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Sensorineural hearing loss and ulcerative colitis

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Abstract
The association of sensorineural hearing loss and ulcerative colitis is well documented and it is speculated that this is autoimmune in origin. A case in a 12-year-old boy is described, that initially responded to steroid therapy, but four years later resulted in bilateral, profound sensorineural hearing loss in spite of good control of his bowel disease. Immunological tests may provide a clue as to the aetiology of suspected cases of autoimmune inner ear disease. Immediate treatment with steroids with or without immunosuppressive therapy is essential as delay may lead to irreversible hearing loss.

Key words: Autoimmune diseases; Colitis, ulcerative; Hearing loss, sensorineural

Introduction

Autoimmune sensorineural hearing loss was first described in 1979 (McCabe). Since then there have been reports in the literature of sensorineural hearing loss in association with systemic immune diseases such as rheumatoid arthritis, Wegener's granulomatosis, giant cell arteritis and inflammatory bowel disease (Weber *et al.*, 1984; Dowd and Rees, 1987; Jacob *et al.*, 1990; Luqmani *et al.*, 1991). We report a case of autoimmune inner ear disease, review the literature and stress the urgency of the treatment of the hearing loss.

Case report

A 12-year-old boy developed acute onset asymmetrical hearing loss, at the same time as a diagnosis of ulcerative colitis was made. Clinical examination showed normal tympanic membranes and a positive Rinne test in both ears. Pure tone audiometry showed hearing thresholds of 90 dB in the right ear and 50 dB in the left ear (average threshold at 0.5, 1, 2 and 4 kHz) (Figure 1), which was confirmed by evoked response audiometry. Impedance audiometry showed normal compliance. All immunological tests were normal and the ESR was elevated (40 mm/hour). A computed tomography (CT) scan with contrast enhancement of the cerebello-pontine angle and brain was normal. He was treated with olsalazine and prednisolone (40 mg/day) for three months. His bowel symptoms were controlled and his hearing improved almost immediately to 50 dB in the right ear and to normal in the left ear (Figure 2). Prednisolone was reduced gradually and stopped after three months and he was continued on a maintenance dose of olsalazine.

Four years later he presented to his general practitioner, four weeks after the onset of sudden, profound, bilateral sensorineural hearing loss (Figure 3) and was immediately referred to the ENT clinic. Tests of immune function

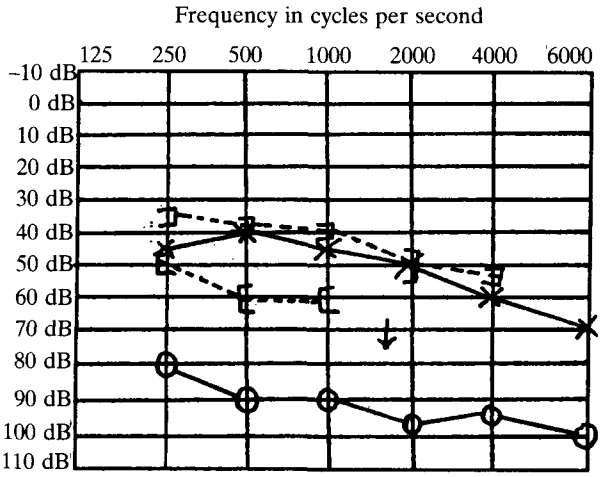

FIG. 1
Pre-treatment pure tone audiogram (1992).

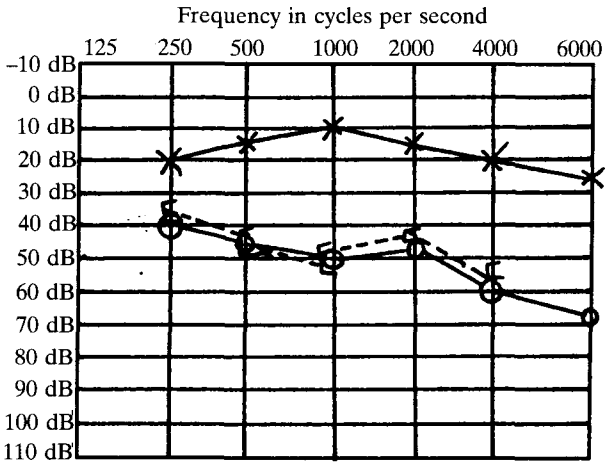

FIG. 2
Post-treatment pure tone audiogram (1992) showing hearing improvement in both ears.

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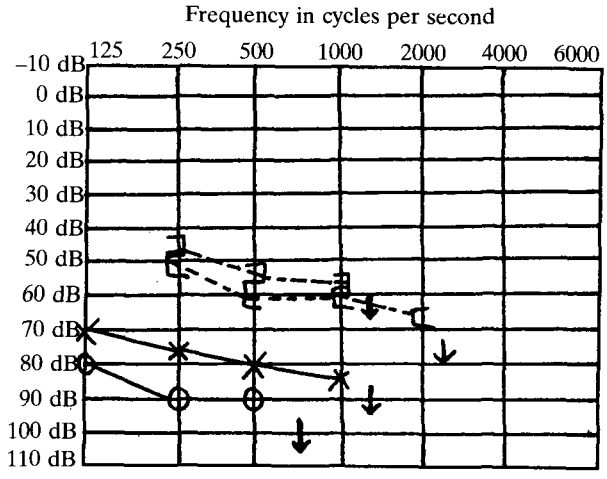

FIG. 3
Pure tone audiogram (1996) showing bilateral, profound sensorineural hearing loss.

showed a raised IgG suggestive of a chronic immune response, positive anti-nuclear antibodies and an ESR of 35 mm/hour. The patient's hearing thresholds did not improve following maximal steroid therapy. Treatment with immunosuppressive drugs was offered to the patient but he refused this line of treatment after the potential side-effects were explained to him. He is at present being considered for cochlear implantation.

Discussion

The association between ulcerative colitis and sensorineural hearing loss is well documented (Summers and Harker, 1982; Weber *et al.*, 1984; Jacob *et al.*, 1990; Herdman *et al.*, 1991). The hearing loss often recovers, at least in part, with aggressive treatment with steroids and cyclophosphamide or azathioprine (Summers and Harker, 1982). Plasmapheresis has however been disappointing in the treatment of patients with sensorineural hearing loss and ulcerative colitis (Luetje, 1989).

Autoimmune inner ear disease may occur due to loss of immunological tolerance to self antigens, cross-reacting antibodies, self antigen modification by infection or drugs or autoimmunity due to hidden self antigens which are exposed due to tissue damage (Herdman *et al.*, 1991). There have been reports of raised circulating immune complexes in such cases (Kanzaki and O-Uchi, 1983; Brookes, 1985).

Though there is no doubt that autoimmune inner ear disease exists, the predictive value of laboratory tests remains poor. McCabe who originally described autoimmune hearing loss now recommends a policy of treating all patients that he suspects clinically of having autoimmune inner ear disease with steroids and cyclophosphamide (McCabe, 1989).

This report illustrates a case of sensorineural hearing loss in association with ulcerative colitis, that initially responded to steroid therapy, possibly suggesting an autoimmune aetiology. It suggests that there is unlikely to be an association between the activity of the colitis and impairment of hearing, as evidenced by this patient who developed profound, bilateral hearing loss in spite of good control of his bowel disease. In addition it shows that laboratory tests for immune disorders may sometimes provide a clue as to the aetiology. We recommend immediate treatment of such sensorineural hearing loss with steroids, with or without immunosuppressive therapy (McCabe, 1979; Summers and Harker, 1982; Weber *et al.*, 1984; Dowd and Rees, 1987; McCabe, 1989; Jacob *et al.*, 1990) to salvage residual auditory function, as sudden loss of hearing, especially in a young patient, results in severe disability. Patients with suspected autoimmune inner ear disease must be advised to present immediately for treatment in the event of a sudden deterioration in their hearing. This case report provides further evidence in favour of the administration of empirical steroid therapy in cases of autoimmune and possibly also idiopathic sensorineural hearing loss.

References

Brookes, G. B. (1985) Immune complex-associated deafness: preliminary communication. *Journal of the Royal Society of Medicine* **78:** 47–55.

Dowd A., Rees, W. D. W. (1987) Treatment of sensorineural deafness associated with ulcerative colitis. *British Medical Journal* **295:** 26.

Herdman, R. C. D., Hirari, M., Ramsden, R. T. (1991) Autoimmune inner ear disease and ulcerative colitis. Letter to the Editor. *Journal of Laryngology and Otology* **105:** 330–331.

Jacob, A., Ledingham, J. G., Kerr, A. I. G., Ford, M. J. (1990) Ulcerative colitis and giant cell arteritis associated with sensorineural deafness. *Journal of Laryngology and Otology* **104:** 889–890.

Kanzaki, J., O-Uchi, T. (1983) Circulating immune complexes in steroid-responsive sensorineural hearing loss and the long-term observation. *Acta Otolaryngologica* **(Suppl.) 393:** 77–84.

Luetje, C. M. (1989) Theoretical and practical implications for plasmapheresis in autoimmune inner ear disease. *Laryngoscope* **99:** 1137–1146.

Luqmani, R., Jubb, R., Emery, P., Reid, A., Adu, D. (1991) Inner ear deafness in Wegener's granulomatosis. *Journal of Rheumatology* **18:** 766–768.

McCabe, B. F. (1979) Autoimmune sensorineural hearing loss. *Annals of Otology, Rhinology and Laryngology* **88:** 585–589.

McCabe, B. F. (1989) Autoimmune inner ear disease. Therapy. *American Journal of Otology* **10:3,** 196–197.

Summers, R. W., Harker, L. (1982) Ulcerative colitis and sensorineural hearing loss: is there a relationship? *Journal of Clinical Gastroenterology* **4:** 251–252.

Weber, R. S., Jenkins, H. A., Coker, N. J. (1984) Sensorineural hearing loss associated with ulcerative colitis. *Archives of Otolaryngology* **110:** 810–812.

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