-39
Moderate	40-69
Severe	70-94
Profound	≥95

Figure 5 Classification of level of hearing impairment from the RNID⁽¹⁵⁾

(RNID),⁽¹⁵⁾ (see figure 5). For the purpose of this study, patients with hearing thresholds of 21-24dB were included in the 'normal' category. Right and left ears were compared to determine whether there was a difference between the side with pathology and the normal side.

Literature Review

Web of Science, The Cochrane Library and MEDLINE (Ovid) were searched using the keywords 'cerebellopontine' and 'pathology' for reviews and research articles in the English language published since 1999, which yielded 29 results. Only a handful of the articles were relevant so the search was modified to 'cerebellopontine' and 'screening'. Finally, studies that focused on the commonest pathology, VS, were sought using the search terms 'acoustic neuroma' OR 'vestibular schwannoma' and 'screening'.

RESULTS

A total of 484 MRI head/IAM scans were found and searched. Forty-four patients were found to have abnormal findings and were included in the study. Fifteen of the 44 were found to have vascular loops and were included in the analysis.

In the study population, there were four different pathologies found in the CPA on MRI; the commonest was VS (27 patients), followed by vascular loops (15 patients), one meningioma and an epidermoid cyst. Letters were not available for eight of the patients, but three of them had undergone VS excision, as reported in their surveillance MRI. Pure-tone audiograms were available for 33 of the patients.

Presenting symptoms and pathology

There were a variety of presenting symptoms in patients, which included hearing loss (bilateral/unilateral), tinnitus, balance problems, vertigo, a sense of ear fullness and facial numbness (see figure 6). Hearing loss was the commonest complaint, reported by 33 patients (89.2%), with approximately

half (53.1%) of them reporting bilateral hearing loss and 48.6% reporting unilateral hearing loss. Half of the 18 patients with bilateral hearing loss reported asymmetrical hearing loss. Over half of the patients (21, 56.8%) experienced tinnitus. Balance problems were less common, affecting only seven patients (18.9%) and vertigo was reported by five patients (13.5%).

Pure-tone audiometry

Audiograms were available for 33 out of 44 patients in the study sample. The level of hearing was calculated by taking the average amplitude for 500, 1,000 and 2,000Hz. Two of the patients had bilateral pathology and the remaining 31 had unilateral pathology reported. In the patients with unilateral pathology, the average threshold for the affected side was 46.6Hz, compared to 29.1Hz in the unaffected ear. This is concordant with previous studies and guidance that suggest that asymmetric auditory symptoms should raise suspicion of a CPA lesion and prompt an MRI scan.^(3,9,12,13) Only two of the 31 patients had not reported any symptoms of hearing loss, but both their audiograms showed asymmetry; one patient with a VS had a mild hearing impairment in the affected ear and the second had a vascular loop but worse hearing in the opposite ear.

The data was analysed to see if there were any differences in presentation between each condition.

Vestibular schwannoma

All but one of the VS patients presented with hearing loss (eight unilateral, 12 bilateral) and approximately half had tinnitus. Balance problems, vertigo and ear pain were much less frequent, occurring in less than a fifth of the patients (see figure 7).

Audiograms were available for 20 of the VS patients. Nineteen had unilateral VS, with an average hearing threshold of 55.5dB in the VS ear compared to 27.2dB in the unaffected ear. When classified with the RNID definitions of deafness, 15 of the patients (80%) had worse hearing in the VS ear; two had equal hearing levels and two had worse hearing in the non-VS ear. In the 15 patients with worse hearing in the VS ear, the difference in hearing thresholds varied from 10 to 90dB (mean 30.3dB). Figure 8 shows the audiometric values for each ear for the patients with right-sided and left-sided VS. The audiograms vary between patients and there is no clear trend to show correlation between the audiometry results and the presence of VS. The high frequency loss reported in the literature as characteristic of VS was evident in all of the patients.⁽¹⁹⁾

Figure 9 compares the audiometric findings for the VS ear with its size (including the patient with bilateral VS). Eighteen of the

20 VS had the size of the tumour reported on the MRI scan, which ranged from 4 to 18mm (maximum length), with an average of 10.3mm. There is positive correlation between severity of hearing loss and size, but it is a weak one. Two of the largest tumours (maximum lengths 17 and 18mm) in the sample had a mild hearing impairment, whereas one of the smallest tumours measuring 4mm had a moderate hearing impairment.

The patient with bilateral VS had a much larger tumour on the left than on the right (10 × 18mm compared with 3 × 4mm). The hearing threshold was more severely affected on the left

compared to the right (25 versus 43.3dB), which supports the correlation between level of hearing impairment and VS size.

Vascular loops

Figure 10 illustrates the frequency of symptoms reported by patients with vascular loops. Tinnitus and hearing loss were equally common symptoms. Two of the 14 patients had bilateral loops noted on MRI. One of these patients presented with bilateral hearing loss but the other presented with right-sided tinnitus.

Eleven patients had audiograms available. The average hearing threshold for the patients with vascular loops was slightly higher for the affected ear compared to the unaffected ear (34.6 versus 31.9). However, three patients' audiograms showed worse hearing in the ear without a vascular loop on MRI. The differences between affected and unaffected ears were variable, ranging from a small difference of 3.3dB to a larger discrepancy of 28.3dB (average 14.4dB) for the eight patients with worse hearing in the ear with a vascular loop.

Meningioma

The patient with a right meningioma presented with right-ear pain, hearing loss and vertigo. The audiometry findings showed asymmetrical hearing loss, but this was in fact worse in the left ear, suggesting that the meningioma was not causing compression of the right acoustic nerve.

Epidermoid cyst

This patient's only complaint was bilateral hearing loss, which was reflected in the audiogram. The epidermoid cyst was right-sided and the average hearing threshold in that ear was 56.7dB compared to 41.7dB in the left ear. In this case the degree of hearing loss was greater in the affected ear.

DISCUSSIONS

According to the literature, approximately 75% of masses in the CPA are VS, and 10-15% are meningiomas.^(16,5) In this study, the incidence of VS was more common, comprising 93.1% of the masses (27 out of 29) and meningiomas were less frequent, constituting only 3.4%.

The data suggests a higher pick-up rate for CPA pathology. It has to be borne in mind that this data was collected over three years and a few patients would have undergone a repeat annual surveillance scan in that period. Also the histology of the tumours that were surgically removed is not available to be included this study. There is a valid point to be made regarding the number of scans undertaken and hence the obvious higher numbers of small VS that were detected. However, there does, despite the above, remain a marginally higher incidence of VS in our catchment area. The incidence has been commented on by our regional Skull Base MDT Team as well. It would be of interest to further analyse the demographics of the patients diagnosed with VS in this review.

“ Our data suggests our local incidence of VS is greater than other reported incidences ”

Our data suggests our local incidence of VS is greater than other reported incidences. We are unsure why this is the case, but it may be because we have included longterm follow-up patients in our data. It is a question, however, which demands an answer with more detailed epidemiological data.

Hearing loss and tinnitus were the two commonest presenting. Other symptoms such as balance problems and vertigo were less common.

The majority of patients with VS in this study presented with hearing loss, which is slightly higher than the findings of

previous studies. Tinnitus was present in almost half of the patients, but reports of the incidence of this symptom vary from one-third to over 80% in other studies,^(2,6,10) so its actual frequency in VS is unclear.

We noted that several patients had normal audiograms. We also note that high-frequency hearing loss occurs with normal aging, when it is termed 'presbycusis'.

The weak co-relation between size of VS and hearing deficit has been reported elsewhere.⁽⁹⁾

This would support the findings of a 2008 systematic review, whose authors suggest that vascular loops are associated with some cases of unilateral hearing loss and pulsatile tinnitus.⁽¹⁷⁾ It is not possible to determine whether there is a common clinical picture seen in patients with epidermoid cysts or meningiomas because there was only one patient with each of these conditions in this study. Future research in this area would need a multicentre study to gather sufficient patients with these uncommon conditions.

LIMITATIONS OF THE STUDY

This is a larger study sample than other reported series but suffers from deficiencies in availability of notes and audiometry records.

Collecting data from clinic letters helps to avoid recall bias associated with patient-completed questionnaires, but the letters may not have included all the symptoms the patient presented with, especially if they were infrequent attacks of vertigo or dizziness. Therefore, the clinical picture could be inaccurate. This could be partly avoided by using the patient's hospital notes for data collection, although they may still not give a complete picture.

Finally, the patient's diagnoses were made from the MRI scan report and not confirmed pathologically. Therefore, some of the initial diagnoses may have been incorrect, thus the study results are not a true representation of the different CPA pathologies.

CONCLUSION

CPA lesions can present in a number of ways, including audiovestibular symptoms, changes in facial sensation and headache. Ear pain and ear fullness are infrequently reported symptoms but are suggestive of a CPA mass. This study concurs with guidelines recommending that patients presenting with hearing loss or tinnitus without an obvious cause should be investigated for the presence of VS. However, it is important to bear in mind that 10% of VS present in atypical ways, such as fluctuating hearing loss with normal or symmetrical pure-tone audiograms.⁽¹¹⁾ A normal audiometry result does not exclude the possibility of a CPA mass and clinicians should still use their clinical judgement for patients with suspicious symptoms.

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