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Clinical Record

Sweet's disease and profound, bilateral, sensorineural hearing loss

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Abstract

Objective: We report a case of Sweet's disease associated with rapid, profound loss of hearing, against a background of progressive, bilateral, sensorineural hearing loss.

Results: The clinical features were indistinguishable from those of immune-mediated inner ear disease. Establishment of a definitive diagnosis was a challenge due to the absence of a reliable diagnostic test. The patient was unresponsive to extensive immunosuppressive therapy and subsequently underwent cochlear implantation, with good hearing outcomes.

Conclusions: Profound, bilateral, sensorineural hearing loss in the context of Sweet's disease may be related to the underlying immunological aetiology. Cochlear implantation can successfully restore hearing when immunotherapy fails.

Key words: Sweet's Disease; Sensorineural hearing loss; Cochlear Implant

Introduction

Sweet's disease, or acute febrile neutrophilic dermatosis, consists of the following clinical triad: erythematous cutaneous plaques, infiltrated by mature neutrophils, in association with fever and leukocytosis.[1–3] The pathogenesis of this condition has yet to be clearly elucidated, but an immune aetiology has been implicated based on clinical response to corticosteroids. As yet, there have been few reports of progressive, bilateral, sensorineural hearing loss (SNHL) in the context of Sweet's disease.[4,5] An autoimmune process targeting the inner ear may be responsible for this phenomenon, similar to the process described in autoimmune inner ear diseases. Sweet's disease together with progressive, bilateral SNHL is a potentially treatable condition, and timely initiation of immunotherapy and evaluation for cochlear implantation for the profoundly deafened can significantly improve auditory outcomes and quality of life.

We present a patient with Sweet's disease who was affected by sudden, rapid, profound hearing loss, against a background of progressive, bilateral SNHL. The patient was unresponsive to extensive immunotherapy and subsequently underwent cochlear implantation, with considerable improvement in speech perception.

Case report

A 63-year-old woman with a 10-year history of Sweet's disease, associated with myelodysplasia, visual disturbance, mild parkinsonian symptoms and skin ulcerations, was referred for investigation of sudden, rapid, bilateral hearing loss. The patient's diagnosis of Sweet's disease had initially been confirmed by a skin biopsy; prednisone (10 mg daily) had been commenced and continued long term for disease control. Bilateral SNHL had first been noted seven years prior, during a particularly severe exacerbation of Sweet's disease. Over the subsequent three years, serial pure tone audiograms indicated progressive, symmetrical, bilateral SNHL affecting the higher frequencies. A hearing aid had been fitted for the right ear, followed by the left, but their effectiveness had declined over time.

Over the few months prior to the current presentation, the patient reported the rapid onset of sudden, profound hearing loss, with no accompanying tinnitus or vertigo.

The patient appeared cushingoid and profoundly deafened, but clinical examination was otherwise unremarkable.

Pure tone audiography demonstrated near-symmetrical, severe to profound SNHL bilaterally, with no recordable speech perception (Figure 1). Lip-reading was compromised by poor vision. Anti-nuclear antibody testing was positive, with a homogeneous, speckled pattern and a titre of 160; however, the rest of the autoimmune screen was unremarkable, as was blood biochemistry and inflammatory marker analysis. Assay for heat shock protein 70 was negative. A magnetic resonance imaging scan of the brain, performed two years previously, was within normal limits.

The patient's hearing loss was suspected to be due to an autoimmune aetiology in the context of Sweet's disease. Three doses of 500 mg pulsed intravenous methylprednisolone were administered, and the patient's subjective hearing improved remarkably; she was able to use the telephone within 24 hours. Unfortunately, her hearing loss again deteriorated to profound levels over the next two to three weeks, despite ongoing immunosuppression with oral prednisone (50 mg daily), mycophenolate (250 mg twice daily) and a course of intratympanic dexamethasone injections to the right ear. Cyclosporin was

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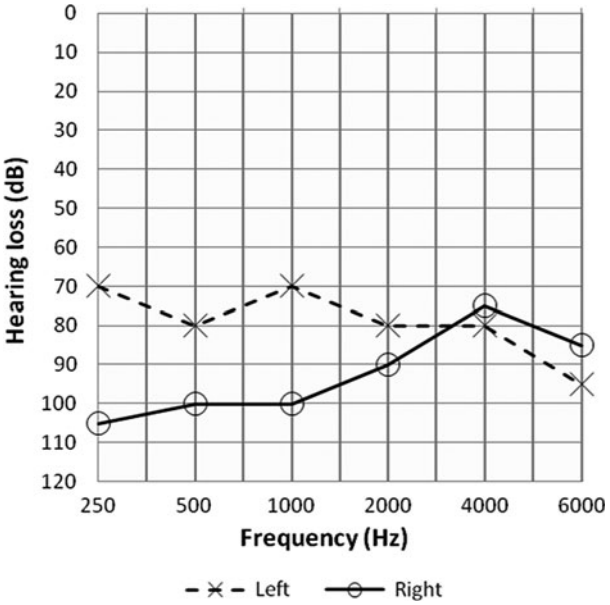

FIG. 1

Pure tone audiogram recorded prior to cochlear implantation, showing severe to profound, sensorineural hearing loss (SNHL) for the left ear, and for the right ear a profound SNHL in the low frequencies, sloping up to a severe SNHL in the high frequencies.

trialled but was ceased due to side effects. The dose of mycophenolate was increased to 500 mg twice daily, but this was ceased after three months when the patient's hearing did not improve. Low dose prednisone (8 mg daily) was continued for control of Sweet's disease. A repeated analysis of inflammatory markers was within normal limits.

As she had no residual hearing despite hearing amplification, the patient was thoroughly assessed for cochlear implant candidacy. No measurable response was obtained in either ear on electrocochleography. Tympanometry was normal, and stapedial reflexes were absent bilaterally. No consistent responses were obtained upon promontory stimulation. Brainstem evoked response audiometry demonstrated normal auditory nerve and brainstem conduction. Pre-operative computed tomography imaging of the petrous temporal bones was unremarkable.

The patient underwent cochlear implantation of the right ear, using a Nucleus CI-24RE(ST) implant (Cochlear Ltd, Lane Cove, NSW, Australia), 12 months after initial presentation. Post-operative recovery was uncomplicated.

When the patient's device was switched on, three weeks post-operatively, her speech discrimination scores were greatly increased. At three months post-operatively, her speech perception with the cochlear implant (auditory mode only), as assessed by the City University of New York sentence test (an open-set speech perception test, delivered via pre-recorded compact disc in quiet (65 dB SPL) and in noise (+10 signal to noise ratio)), was 99 per cent correct in quiet conditions and 41 per cent correct in noisy conditions. The aided average for the right ear across four frequencies (0.5, 1, 2 and 4 kHz) was 23.75 dB. The patient conversed well with others and could use the telephone via her cochlear implant. She reported considerable improvements to her overall quality of life.

At the time of writing, the patient was awaiting cochlear implantation of her left ear.

Discussion

Sweet's disease

Described by Robert Douglas Sweet in 1964, Sweet's disease (also known as acute febrile neutrophilic dermatosis or Sweet's syndrome) is a multi-system, inflammatory disorder characterised by a clinical triad comprising painful, raised erythematous plaques, infiltrated by mature neutrophils, associated with fever and leukocytosis.[1–3] The postulated pathogenesis relates to abnormal regulation of the immune system, involving various inflammatory cells, keratinocytes and cytokines. An association with particular human leukocyte antigen subtypes, altered T-cell functions, up-regulation of cytokines and adhesion molecules, and altered neutrophil chemotaxis has been proposed. However, a rapid therapeutic response to systemic corticosteroids and immunosuppressants remains the most convincing evidence for this immunological aetiology.[1–4] Multi-system involvement can affect the cardiovascular, central nervous, gastrointestinal, hepatic, musculoskeletal and pulmonary systems. Cases of bilateral SNHL in association with Sweet's disease have rarely been reported.[4,5]

Sweet's syndrome and bilateral, progressive, sensorineural hearing loss

First described by McCabe in 1979, idiopathic, progressive, bilateral SNHL is a rare form of rapidly progressive, bilateral SNHL which responds clinically to high doses of dexamethasone and cyclophosphamide.[6] The clinical picture is often variable, with rapidly fluctuating, progressive loss of hearing in both ears over a period of weeks to months. The diagnosis is made challenging by the absence of a definitive diagnostic test. Sensorineural hearing loss occurring in the context of systemic autoimmune diseases (such as relapsing polychondritis, Cogan's syndrome, systemic lupus erythematosus and Wegner's granulomatosis) has been well described in the literature. Also known as immune-mediated inner ear disease, the observed clinical response to corticosteroids and immunosuppressants supports an immunological aetiology targeting the inner ear.[7]

- **Sweet's disease, or acute febrile neutrophilic dermatosis, is characterised by a clinical triad comprising erythematous cutaneous plaques, infiltrated by mature neutrophils, in association with fever and leukocytosis**
- **This paper describes a case of Sweet's disease associated with rapid, progressive, bilateral, sensorineural deafness**
- **Prompt referral for audiological assessment and a trial of immunotherapy is warranted**
- **Early referral for cochlear implantation may be indicated**

The precise association between Sweet's disease and progressive, bilateral SNHL has not been clearly elucidated, due to the rare occurrence of such cases. An immune process targeting the inner ear has been suspected, given the immunological aetiology of Sweet's disease. The rapid, fluctuating and relentless nature of the SNHL observed in our patient mirrored the clinical picture of other reported immune-mediated inner ear diseases. In such cases, thorough investigation and timely initiation of first-line corticosteroid therapy is warranted, given the potential to preserve residual inner ear function and

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reverse hearing loss. In cases unresponsive to immunotherapy, auditory rehabilitation with hearing aids and cochlear implants should be considered. Highly successful outcomes of cochlear implantation have been demonstrated in adults post-lingually deafened by immune-mediated SNHL. Such success may be due to: the relatively short duration of profound deafness; the pathological site of deafness being limited to the organ of Corti; preservation of the spiral ganglion cell population, auditory nerves and higher pathways; and retained memories of sounds.[8] A theoretical risk of post-operative infection and wound dehiscence associated with a considerable history of immune-suppression has been reported; however, such complications were not evident in our patient.

Conclusion

To date, there have been few reports of Sweet's disease associated with rapid, progressive, bilateral SNHL. Such audiological effects of Sweet's disease may be potentially treatable, and prompt referral for audiological assessment and trial of immunotherapy is warranted. For the profoundly deafened, timely evaluation for cochlear implantation can lead to significant improvement in speech perception and quality of life.

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Dr S Cheng takes responsibility for the integrity of the content of the paper.
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