HEARING LOSS IN VESTIBULAR SCHWANNOMA: SIZE ISN’T EVERYTHING

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BACKGROUND

Vestibular schwannoma (formerly known as acoustic neuroma) is a benign tumour most commonly arising from the inferior vestibular division of the VIIIth (vestibulocochlear) cranial nerve. Traditionally, symptoms were thought to result from direct tumour compression of the nerve within the internal auditory meatus (IAM). However, if this was true:

- Why do 95% of patients with a benign tumour of the vestibular nerve present with hearing loss (a function of the adjacent cochlear nerve) rather than vestibular symptoms?
- How can some large tumours produce less severe hearing impairment than smaller ones?
- Why do some patients experience a deterioration in hearing despite a lack of tumour growth?

We noted reduced T2 signal in the ipsilateral cochlea in several cases of vestibular schwannoma (figure 1) prompting the question.

IS COCHLEAR T2W SIGNAL RELATED TO HEARING LOSS AND THE SIZE OF THE VESTIBULAR SCHWANNOMA?

The following summary of the current literature aims to answer these questions and improve our understanding of the mechanisms underlying hearing loss in vestibular schwannoma.

HEARING LOSS IN VESTIBULAR SCHWANNOMA

Patients with vestibular schwannoma (VS) most commonly present with sensorineural hearing loss (SNHL). This type of hearing loss can be seen when there is damage to any structure along the auditory pathway from the inner ear to the brainstem (figures 1 and 2). The mechanisms underlying SNHL in patients with VS remain poorly understood.

A growing body of evidence suggests that direct neural compression within the IAM is not the sole cause of symptoms and the underlying process is likely to be multifactorial with cochlear pathology playing a significant role. In support of this:

- Tumour size, location and nerve of origin do not correlate with the degree of hearing loss1,2,3.
- Clinical audiologic testing can differentiate cochlear from retrocochlear hearing impairment and both processes have been shown to play a part in SNHL associated with VS, in some cases cochlear deficits occurring independently of retrocochlear deficits4,5.
- Pathological examination in patients with VS have revealed cochlear degeneration (including damage to the outer hair cells, stria vasculosa and spiral ligament) in addition to, and in some cases independent of, expected vestibulocochlear nerve degeneration.
- Cochlear perilymph protein content has been shown to be 5-15 times higher in patients with VS than controls.
- Histologically, perilymph was sampled by ‘labyrinthine tap’ as a diagnostic tool prior to the development of modern non-invasive imaging techniques.

ALTERED COCHLEAR SIGNAL IN VESTIBULAR SCHWANNOMA

Increased protein content within perilymph explains reduced signal observed in the cochlea on T2 weighted MR sequences (figure 1). Conventional fluid-attenuation inversion recovery (FLAIR) and more recently 3D-FLAIR sequences which have a higher sensitivity for the detection of increased protein content, have been used by several authors in an attempt to quantify protein levels within the perilymph and correlate this with observed hearing deficits4,5,6. In summary:

- Most authors have shown either no correlation or only very weak correlation between cochlear signal and the degree of hearing impairment4,6.
- Kim DY et al (2014) suggest a moderate correlation with intracanalicular VS cochlear signal and HL as measured by pure tone audiometry, but no correlation when looking at tumours extending to the cerebellopontine angle. They conclude cochlear signal may be an additional parameter to use when monitoring functional impairment during follow up of patients with small intracanalicular VS5,6.
- Petrovic BD et al (2010) suggest the increased inner ear FLAIR signal can be used as a diagnostic adjunct to alert the radiologist to the presence of an otherwise subtle VS5.
- Van de Langenberg R et al (2011) suggest patients who demonstrated hypointensity of the labyrinth at presentation will have faster rate of deterioration in hearing12.

WHAT IS THE CAUSE AND SIGNIFICANCE OF INCREased PERILYMPH PROTEIN?

The underlying cause and significance of the increase in perilymph protein content remains uncertain. The most commonly described theories include:

- Breakdown of the blood-perilymph barrier due to high venous pressure or arterial stasis as a result of vascular compression by tumour4.
- Blocked neuroaxonal protein transport along the cochlea nerve secondary to tumour compression.
- Cell mediated immune reaction within the inner ear to the antigentic properties of VS.

Of interest there has been a recent focus on the use of molecular biology techniques13,14.

CONCLUSION

Sensorineural hearing loss in VS is multifactorial and involves both cochlear and retrocochlear processes.

Reduced T2 signal within the cochlea in VS is due to an increase in perilymph protein. The nature and significance of this remains uncertain at present, although it is the subject of much ongoing research. The size of the tumour does not appear to correlate with changes in protein content.

Radiologically, altered protein content can be most accurately assessed using 3D-FLAIR which demonstrates higher sensitivity and reduced artifact compared with traditional sequences performed in the initial investigation of SNHL. No studies to date have demonstrated a convincing correlation between imaging signal i.e. protein content and the degree of hearing impairment.

Currently treatment options are limited to ‘watchful waiting’ and surgery. Recent molecular studies offer the hope of medical therapy in the future.

As our understanding of the mechanisms of hearing loss in VS and our ability to predict patterns of hearing loss improves, the patient can be better informed in their decision regarding invasive hearing preservation surgery. What is currently clear however is that:

- SIZE DOESN’T MATTER WITH RESPECTS TO VESTIBULAR SCHWANNOMA, COCHLEAR PROTEIN CONTENT AND SNHL

REFERENCES

5. Lassalexa SF, Goh AC, VYR et al. Secreted factors from human vestibular schwannomas ( ) and reduced signal in the (polateral cochlea (arrow). In (a) note the large tumour extends into the cerebellopontine angle compressing the adjacent middle cerebellar peduncle and brainstem and partially effacing the fourth ventricle.

FIGURE 1: Axial T2 constructive interference in steady state (CISS) images at the level of the internal acoustic meatus in a series of four patients (a-d) demonstrating left sided vestibular schwannomas ( ) and reduced signal in the (polateral cochlea (arrow). In (a) note the large tumour extends into the cerebellopontine angle compressing the adjacent middle cerebellar peduncle and brainstem and partially effacing the fourth ventricle.

FIGURE 2: Axial schematic of an intracanalicular VS.


- Sensory cochlear hearing loss (SNHL)
- Pathway can involve any structure from inner to outer nuclear layer to cochlear nucleus.

- Blood perilymph barrier (BPB)
- Neuroaxonal transport (NAT)
- Immunological reaction (IR)
- Proteolysis (P)

- Secreted factors from human vestibular schwannomas ( ) and reduced signal in the (polateral cochlea (arrow). In (a) note the large tumour extends into the cerebellopontine angle compressing the adjacent middle cerebellar peduncle and brainstem and partially effacing the fourth ventricle.

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