Sudden sensorineural hearing loss:

early diagnosis improves outcome

INTRODUCTION

Sudden sensorineural hearing loss (SSNHL) is one of the most under recognised medical emergencies. Unilateral loss of hearing does not produce the same immediate concerns as unilateral loss of vision and the differential includes very common and benign conditions such as wax obstruction and otitis media with effusion. These two factors often conspire to delay the patient's initial consultation and subsequent referral to the ear, nose, and throat (ENT) specialist. This is unfortunate as there is now evidence that early action significantly improves the chances of obtaining some recovery in hearing thresholds.

DEFINITION

There is no universal definition of SSNHL but based on the patient's pure tone audiogram, it is often defined as an increase in pure tone threshold of greater than 30 decibels (dB) in at least three adjacent frequencies occurring within 72 hours. Its peak incidence is between 50 and 70 years of age although it can occur at any age. Figure 1 shows a typical audiogram of someone with a sudden unilateral sensorineural hearing loss.

DEMOGRAPHICS AND AETIOLOGY

SSNHL is uncommon with around 20 people in every 100 000 being affected every year. The cause is not known in around 70% of cases.¹ In these idiopathic cases the assumption is that there is either a viral or a vascular aetiology. Table 1 shows the more common pathologies that make up the 30% of cases that have an identifiable underlying cause for their SSNHL.

The hearing loss may be associated with tinnitus, vertigo, and aural fullness. Other otological symptoms are rare.

A number of negative prognostic factors have been suggested including advanced age, the presence of vertigo, audiogram shape, and a family history.² A recent review of the literature undertaken by Lin *et al* also suggests that there is greater risk of SSNHL in patients with risk factors for cardiovascular disease.³

INVESTIGATION

Certain investigations for specific causes of SSNHL should be undertaken even though they only infrequently identify any underlying pathology. The most important investigation is a magnetic resonance scan of the internal auditory meati to exclude a vestibular schwannoma. Serological investigations are of limited use although rare treatable infections such as syphilis and toxoplasmosis should be excluded.

TREATMENT

There are a number of different treatments that have been used in sudden sensorineural hearing loss. This includes oral or intravenous steroids, antiviral agents, vasodilating agents such as carbogen gas inhalation (this is a mixture of 95% oxygen and 5% carbon dioxide), and diuretics. There is no evidence that any of these have any material effect on hearing.

Recently, however, there has been a great deal of interest in the use of intratympanic (IT) injection of steroids through the tympanic membrane. This has been shown to deliver up to a 250 times greater dose of steroid to the inner ear than oral steroids, and is not associated with the potential adverse effects of systemic steroids; although some clinicians use this technique in conjunction with systemic steroids. There are a large number of papers investigating the influence of IT steroids for the management of SSNHL.⁴ However, many of the studies are flawed, not least because up to 60% of SSNHL can make some degree of spontaneous recovery and large scale studies are required to obtain useful results in what is a fairly rare condition.

A meta-analysis of the literature regarding the use of IT steroid in SSNHL showed that in general, between 50 and

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The blue triangles denote inner ear hearing function in the better hearing ear (right). The blue brackets represent the inner ear function of the left ear and the downward pointing arrows mean that there is no measureable inner ear function on the left.

Figure 1. A typical pure tone audiogram of a patient with a sudden unilateral sensorineural hearing loss. For each frequency of sound the tester reduces the sound intensity of the tone to a level at which the patient can only just hear it. This is the hearing threshold. The overall hearing threshold is marked as a red circle for the right and a blue cross for the left ear. In this case, there is no measurable hearing threshold at any frequency in the left ear but normal compared to 0–15dB in controls. The mean thresholds in the right ear. The pattern is variable however.

75% of patients show an improvement in hearing threshold of 10–15dB or more with IT steroid, compared to 0-50% in controls who in the main received oral steroids.⁴ The mean improvement in pure tone average in the IT steroid group ranged from 15-30dB benefit of IT steroid relative to controls

Table 1. Identifiable causes of sudden sensorineural hearing loss

Infection Viral infection, for example, mumps, measles, herpes zoster Bacterial labyrinthitis secondary to chronic middle ear disease Meningitis	44%
Ear disease Ménière's disease Ear surgery Ototoxicity, for example, aminoglycosides Autoimmune disease, for example, autoimmune inner ear disease (AIED) Otosclerosis	17%
Trauma Head injury (temporal bone fracture or cerebral trauma) Sudden loud noise exposure Barotrauma resulting in round window rupture, for example, from diving or flying	15%
Neurological and vascular disease Cardiovascular disease Transient ischaemic attack Multiple sclerosis	12%
Neoplastic Vestibular schwannoma Meningioma Other	8%

was 13dB. Some evidence suggests that IT steroid therapy is more useful to salvage hearing following ineffective oral steroid therapy than if it had been used as a primary treatment.^{4,5} Various types of steroids have been used but there is no evidence that one is better than another. The key is to treat early. The earlier the treatment is commenced the higher the chance of obtaining a significant improvement in hearing thresholds. By 2 weeks the chances of gaining any significant benefit are very low.

The injections are performed under local anaesthetic. The tympanic membrane is anaesthetised by using either 10% xylocaine spray, phenol, or a eutectic agent such as Ametop® (Smith & Nephew Healthcare) gel or EMLA® (AstraZeneca) cream. After the injection, the patient lies on their side for half an hour to allow diffusion of the steroid through the round window membrane.

The subsequent protocol varies from centre to centre but a course of daily injections over 3 days followed by an audiogram is common practice. If there is ongoing improvement in the audiogram then injections can be continued until the audiogram has stabilised or returned to normal.

OPTIMISING OUTCOMES

Many of the common differential diagnoses of sudden hearing loss are conductive in nature. Therefore tuning-fork tests such as Reine's and Weber's tests can be useful in differentiating a sudden sensorineural loss from a conductive loss and may be used to determine the urgency of the referral. If the sudden hearing loss is apparently sensorineural then an urgent referral to the local ENT department should be made so that early treatment can be started. The key to effective treatment is patient and doctor awareness of the need for early presentation and rapid onward referral.

Provenance

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