Menière's Syndrome in Otosclerotics

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Deafness, tinnitus, and intermittent vertigo are the well-known classical symptoms of Menière's disease. This triad is so dramatic and so characteristic of the disease that one may sometimes be tempted to make the diagnosis on the history alone and to omit a time-consuming investigation of hearing and vestibular function.

The purpose of this paper is to show that sustained bouts of giddiness may occasionally occur in patients suffering from otosclerosis and to emphasize the need for accurate audiometric assessment in all cases of aural vertigo. Reports of three cases are given. The patients presented with symptoms typical of Menière's disease, but the diagnosis was confounded by the hearing-loss being proved to be conductive rather than perceptive in nature. Exploration of the middle ear in each instance revealed fixation of the footplate of the stapes by otosclerotic bone.

Case 1

A female shop-assistant aged 24 attended the Radcliffe Infirmary, Oxford, in May 1960. She was accompanied by her mother, who was manifestly concerned about the attacks of giddiness from which her daughter had been suffering for the previous three months. Every fortnight or so she would suddenly and without warning find that everything was going "round and round" with objects moving from left to right, causing her to lose her balance and often to fall over. Each bout would last about 10 minutes, culminate in vomiting, and leave the patient feeling unsteady on her feet for 24 hours afterwards. She had also noted deafness in the left ear for six months and that this was much worse during and for some time after an attack of vertigo. Tinnitus in the left ear had been noted for 10 days only. The patient herself had had no previous history of ear disease, but her paternal grandmother, aged 70, had been deaf in both ears for at least 40 years for no obvious reason.

The patient was a rather anxious young woman of average build. Examination of the central nervous system showed no abnormality and the tympanic membranes were healthy. The Weber tuning-fork test was lateralized to the left. The Rinne test was negative on the left and positive on the right. An audiogram showed a conductive hearing loss on both sides, more severe on the left, with a normal hearing level by bone conduction. Caloric testing revealed normal vestibular function on each side.

On the assumption that the patient was suffering from some condition allied to Menière's disease, she was treated by sedation and restriction of salt and fluid intake. The attacks continued and a course of nicotinic acid was tried, but without benefit. During the six months after she was first seen she noticed a considerable further deterioration in her hearing: by November 1960 the Rinne test had become negative on the right as well as on the left side. Audiometry (Fig. 1) showed that this further loss was in the air-conduction level and that the bone-conduction level had remained unaltered.

In view of this deterioration in the patient's hearing, and in order to clarify the diagnosis, exploration of the left middle ear was carried out under local anaesthesia in May 1961. The mucosa was healthy, but the footplate of the stapes was completely fixed by a large anterior focus of otosclerotic bone. The stapes was removed and replaced by a polyethylene tube prosthesis with a vein-graft over the oval window.

After operation there was a satisfactory improvement in hearing, and this was found to be fully maintained when the patient was last seen in November 1963. During the first six months after operation she had two momentary attacks of giddiness but none at all subsequently. The post-operative caloric test gave a normal response on each side.

FIG. 1.—Case 1. Audiogram showing air- and bone-conduction hearing levels on the left side only. The higher air-conduction curve was recorded in May 1960 and the lower curve shows the hearing level six months later. It is interesting to note that during this period of rapidly increasing deafness the attacks of vertigo were of maximal frequency and intensity.

Case 2

A housewife aged 38 attended the War Memorial Hospital at High Wycombe in April 1963 complaining of severe "giddy turns" for the previous two years. The attacks would vary in frequency between two a day and two a week, each lasting about 20 minutes and leaving the patient feeling "wobbly" for some hours. The onset was sudden, and frequently she would fall to the ground, where she would lie with her eyes closed so as to avoid having to look at her apparently whirling surroundings. Nearly always she would vomit a few minutes after the onset of the attack.

Eighteen years previously, at the age of 20, she had noted the gradual onset of bilateral deafness, which had become markedly worse during the early stages of her last two pregnancies. Her hearing was very much better when there was a background noise, and it was always worse on the right side during and for about 24 hours after an attack of vertigo. Tinnitus, "like a bee humming," had been present on the right side for 12 months, and this would become much louder for some minutes before a bout of giddiness. She said that for no obvious reason her father and eldest brother had also become deaf at an early age.

She was a healthy-looking woman of slight build. The tympanic membranes were normal and tuning-fork tests showed a bilateral conduction hearing loss with normal cochlear function, and this was confirmed by audiometry. Caloric testing gave responses which were within normal limits on each side.

The right middle ear was explored in June 1963. The footplate of the stapes was found to be white, fixed, and grossly thickened

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by otosclerotic bone. A small hole was drilled in the postero-superior part of the footplate and a McGee stainless-steel piston was placed in position so that one end was fixed to the incus and the other protruded a small distance into the vestibule through the hole in the footplate.

When she was last seen in November 1963, five months after her operation, the hearing in the right ear had returned to normal (Fig. 2) and the tinnitus had disappeared. Apart from some unsteadiness for a few days in the immediate post-operative phase there had been no attacks of vertigo of any description since her operation. The post-operative caloric test, like the pre-operative one, was quite normal.

FIG. 2.—Case 2. Audiogram showing air- and bone-conduction hearing levels in the right ear only. The lower air-conduction curve represents the pre-operative, and the upper the post-operative, hearing threshold. The shaded area shows the gain in hearing resulting from stapedectomy.

Case 3

A brickmaker aged 55 had numerous brothers and other relatives who became deaf during early life, and he was not surprised, therefore, to find that he himself developed some difficulty with hearing in his late thirties. During subsequent years this bilateral deafness remained more or less the same and his general health was good until 1959, when he began to have bouts of giddiness, tinnitus in the right ear, and a considerable worsening of his hearing. The giddy turns would strike him suddenly, causing him to fall to the ground if there was no readily available means of support. He described it as a feeling of "everything going round and round." The noises in the right ear became very loud, "like escaping steam," and always he would vomit at least once during each attack. He said that he did not recall any deterioration in hearing either during or after a bout of vertigo, but that there was a considerable overall increase of the deafness at the time when the giddy turns began in 1959. Each vertiginous episode lasted about half an hour and left the patient feeling somewhat unsteady for the rest of the day.

He was treated by salt-and-fluid restriction and also a number of drugs, including promethazine chlorotheophyllinate and dimenhydrinate, but these did not materially alter the course of the disease, and he continued to have giddy turns ranging in frequency from two a week to one a month. Things came to a head in the spring of 1961, when on two occasions he developed acute vertigo in the street, causing him to fall over and his having to be carried home. He was then referred to Radcliffe Infirmary with a view to a surgical destruction of the offending labyrinth.

He was a robust labourer with normal blood-pressure and no abnormal central nervous system signs. The tympanic membranes were healthy and the Rinne test was negative on both sides. Audiometry (Fig. 3) showed that in addition to the conductive deafness there was also a moderate perceptive loss, as shown by the depressed bone-conduction curve, on both sides. A Hallpike caloric test gave

a normal response on the left, but showed a marked reduction of vestibular function on the right.

The right middle ear was explored in November 1961. The stapes was found to be fixed and the footplate was grossly thickened by otosclerotic bone. Because of its thickness it was not possible to effect a complete removal of the footplate, but as large an opening as possible was made into the vestibule by the use of the

FIG. 3.—Case 3. Pre-operative audiogram showing a mixed deafness which is more severe on the right side. Air conduction: X=left; O=right. Bone conduction:]=left; [=right.

micro-drill, and a vein-graft and polyethylene tube were employed to re-establish the continuity of the ossicular chain.

Post-operatively the patient had a good improvement in hearing with closure of the air-bone gap, but when he was last seen in January 1963 this had not been fully maintained. Up to the time of his operation he had continued to suffer from severe attacks of vertigo, but afterwards these had dramatically ceased, though he was still not absolutely sure of his balance at all times. The post-operative caloric test was the same as the pre-operative one, and showed marked diminution but not complete absence of vestibular function on the right side.

Discussion

Search of the literature revealed only occasional reference to vertigo as a symptom of otosclerosis. Cohen (1959) reported the case of a patient with otosclerosis and severe giddiness who was relieved of both deafness and vertigo after a stapes mobilization operation.

Cawthorne (1955) states that patients with otosclerosis sometimes give a history of slight and momentary giddiness when stooping, and attributes this to the effect of postural changes on the labyrinth. He is of the opinion that these minor episodes of postural vertigo are more common in otosclerotic patients than in the normal population.

The three cases recorded here, however, do not fall into this category, as the attacks of vertigo were much more prolonged and of much greater severity, causing the patients to fall to the ground and vomit.

It might be said that these three patients were suffering from both otosclerosis and the labyrinthine hydrops of Menière's disease as two separate and aetiologically unrelated entities. In Case 3, where there was a substantial sensori-neural deafness and a diminished caloric response on one side, it is possible, though statistically unlikely, that the patient did have these two separate diseases coexisting in his right labyrinth. In Cases 1 and 2, however, the presence of normal cochlear function and normal caloric reactions exclude a diagnosis of labyrinthine hydrops, and it must be concluded

that the otosclerotic process was in some way directly responsible for the vestibular disturbances.

Exactly how otosclerosis produces vestibular disturbances is not known, but from our knowledge of the histopathology of Menière's disease and of otosclerosis, and by a careful analysis of the findings in the three cases recorded here, it is possible to make a few valid observations.

It is known that otosclerotic disease can and commonly does extend to involve the cochlea (Ruedi, 1962) and that this can produce a perceptive loss of hearing. It is also known that otosclerosis may involve the semicircular canals (Seligman and Shambaugh, 1951), but the effect of this on vestibular function has not yet been assessed. Schuknecht *et al.* (1962) have compiled considerable histological evidence to show that the attacks of vertigo in Menière's disease are due to rupture of the membranous labyrinth, causing a sudden admixture of the endolymph and perilymph. It has also been shown (Benitez and Schuknecht, 1962) that labyrinthine otosclerosis can cause rupture of the basilar membrane through shrinkage of that part of the spiral ligament which lies adjacent to an otosclerotic focus.

It seems likely, therefore, that the explosive attacks of vertigo are due to sudden rupture of the membranous labyrinth, which has been rendered atrophic by its proximity to a focus of otosclerotic activity.

More difficult to explain is why the vertigo should cease after stapedectomy. Clearly, the only beneficial effect that this operation can confer upon the labyrinth is one of mechanical decompression.

By accommodating any alteration in the volume of the fluids, a mobile stapes footplate must help to prevent violent pressure changes within the labyrinth. If the footplate is fixed, however, this compensatory adjustment no longer operates and the various membranous partitions are subjected to much greater and therefore more disruptive changes in pressure. A healthy endolymphatic labyrinth might be able to withstand these abnormal pressures, but if rendered "fragile" by adjacent otosclerotic disease then rupture could ensue. By restoring the normal "safety-valve" mechanism, stapedectomy mitigates the mechanical stresses imposed upon this delicate

membranous labyrinth and thereby tends to prevent further episodes of vertigo.

As the three cases had bilateral otosclerosis, it would seem to have been a fortunate "fluke" that in each instance we operated on the ear that was causing the attacks of vertigo. In all three patients, however, there were indications which pointed to one ear as being responsible for the vestibular disturbance; loss of balance was accompanied by a temporary worsening of the deafness on one side in Case 1 and by an increase in the intensity of the unilateral tinnitus in Cases 2 and 3. In addition, stapedectomy was always carried out on the ear that showed the greater degree of deafness and was therefore presumed to be the more severely affected by otosclerosis.

Summary

Three cases of otosclerosis are described in which the patients suffered from recurrent severe episodes of vertigo similar to those of Menière's disease. Labyrinthine otosclerosis induces atrophic changes in the membranous labyrinth, and it is postulated that vertigo is due to sudden rupture of the latter. In each patient the vertigo ceased after stapedectomy, and this was probably due to a decompression effect on the labyrinth.

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Immunoassay of Serum Growth Hormone in Acromegalic Patients

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Immunological methods for the assay of growth hormone in human serum have been extensively studied since the introduction of an immunoassay by Read (1959), using the haemagglutination technique. The original assay has proved unsatisfactory owing to interference from substances, probably protein in nature, present in serum (Grumbach and Kaplan, 1962); Dominguez and Pearson (1962), however, have used the method after a preliminary extraction procedure. Immunoassays using radioiodine-labelled human growth hormone (H.G.H.) have been described by Hunter and Greenwood (1962a, 1964), Utiger *et al.* (1962), and Glick *et al.* (1963); we have used a technique very similar to that of Utiger *et al.*

In this paper our technique is described, together with the results obtained during glucose-tolerance tests and insulin-tolerance tests on acromegalic patients.

Methods

Antigen and Antibody Used

Ampouled H.G.H., prepared by the Department of Biochemistry, University of Cambridge, using the Raben (1957) technique, was supplied by the Clinical Endocrinology Committee of the Medical Research Council. It was labelled each week with radioiodine, ¹³¹I, using the method of Hunter and Greenwood (1962b), and specific activities of 150-200 $\mu\text{c}/\mu\text{g}$.

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