Sensorineural deafness – a new complication of ulcerative colitis?

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Summary: A case of sudden bilateral sensorineural deafness associated with active ulcerative colitis is reported. Evidence for an autoimmune basis for this condition is reviewed and the potential benefit of systemic corticosteroids emphasized. The condition may represent a recently recognized extra-intestinal complication of ulcerative colitis.

Introduction

Sensorineural hearing loss is a well recognized condition of diverse aetiology. In 1979 McCabe described a series of 18 patients in whom the onset of deafness was sudden and did not fit well with known categories of nerve deafness. He proposed the new entity of 'autoimmune sensorineural hearing loss' to explain the condition though the evidence for this was incomplete. Since then two independent cases of sudden bilateral nerve deafness associated with active ulcerative colitis have been reported (Summers & Harker, 1982; Weber *et al.*, 1984). Though the aetiology of ulcerative colitis is unknown there is a growing body of evidence to suggest that there are immunological changes in this condition which may be wholly or in part the basis of the disease. This raises the possibility of a direct link between sudden deafness and active colitis perhaps by way of an immune complex vasculitis affecting the inner ear.

If confirmed, such an association could represent a newly recognized extra-intestinal complication of ulcerative colitis. I now report what I believe to be the third case of sudden progressive, bilateral sensorineural hearing loss in a patient with active ulcerative colitis.

Case report

A 13 year old boy of previously good health and normal hearing noticed a short episode of bleeding per rectum in late 1970. Diarrhoea and further bleeding recurred in June of 1971 and he was referred to this hospital for investigation. At that time haemoglobin was 9.7 g/dl, barium enema was normal but proctos-

copy revealed an inflamed and bleeding rectal mucosa. He was given oral iron and multi-vitamins. In August 1971 he complained of dizziness and tinnitus and over the next 24 hours experienced severe loss of hearing in the left ear. By December 1971 a milder and more slowly progressive deafness occurred in the right ear with temporary recurrence of dizziness and tinnitus. Throughout this period he continued to have mild diarrhoea with the passage of blood per rectum. Specific treatment for his colitis was not given until after the deafness was well established and consisted of prednisolone suppositories provided in March 1972 and sulphasalazine 1 g b.d. towards the end of the same year.

Four months after the original onset of deafness the patient was seen by a consultant otolaryngologist who found both auditory meati and tympanic membranes to be intact and healthy. Rinne's test suggest bilateral nerve deafness. X-rays of skull and internal auditory meati were normal and the Wassermann reaction was negative. Audiograms confirmed the presence of a sensorineural deafness with hearing loss of 90 db in the left ear and 50 db on the right. Examination by a consultant neurosurgeon failed to reveal any further neurological abnormalities. At that time no identifiable cause for the deafness could be found.

Follow-up of the patient has continued to the present with severe hearing loss persisting though partly off-set by the development of skilled lip-reading. The diagnosis of ulcerative colitis has been well established on clinical, radiological and histological grounds and was managed medically until 1980. At that time the disease process had spread to involve the whole colon and a pan-proctocolectomy was carried out. Examination of the resected bowel showed mild lymphoid hyperplasia in the terminal ileum. The colon was severely inflamed throughout its length with

ulceration and pseudopolyp formation. Histological examination revealed the features of severe active ulcerative colitis. Since surgery the patient has remained well to the present but with no improvement in hearing.

Discussion

McCabe's original suggestion in 1979 of an immunological basis for some types of sensorineural hearing loss has since received some support. Other conditions known to be associated with an immunological disturbance such as rheumatoid arthritis (Goodwill *et al.*, 1972), Sjögren's syndrome (Doig *et al.*, 1971) and polyarteritis nodosa (Gussen, 1978) have all been shown to have an increased incidence of sensorineural deafness. In the case of polyarteritis nodosa the hearing loss may be due to an immune vasculitis of labyrinthine vessels (Jenkins *et al.*, 1981). Circulating immune complexes in patients presenting with sudden sensorineural hearing loss have been detected by Kanzaki & O-Uchi (1983). These workers report five cases in whom immune complexes were found by the Raji cell immunofluorescence assay so allowing the possibility of a type III immune reaction leading to vasculitis and inner ear damage.

Yoo *et al.* (1983) describe an interesting animal model of immunologically mediated sensorineural hearing loss. They show that immunization of rats with native type II collagen leads to deafness in a large proportion of their experimental animals as compared to controls. Such immunization has previously been shown to induce both arthritis and chondritis of the ear lobe in rats. Circulating antibody to type II collagen is readily detectable by ELISA in immunized animals and immunofluorescence studies demonstrate the deposition of IgG in the wall of the cochlear artery and perivascular fibrous tissue in all immunized rats but not in the control animals. Histologically there was vasculitis of the cochlear artery and degenerative

changes in the spiral ganglion cells of the cochlea. Overall, these findings were taken to indicate an antibody-mediated vasculitic mechanism underlying the deafness, although a Type IV or cell-mediated hypersensitivity reaction cannot be fully ruled out.

The immunology of ulcerative colitis has been extensively studied though whether the changes described are aetiologically important remains uncertain. Both immune complex phenomena (Hodgson *et al.*, 1977; Kemler & Alpert, 1980a) and cell-mediated cytotoxicity (Kemler & Alpert, 1980b) are well described and probably underlie many of the extra-intestinal complications of this condition such as arthritis, erythema nodosum and cutaneous vasculitis. That similar mechanisms could cause deafness via vasculitic or parenchymal change to the organs of the inner ear in some cases of active ulcerative colitis is by no means impossible.

Finally, evidence suggesting a response in immune sensorineural deafness to steroid treatment should be mentioned. This would again be consistent with such deafness having an immune basis. Both McCabe (1979) and Kanzaki & O-Uchi (1983) report that systemic corticosteroids, sometimes in combination with cyclophosphamide, produce marked alleviation of this type of deafness if given soon after its onset. In both previous cases in which deafness was reported in association with active ulcerative colitis, the hearing loss responded to treatment with dexamethasone and cyclophosphamide (Summers & Harker, 1982) and methylprednisolone (Weber *et al.*, 1984). Regarding the patient presently reported, no conventional treatment for colitis was provided for some 16 months after the onset of symptoms. It was during this period of continued colitic activity that deafness of the left ear followed by the right made an appearance. If sensorineural deafness is a newly recognized complication of ulcerative colitis its confirmation as such and general recognition is important not least because it represents a treatable cause of otherwise permanent nerve deafness.

References

- DOIG, J.A., WHALEY, K., DICK, W.C., NUKI, G., WILLIAMSON, J. & BUCHANAN, W.W. (1981). Otolaryngological aspects of Sjögren's syndrome. *British Medical Journal*, **4**, 460.
- GOODWILL, C.J., LORD, I.J. & KNILL-JONES, R.P. (1972). Hearing in rheumatoid arthritis. A clinical and audiometric survey. *Annals of the Rheumatic Diseases*, **31**, 170.
- GUSSEN, R. (1978). Polyarteritis nodosa and deafness: A human temporal bone study. *Archives of Otorhinolaryngology (NY)*, **217**, 263.
- HODGSON, H.J.F., POTTER, B.J. & JEWEL, D.P. (1977). Immune complexes in ulcerative colitis and Crohn's disease. *Clinical and Experimental Immunology*, **29**, 187.
- JENKINS, H.A., POLLAK, A.M. & FISCH, U. (1981). Polyarteritis nodosa as a cause of sudden deafness. A human temporal bone study. *American Journal of Otolaryngology*, **2**, 99.
- 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.
- KEMLER, B.J. & ALPERT, E. (1980a). Inflammatory bowel disease associated circulating immune complexes. *Gut*, **21**, 195.
- KEMLER, B.J. & ALPERT, E. (1980b). Inflammatory bowel

- disease: study of cell mediated cytotoxicity for isolated human colonic epithelial cells. *Gut*, **21**, 353.
- McCABE, B. (1979). Autoimmune hearing loss. *Annals of Otology, Rhinology and Laryngology*, **88**, 585.
- SUMMERS, R.W. & HARKER, L. (1982). Ulcerative colitis and sensorineural hearing loss: Is there a relationship? *Journal of Clinical Gastroenterology*, **4**, 251.
- WEBER, R.S., HERMAN, A.J. & NEWTON, J.C. (1984). Sensorineural hearing loss associated with ulcerative colitis. *Archives of Otolaryngology*, **110**, 810.
- YOO, T.J., CREMER, M.A., TOMODA, K., TOWNES, A.S., STUART, J.M. & KANG, A.H. (1983). Type II collagen-induced autoimmune sensorineural hearing loss and vestibular dysfunction in rats. *Annals of Otology, Rhinology and Laryngology*, **92**, 267.