Aetiology, investigation and acute management of sudden sensorineural hearing loss

BY EDWIN HALLIDAY

The cause of a single sided sensorineural hearing loss has a wide variety of aetiologies. This review by **Edwin Halliday** looks specifically at the differential diagnostic causes of a sudden sensorineural hearing loss, the relevant investigations and the management should no obvious treatable cause be found.

Hearing loss can be defined by site of disease, severity, audiometric configuration and method of onset. Sudden sensorineural hearing loss (SSHL) is rare with the incidence estimated at five to 30 cases per 100,000 per year [1]. There is no strict definition, but it is often defined as a hearing loss of at least 30 dB in three sequential frequencies in the standard pure-tone audiogram (PTA) which has occurred in less than three days [2].

Clinical presentation

The age distribution of patients varies widely, with an average age of between 50 and 60 years and no sex preference [1]. Typically the hearing loss is unilateral, but in less than 5% of cases it is bilateral [3]. The severity of hearing loss can be defined based on the audiometry as mild, moderate, severe or profound [1] and can affect high, low or all frequencies. Associated symptoms include tinnitus in 80% of patients and vertigo in approximately 30%. SSHL can present as an isolated problem or can be associated with a systemic disease.

Causes

A number of causes of SSHL have been described, together with theories about potential underlying pathological mechanisms [1]. Detailed investigation will only identify a cause of SSHL in around 10% of patients [4]. In the remainder, no obvious cause is found and the term idiopathic SSHL (ISSHL) is used.

Many viruses have been suggested as causative organisms of SSHL although serological, epidemiological and histopathological data are inconclusive. These viruses include cytomegalovirus, enterovirus, Epstein-Barr virus, herpes simplex type I and II, influenza, mumps, rubella and varicella-zoster virus. Other infectious causes include bacterial meningitis, Lyme disease, fungal meningitis, otosyphilis, HIV and Creutzfeldt-Jakob disease.

The cochlea is supplied by an end artery and vascular occlusion has been suggested as another cause for SSHL.

Hyperviscosity syndromes such as sickle-cell syndrome, Waldenstrom's macroglobulinaemia and myeloma may be associated with SSHL. A vascular pathology may be the underlying mechanism for SSHL in giant-cell arteritis, Takayasu's arteritis, Wegener's granulomatosis, systemic sclerosis, Sjögren's syndrome, relapsing polychondritis and polyarteritis nodosa. SSHL is common in patients with Behçet's disease and asymptomatic hearing loss can be seen in systemic lupus erythematosus.

Cogan's syndrome is the rare combination of hearing loss and inflammatory eye disease. Half of patients present with SSHL, which is usually bilateral, asymmetric and often progressive over one to three months. It is often accompanied by acute vertiginous episodes and balance loss. One complication includes medium to large vessel vasculitis, including aortitis which can lead to aneurysmal dilatation and aortic regurgitation.

Vestibular schwannoma is the most common neoplastic cause of SSHL. It usually presents with unilateral tinnitus and progressive sensorineural hearing loss, although it can present with sudden loss.

Trauma may also account for SSHL; causes include barotrauma, acoustic trauma, ear surgery and temporal bone fracture [5].

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Drug-induced hearing loss is usually gradual. However, it is recognised that some drugs, including penicillin, oestrogens, phosphodiesterase-5 inhibitors, pegylated interferon, ribavirin, aminoglycosides and chemotherapeutic agents, can occasionally cause SSHL.

Unilateral or bilateral SSHL can present during the course of demyelination or as the initial symptom. Hearing loss can occur in neurosarcoidosis and SSHL can be the presenting symptom alone or in combination with other neurological signs.

Pseudohypacusis, where hearing loss is apparent in the absence of clinical or audiological evidence, is not an infrequent cause of apparent sudden hearing loss in school-aged children and young women.

Clinical assessment

History and examination

Assessment should begin with a thorough history and clinical examination. Examination should include otoscopy and tuning fork tests. A sensorineural hearing loss can be confirmed with masked audiometry, which also documents the severity of the hearing loss and provides a baseline for assessment of recovery and efficacy of treatment.

If SSHL has been confirmed, patients should undergo a full neuro-otological examination to establish the presence of any peripheral or central vestibular dysfunction [1].

Blood tests

There is little evidence that routine blood tests will alter the diagnosis, treatment or prognosis of SSHL. However, they may provide additional evidence to support clinical suspicion of systemic disease.

Imaging

The British Association of Otorhinolaryngologists – Head and Neck Surgeons (BAO-HNS) recommends that any patient complaining of unilateral or asymmetrical auditory symptoms (either hearing loss or tinnitus), whether of progressive or sudden onset, in whom there is no other obvious cause, should be screened with MRI [6]. In patients presenting with SSHL, the MRI will detect a vestibular schwannoma in about one in 20 patients [7]. The MRI will also allow other abnormalities to be detected.

Management

If a systemic or local cause of the hearing loss is identified then treatment should be targeted towards this. If the cause of the hearing loss is idiopathic then a number of general treatment regimens have been suggested. The evidence for many of these treatments is mixed and there is a high rate of spontaneous recovery, varying from 30 to 68% [8].

Oral steroids

Oral steroids are the most common therapeutic intervention in ISSHL. They are thought to decrease inflammation and oedema [5]. One suggested regimen is to use oral prednisolone at a dose of 1mg/kg/day (up to a maximum dose of 60mg daily) for seven to 14 days and then to taper over a similar time period.

The best evidence for the use of oral steroids in ISSHL is from a Cochrane review (updated in 2013) [9] which concluded that "the value of steroids in the treatment of idiopathic sudden sensorineural hearing loss remains unclear". The review identified only three suitable trials involving a total of 267 patients. All three studies were at high risk of bias. One trial showed a significant improvement in hearing in 61% of patients receiving oral steroids, compared to only 32% of patients in the control group [10]. The other two trials showed a lack of effect of oral steroids compared with the placebo. Another meta-analysis suggested an improvement with oral steroid therapy, but this failed to reach statistical significance [8].

The systemic administration of steroids is not without risk and their use must be balanced against potentially hazardous side-effects in certain patient groups such as those with hypertension, diabetes or glaucoma.

Intratympanic steroids

The rationale for intratympanic administration of steroids is the same as for oral steroids, although as it is organ-specific treatment there are two theoretical advantages: reduced systemic steroid uptake and the potential of direct steroid uptake through the round window membrane resulting in higher perilymph concentration [11, 12]. There is no clear consensus for intratympanic therapy and regimens using dexamethasone or methylprednisolone every three to seven days for three to four sessions have been suggested [5].

The evidence appears to suggest that intratympanic steroids are at least as effective as oral steroids when used as a primary treatment [13]. Alternatively intratympanic steroids can be used as salvage therapy, where there is some evidence that they may provide additional benefit when there has been no response to oral steroids [14].

As there are fewer systemic side-effects compared to oral steroids, intratympanic steroids may be particularly useful in patients who would otherwise have contraindications to oral steroid treatment.

Alternative acute therapies

The use of hyperbaric oxygen, antiviral therapy, vasodilators and vasoactive substances have all historically been utilised in the treatment of ISSHL. There is currently insufficient evidence to support the routine use of these strategies.

Further management

Unilateral hearing loss is typically managed in the outpatient setting while patients with severe bilateral loss may require admission to hospital if there are concerns about ability to cope at home with the new level of hearing disability. Temporary amplification may be required to help the patient communicate. More permanent amplification via hearing aids, bone anchored hearing aids and cochlear implantation should be considered if there is no satisfactory resolution of hearing levels after a period of observation [5].

Conclusion

SSHL is an important condition that can have a significant impact on a patient's quality of life. Careful initial assessment is necessary to exclude potentially treatable causes, although for the majority of patients the cause will be idiopathic. All patients should have an MRI scan to exclude a retrocochlear lesion. There is contradictory evidence about the best therapy to offer patients, but most clinicians would offer oral or intratympanic steroids, with intratympanic steroids being favoured in those with contraindications to systemic treatment. Salvage intratympanic steroids should also be considered in those who have failed to respond to initial therapy with oral steroids. Patients should be well informed about the evidence and side-effects of any therapy offered to them to allow a fully informed decision to be made. There is still much to be learnt about the pathogenesis of SSHL and more clinical trials are necessary to establish evidence-based treatment.

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Declaration of Competing Interests
None declared.

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