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Bilateral acoustic schwannoma: post-operative hearing in the contralateral ear

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Abstract
A patient with neurofibromatosis two (NF2) presented with bilateral acoustic neuroma. Pre-operative audiometry revealed a dead right ear and severe left-sided sensorineural hearing loss. Following surgical removal of the larger right acoustic neuroma we have documented a sensorineural improvement of 45 dB in the contralateral ear on pure tone audiometry, which as far as the authors are aware has not previously been described.

Introduction

A large lesion in the cerebellopontine angle can lead to displacement of the brainstem and its compression against the posterior surface of the contralateral temporal bone. The contralateral VIII nerve emerging from the porus acousticus may also be compressed in this area of the posterior cranial fossa. Both brainstem compression and deformation of the contralateral VIII nerve have been suggested as the aetiological factors of abnormal contralateral auditory function in cases of large acoustic tumours (Zapulla *et al.*, 1982; Musiek and Kibbe, 1986; Johnson and Selters, 1987; Moffat *et al.*, 1989). Much of this work has centred on auditory brainstem responses (ABR) and delays in the later waves arising from the complex central auditory pathways. In this case the extent of the pre-operative bilateral hearing loss meant that ABR recordings were not possible. Previous authors describe a contralateral improvement in electrophysiological audiological function after removal of an acoustic neuroma by the reversion of an abnormal ABR to normal (Deans *et al.*, 1990). In this case it was the pure tone audiogram that improved and this provides confirmation of improving auditory pathway function in the contralateral ear after surgery.

Case report

A 32-year-old female presented with a seven-month history of right sided progressive hearing loss, accompanied by high frequency tinnitus which was continuous and of moderate severity (the patient could hear it during the day above the ambient noise). Over the same time period, she also noticed generalized headaches with acute, right sided exacerbations associated with bilateral blurring of vision. Over the previous three months, she had noticed mild dysequilibrium but no true vertigo. Of note in her past medical history was the removal two years previously of two peripheral neurofibromata, one on the right arm and the other in the right supraclavicular area. There was no family history of neurofibromatosis.

Neuro-otological examination revealed a sensorineural hearing loss on the left and a clinically dead ear on the right side. Positive findings included bilateral absent corneal reflexes, ataxia, past pointing, dysdiadochokinesis, positive Romberg and Unterberger stepping tests and bilateral papilloedema. There was no nystagmus. Pure tone audiometry (Fig. 1) confirmed a dead ear on the right and a left sensorineural hearing loss of 105 dB (average 1, 2, 4 kHz). This hearing loss was too profound for Auditory Brainstem Responses and Speech Audiometry. A computed tomographic (CT) scan (Fig. 2) revealed bilateral cerebellopontine angle lesions, larger on the right, (3×2 cm on the right compared with 2.5×1.8 cm on the left, AP and medial lateral measurements respectively) with evidence of brain stem compression and secondary hydrocephalus. A diagnosis of neurofibromatosis was made. Conseqeuently a left ventriculoperitoneal shunt was inserted with dramatic improvements in her headaches and ataxia. The larger right-sided lesion was removed via a translabyrinthine approach with identification and preservation of the VIIth cranial nerve.

The patient made an excellent recovery, and histopathological examination confirmed that the lesion was a schwannoma, and the patient noticed a subjective improvement in her hearing on the left side post-operatively. This was confirmed by pure tone audiometry (Fig. 3) with a 45 dB average (1, 2, 4 kHz) improvement and the patient was able to change from a BE 53 hearing aid to a BE 34.

Discussion

A diagnosis of neurofibromatosis 2 (NF2) can be made on the basis of a bilateral acoustic neuroma or of a first degree relative with NF2 and either a unilateral acoustic neuroma or two of the following: neurofibroma, meningioma, glioma, schwannoma or juvenile posterior subcapsular lenticular opacity (Martuza and Eldridge, 1988; Adams and Victor, 1989; Glasscock *et al.*, 1989; Baldwin and Le Master, 1989). It is seen in one in 50,000 individuals and is a disorder distinct from neurofibromatosis 1 (NF1), the most common form of neurofibromatosis.

There is a strong autosomal dominant penetrance and linkage to an abnormal gene near the long arm of chromosome 22 (Seinzinger *et al.*, 1987; Wertelecki *et al.*, 1988) has been demonstrated. Glial growth factor and decreased antigenic activity of nerve growth factor (NGF) are also believed to play an important role. Despite the strong autosomal dominant penetrance only half of these patients have a positive family history suggesting a significant number of spontaneous mutations leading to tumour growth (Glasscock *et al.*, 1989).

Symptoms of hearing loss, tinnitus, unsteadiness over uneven ground or in the dark, headache, facial weakness, trigeminal sensory changes, visual disturbances and seizures may

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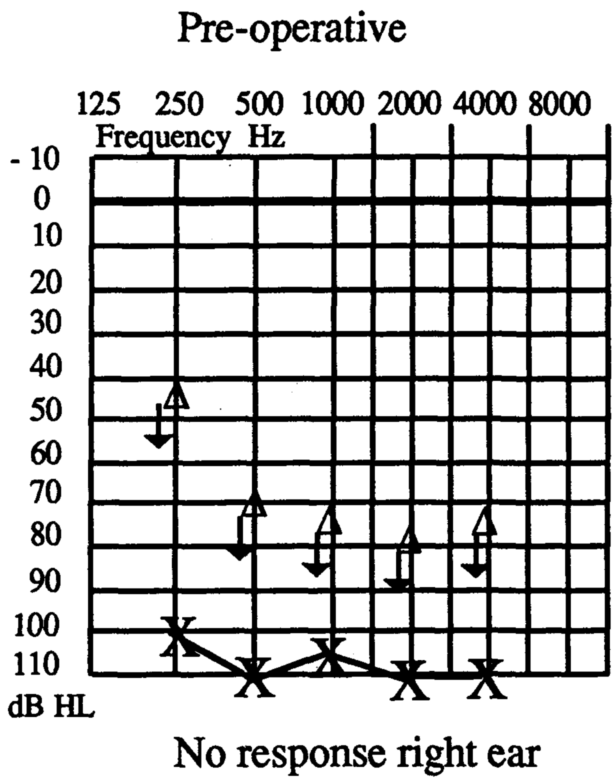

FIG. 1

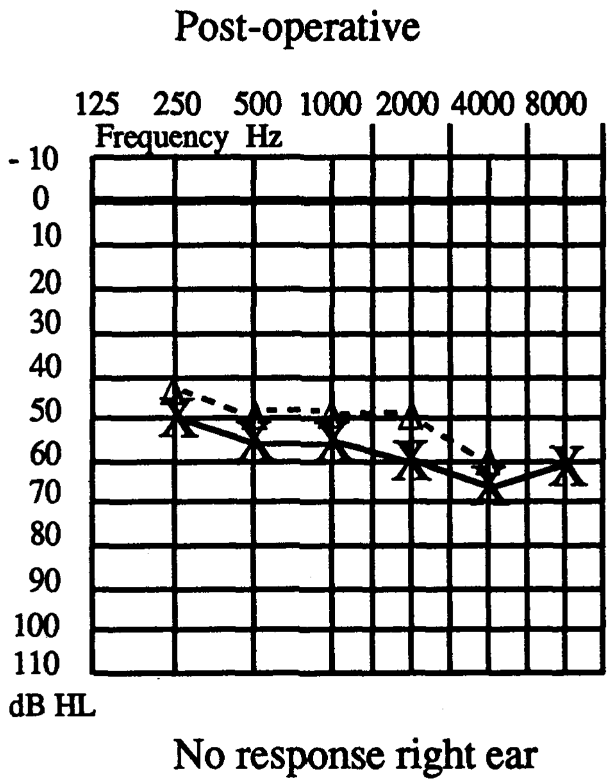

FIG. 3

develop in the 'teens or early twenties but can occur as late as the VIIth decade. Bilateral sensorineural hearing loss in patients younger than 30 years of age should be viewed with a high index of suspicion. Peripheral neurofibroma and *café-au-lait* spots although less common can occasionally be found on thorough investigation. Pure tone audiogram, speech discrimination, ABR and vestibular evaluation may show characteristics consistent with retrocochlear lesions.

Magnetic Resonance Imaging (MRI) with gadolinium DTPA enhancement has largely superseded air meatography in the diagnosis of small intracanalicular and cerebellopontine angle lesions. It is a non-invasive technique and carries the added advantage of being able to image unsuspected contralateral tumours. It produces high definition multiplanar images of soft tissue structures particularly within the internal

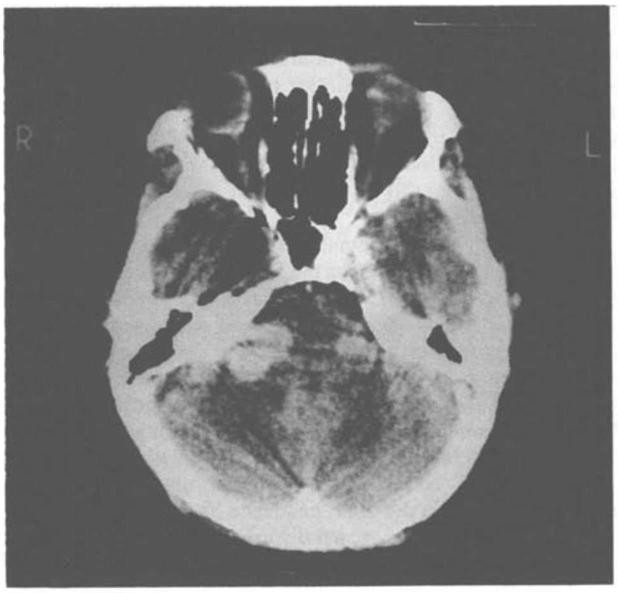

FIG. 2

auditory meatus and bone gives a negative signal. This is in contrast to CT scanning which gives an accurate definition of the bony contour and as such provides an excellent means to demonstrate any temporal bone pathology.

An intimate association of schwannoma with neurofibroma may often be demonstrated with multiple schwannoma being a recognized feature of neurofibromatosis. Histopathologically, in both types of tumour the essential cells are schwannian in origin (Russell and Rubinstein, 1989). Schwannomas are often described as having a pallisading arrangement with interwoven bundles of long bipolar spindle cells (Type A tissue of Antoni). A pleomorphic type is also recognized (Type B tissue of Antoni). Neurofibromas on the other hand have an increased connective tissue content with collagen being a conspicuous element and nerve fibres throughout tumour substance are often both numerous and macroscopically more obvious.

Audiological improvement in the contralateral ear after the removal of one acoustic neuroma in cases of bilateral tumour is presumably due to decompression of the brain stem and contralateral cerebellopontine angle cistern but is difficult to ascribe to one particular part of the complex neuro-anatomical network of the auditory pathway. Since the contralateral tumour is still *in situ* on the functioning nerve the presumptive mechanism of hearing improvement is a decompression of the VIIIth nerve tumour and brainstem complex.

Despite the rarity of bilateral lesions, the same theories propounded by Musiek and Kibbe (1986) and Moffat *et al.* (1989) describing ABR changes at a neural level in contralateral ears with unilateral lesions may well be relevant in understanding the increased audiological perception seen here. This improvement in hearing may have been brought about by the same mechanisms which lead to the contralateral ABR reversion to normality in a post-operative patient as described by Deans *et al*. (1990).

The exact mechanism of the hearing improvement is unknown, but in view of the speed of recovery it may be related to an increase in microvascular supply to the auditory pathway rather than a decrease of intraneuronal oedema as in neuropraxia (which would follow decompression by tumour remo-

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val as described by Moffat *et al.* (1989) and Musiek *et al.* (1986) respectively).

Despite the obvious conjecture about these theories, the final result of improved hearing is irrefutable. It is recognized that the management of patients with bilateral acoustic neuroma is a difficult problem. It is usual to remove the tumor in the worst hearing ear first (this is often but not inevitably the largest tumour) and await the development of deafness or progressive neurological signs before excising the contralateral tumour. The conservation and improvement of hearing in this case is possibly unique as hearing in the contralateral ear is followed closely in these cases. An increased awareness of this finding should ensure that audiological evaluation will be carried out in the contralateral ear in all unilateral lesion removal in cases of bilateral acoustic neuroma.

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