The Journal of Laryngology and Otology
August 1994, Vol. **108**, pp. 671–675

The possible association of pyoderma gangrenosum and progressive sensorineural deafness

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Abstract
Deafness is known to be associated with certain autoimmune disorders. This article describes a hitherto unreported possible association between pyoderma gangrenosum and progressive bilateral sensorineural deafness.

Key words: Pyoderma, gangrenosum; Hearing loss, sensorineural

Introduction

Pyoderma gangrenosum is a condition characterized by the presence of necrotic ulcers often with a bluish undermined margin. These may occur anywhere on the body and may spread rapidly to cover large areas. Histological examination of these lesions is rarely helpful, revealing only nonspecific inflammation (Shuster, 1978). At least half of all patients with pyoderma gangrenosum are found to have ulcerative colitis (Shuster, 1978). There is some evidence to suggest that pyoderma gangrenosum may be a form of vasculitis, and that local involvement of immune complexes may be the trigger for the production of lesions (Holborrow and Reeves, 1983).

Here we report on a woman with pyoderma gangrenosum who developed progressive bilateral sensorineural deafness.

Case report

A 46-year-old woman presented, more than four years ago, with an episode of giddiness and vomiting which lasted 48 hours. No abnormality was found on otoneurological examination, and an audiogram showed normal hearing in both ears. X-rays of the skull and internal auditory meati did not reveal any abnormality. She remained well for a year and then presented to the dermatologists with painful lesions of the face and forearms which were found to be typical of pyoderma gangrenosum. She did not complain of any symptoms pertaining to her gastrointestinal tract nor was there any history of ulcerative colitis. Her skin lesions responded rapidly to a 10 day course of steroids.

Two years later she developed a recurrence of her skin lesions. At the same time she noted left-sided deafness and dizziness on

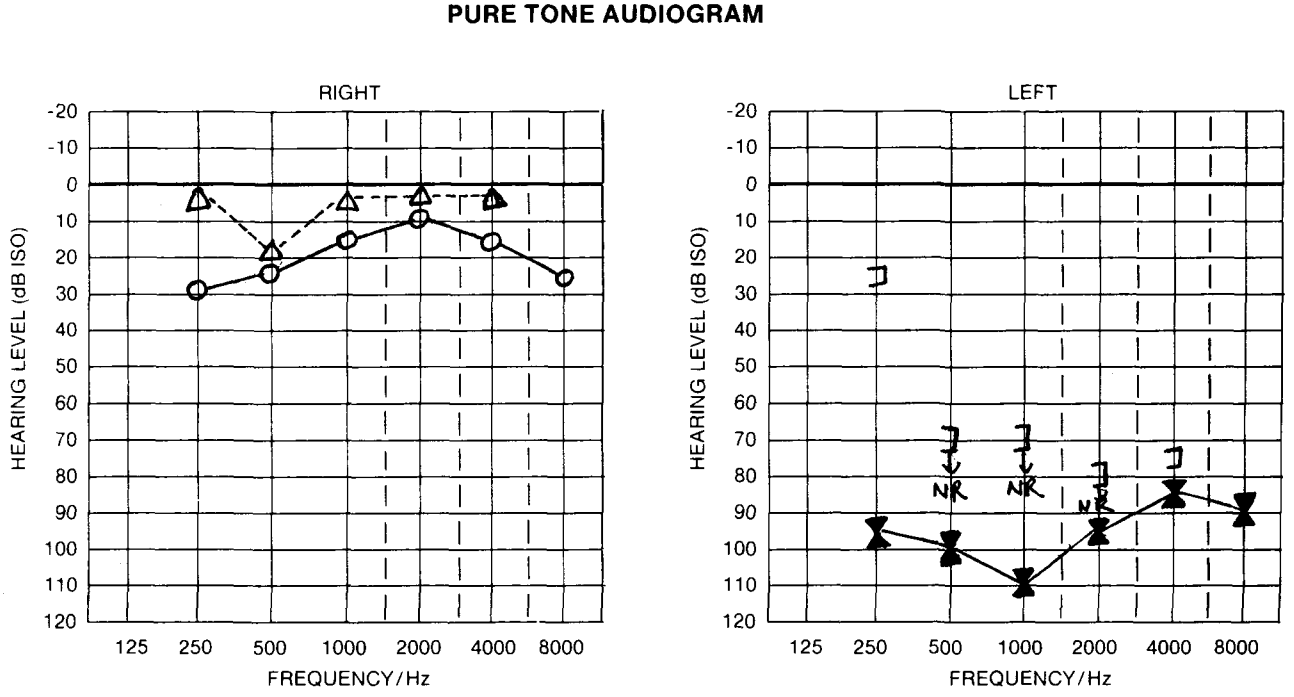

Fig. 1
Audiogram two years after initial presentation.

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Accepted for publication: 11 March 1994.

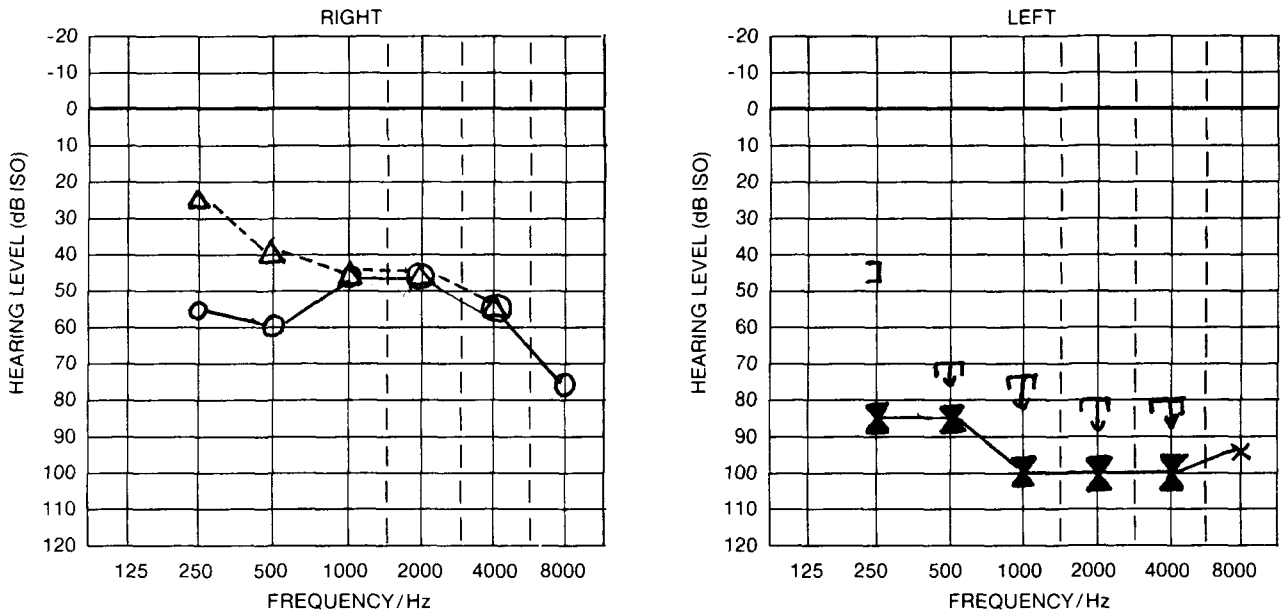

Fig. 2
Audiogram three months later.

sudden movement of her head. Her tympanic membranes were normal, but an audiogram revealed significant deafness in the left ear, while the hearing on the right was normal (Figure 1). Evoked response audiometry did not show significant delay, suggesting the hearing loss was cochlear in nature and caloric tests revealed left canal paresis. CT scan of the posterior fossa and internal auditory meatus were normal. Her pyoderma was being treated with oral prednisolone (60 mg daily) and azathioprine (100 mg daily). Biopsy of a lesion on her cheek revealed non-specific inflammation with evidence of fibrosis and infiltration of the dermis by chronic inflammatory cells. As her skin lesions spread she also noticed deterioration in hearing. An audiogram three months later revealed a 40–60 dB loss in the right ear and profound deafness on the left side (Figure 2). An MRI scan did not reveal a lesion in the cerebellopontine angle. The haemoglobin and differential blood count were normal but the ESR was 81. Urea and electrolytes and liver function tests were normal as before.

While the immunoglobulins level and the albumin globulin ratio have been normal the one consistent abnormality noted on electrophoresis over three years was the presence of a paraprotein of the monoclonal IgG type Kappa light chain. No pro-

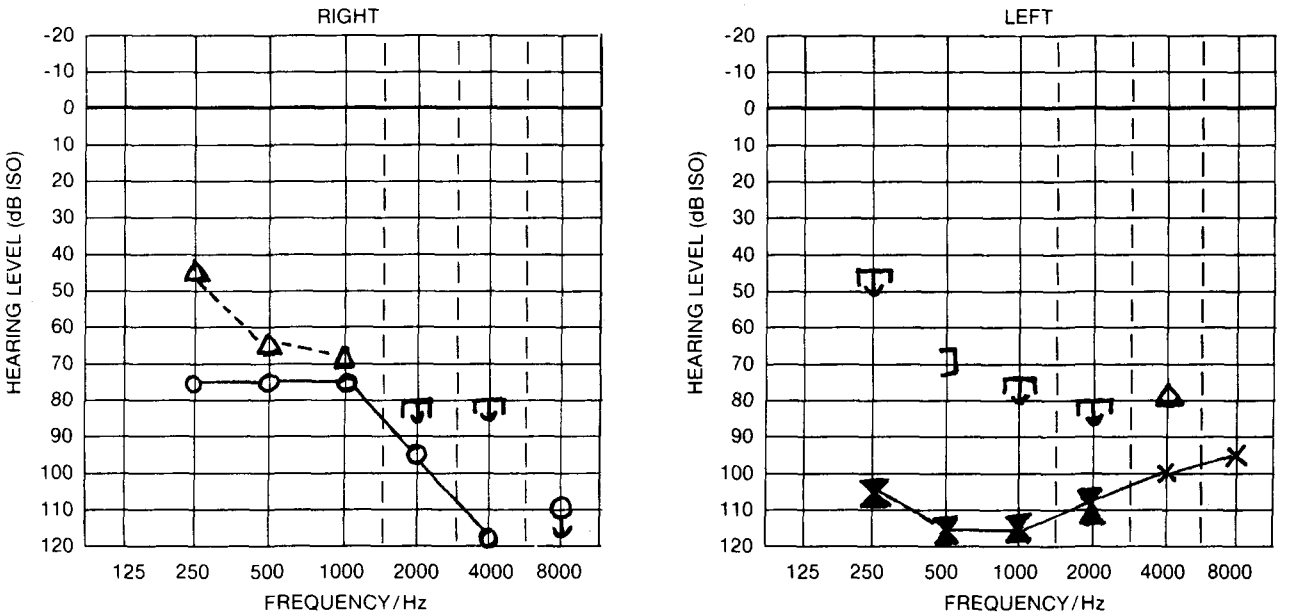

Fig. 3
Audiogram six months later.

https://doi.org/10.1017/S0022215100127793 Published online by Cambridge University Press

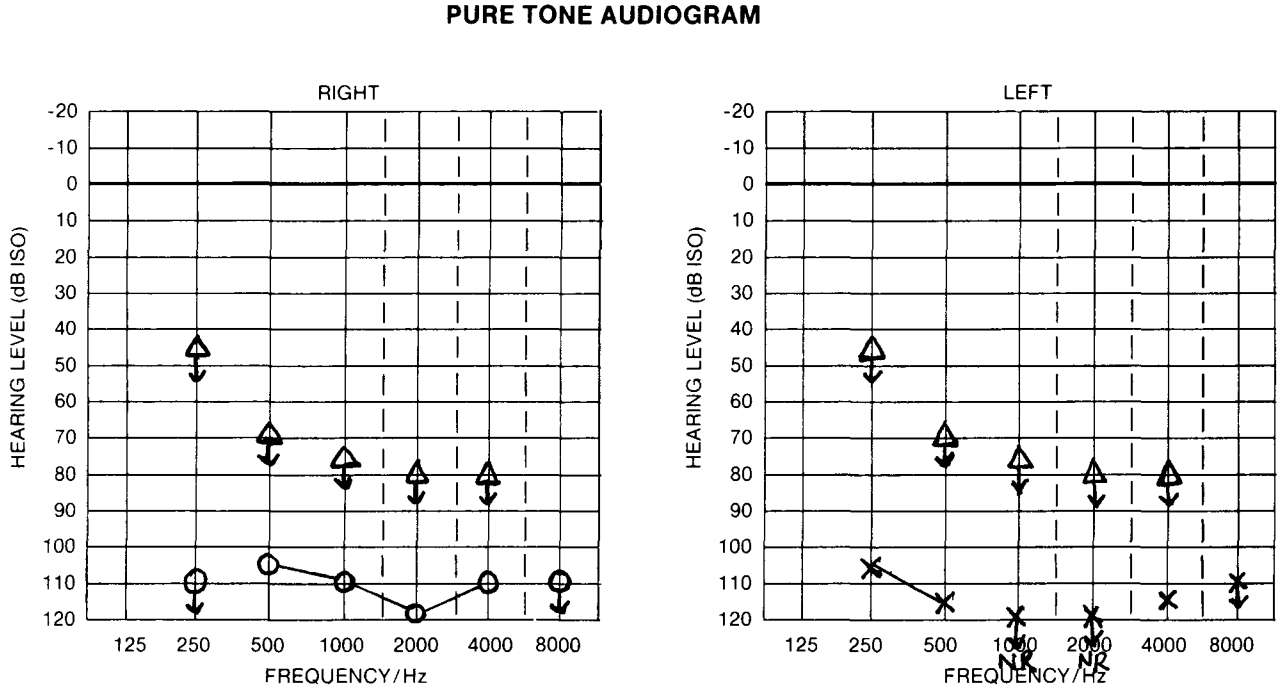

Fig. 4
Audiogram nine months later.

teinuria was present. The hearing continued to deteriorate over the next three months (Figure 3) though the skin lesions had been brought under control and now a right canal paresis was also evident. Within a space of three months she developed severe bilateral sensorineural deafness (Figure 4). An intensive course of oral prednisolone (60 mg daily) and parenteral azathioprine (150 mg daily) produced dramatic subjective improvement and the patient found that she could use her hearing aid much more effectively. Audiograms though not very reliable at this level of deafness showed an improvement of 20 dB in hearing threshold in the right ear (Figure 5). Unfortunately the persistence of nausea and vomiting meant that this treatment had to be terminated and since then her hearing has deteriorated (Figure 6) and skin lesions have spread over her arms and her cheeks (Figures 7 and 8).

This woman has undergone auditory rehabilitation and is using bilateral BE52 hearing aids with little benefit, relying mainly on lip reading. She has been assessed as suitable for a

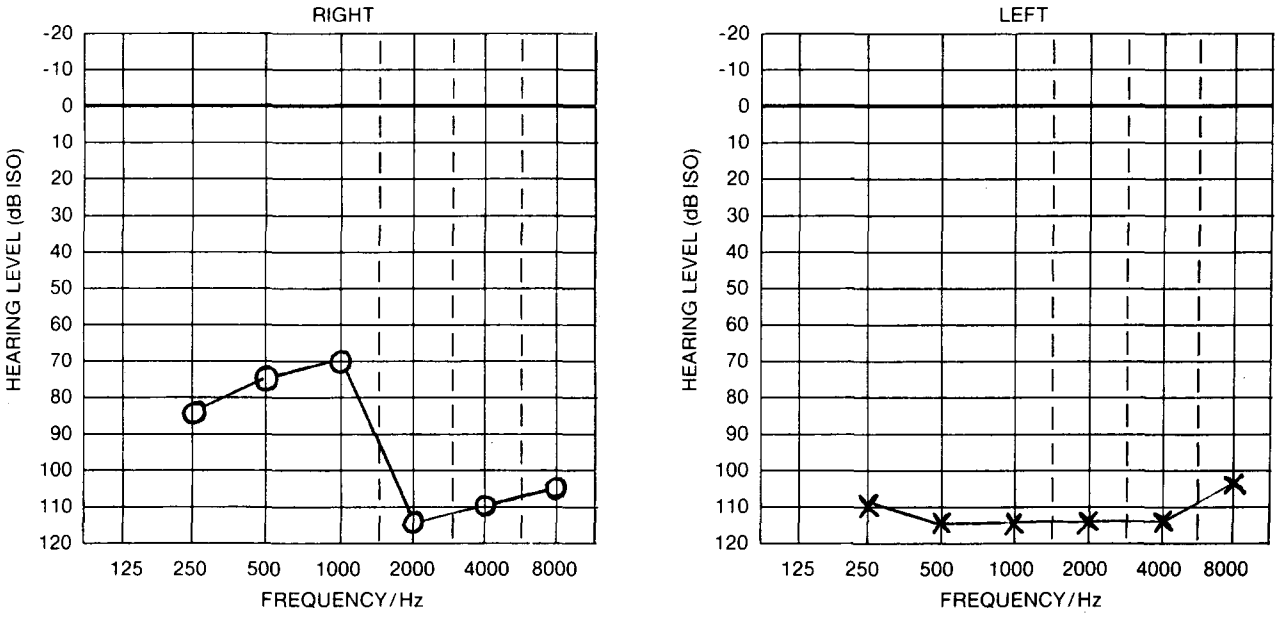

Fig. 5
Audiogram following immunosuppressive therapy.

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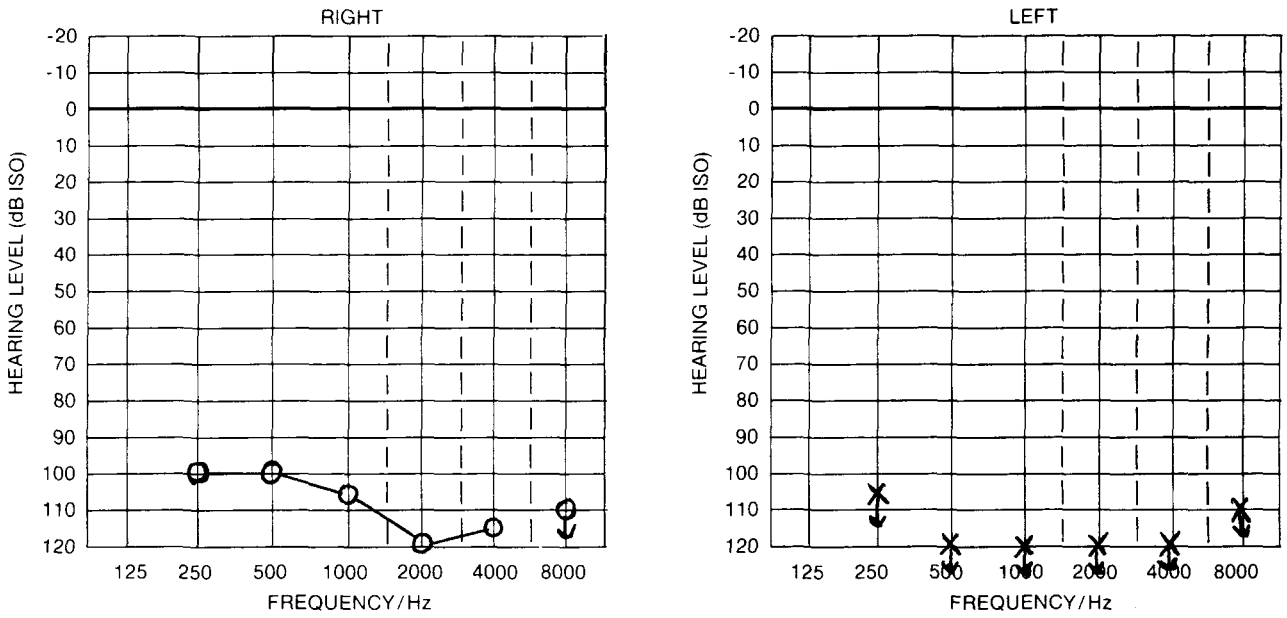

Fig. 6
Audiogram one month after cessation of treatment.

cochlear implant and is awaiting surgery. Her current treatment includes prednisolone (5 mg twice daily), dapsone (50 mg daily), and azathioprine (25 mg alternate days).

Discussion

The relationship between sensorineural deafness and autoimmune disorders is well known. The mechanism by which circulating immune complexes cause deafness is not clearly

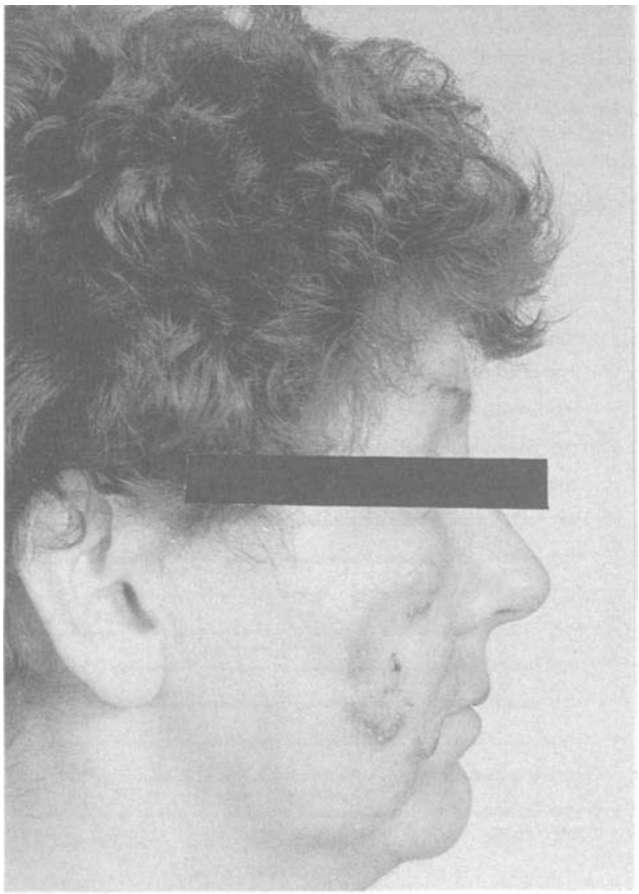

Fig. 7
Pyoderma gangrenosum on cheek.

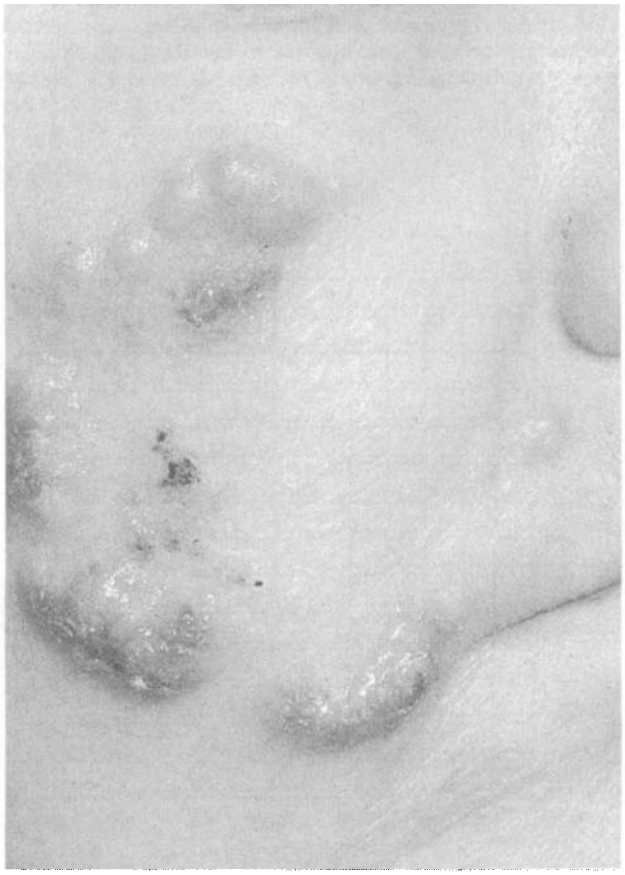

Fig. 8
Close up view of lesions of pyoderma gangrenosum.

https://doi.org/10.1017/S0022215100127793 Published online by Cambridge University Press

understood. The complexes may block the sensitive blood vessels of the stria vascularis leading to ischaemia and hypoxia. Complement activation may also cause more vascular damage and additional tissue inflammatory reaction (Brookes, 1985). Such mechanisms are believed to be involved in Cogan's syndrome, and possibly in syphilis. Several cases of sensorineural deafness associated with ulcerative colitis have been reported (Jacob *et al.*, 1990; Herdman *et al.*, 1991).

Polyarteritis nodosa is another autoimmune condition where damage to the inner ear is caused by circulating immune complexes (Rowe-Jones *et al.*, 1990). Since this condition is associated with significant mortality, autopsy studies of the inner ear have been possible. Extensive vasculitis of the internal auditory artery has been found associated with ischaemic necrosis of the inner ear and fibrosis (Gussen, 1977). Other autoimmune conditions associated with sensorineural deafness include temporal arteritis (Kramer *et al.*, 1988), Wegener's granulomatosis (Clements *et al.*, 1989) and IgA nephropathy, an immune complex glomerulonephritis (Ataya, 1989).

The treatment of deafness associated with autoimmune disorders includes administration of systemic steroids and immunosuppressive agents, such as azathioprine, and varying degrees of success have been reported in different disorders (Summers and Harker, 1982; David and Rees, 1987). Since no satisfactory test exists to confirm the diagnosis of autoimmune inner disease, clinical suspicion alone has been suggested as a sufficient indication in order to treat patients with cyclophosphamide and steroids (McCabe, 1989). Sensorineural deafness in association with ulcerative colitis should be treated similarly (Herdman *et al.*, 1991). Plasma exchange is another option worth considering and has been reported successful in a case of polyarteritis nodosa not responding to steroids and azathioprine (Brookes and Newland, 1986), although disappointing results have been reported by others (Luetje, 1989).

Conclusion

Progressive sensorineural deafness in the absence of any cause may be autoimmune in aetiology and a careful search should be made of any coexisting disorders to establish possible linkage. A trial of steroids and azathioprine is recommended as it may be successful in reversing or at least arresting the progress of sensorineural deafness.

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