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Chronic labyrinthine ischemia

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Abstract
The purpose of the present study was to describe the audiological and vestibular deficit in patients presumed to suffer from chronic impairment of the blood supply to the labyrinth. Thirty-nine subjects affected by various pathologies often impairing systemic blood circulation, which also presented labyrinthine damage of unclear origin, were evaluated for a possible relationship between these two conditions. At the time of this study 80 per cent of the subjects showed also a certain degree of increased blood viscosity. The most common finding in the group was slow and progressive bilateral sensorineural loss of hearing. In most of the subjects the labyrinthine damage began with high-tone loss which, in time, became a flat curve. Less common, but still found in many patients, was a fluctuation in threshold. Short periods of spontaneous subjective improvement in hearing were accompanied by a better result in the Pure Tone Audiogram and Speech Reception Threshold. The discrimination scores remained generally lower than expected. Vestibular examination revealed pathological results for all the subjects in at least some of the tests. The mild subjective complaints concerning equilibrium could be explained by a central nervous system compensation.

Introduction

From an etiological point of view, one can divide ear disease into two categories. The first includes pathologies in which a noxious agent directly affects the ear structures, such as ear infections, acoustic trauma, or Menière's disease. The second category includes pathologies in which the noxious agent primarily affects a body system other than the auditory, with a secondary effect leading to impaired hearing or vestibular function, e.g. cardio-vascular, hormonal or immunological disease. In the latter category, the specialist dealing with the primary disease may not focus on the patient's auditory function, and if the impairment is not too severe, or the patient himself does not complain, hearing and vestibular evaluation may be postponed or neglected. This is unfortunate because there is evidence that labyrinthine damage is partially reversible if identified at an early state and treated.

The relationship between an existing general disease and the impairment of a specific system may be: (a) incidental, i.e. two different diseases existing simultaneously in the same patient, e.g. heart disease and deafness; (b) associated, such as heart disease, with deafness due to quinidine treatment for heart arrhythmia; and (c) casually related, for example, heart disease with decreased cardiac output, leading to insufficient blood supply to the labyrinth and consequent loss of hearing. Needless to say, investigators interested in studying the causal nature of such a relationship should first identify and exclude cases belonging to the first two categories.

The object of the present study was to describe the cochleo-vestibular deficit in patients suspected of having intermittent or continuous labyrinthine ischemia, as may occur in certain general pathologies associated with impaired blood circulation. Labyrinthine ischemia may manifest itself as an acute or chronic disease. An example of the acute form would be 'sudden deafness' in a number of cases. Siegel (1972) estimated the annual number of new cases of 'sudden deafness' in the U.S. to be about 4,000. Most of these cases are probably acute cochlear ischemia, generally of short duration, since most of the patients recover spontaneously. Until now, the chronic form has been neglected by both clinicians and researchers, and is rarely mentioned in literature or textbooks.

In our opinion the number of patients affected by chronic labyrinthine ischemia is much larger than might be suspected. Unfortunately, at the onset, patients, primarily worried about the symptoms of the primary disease, tend to neglect the additional discomfort caused by the mild hearing deterioration. Later, when the hearing loss increases and difficulties in communication become evident, diagnosis is easier but the damage is done and the only option is the use of a hearing aid, even if the hearing deterioration is stabilized as the primary disease comes under control. If such a stablization is obtained in the early stages of the chronic deterioration, a useful degree of hearing could be salvaged.

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Materials and method

Chronic labyrinthine ischemia is generally caused by the combined action of more than one etiological factor. Therefore, the present study sought such cases among patients suffering from various pathologies frequently affecting blood circulation and showing at the same time abnormal blood viscosity.

Fifty-seven patients suffering from different pathologies associated with intermittent periods of high blood viscosity were chosen for this study. All of the patients complained of slow progressive hearing loss and some of them complained of vestibular disturbances. Due to these complaints the patients were under periodical follow-up observations at the Department of Audiology for up to 15 years.

After complete reviews of the files, consultations with attending physicians, and direct contact with the patients, 18 cases were excluded from the study. The reasons for this were: One group included eight patients whose diagnosis of the primary disease had not been fully confirmed at the time of this study. A second group of seven subjects had their first hearing and vestibular examinations within three years of the study. In order to evaluate the rate of deterioration and the fluctuating nature of the hearing loss, it was decided that a minimal period of three years was necessary. Finally, three patients considered their hearing loss to be the result of noise exposure and refused to be re-examined.

The 39 remaining patients formed the subject population of the present study. Their ages ranged from 46 to 71 years, with a mean of 61 years. The nature of their primary pathologies are presented in Table I.

Procedure

After the subjects were examined by their attending specialists for the general disease, and by an otologist, they were scheduled for hearing and vestibular examinations. Each patient underwent Pure Tone (PT) and Speech Audiometry, a test for Recruitment, Tone Decay, Stapedial Reflex, Decay, and some subjects also underwent Brainstem Evoked Response (ABR).

Vestibular evaluation was done for all patients on the basis of the results of Optokinetic, Gaze and Pursuit tests, as well as positional, pre- and post-rotatory and Hallpike Caloric Tests. Blood obtained without stasis was collected on EDTA, and its viscosity was examined within 30 minutes. A Brookfield microviscometer was used, and the examination was done at eight rates of shear between 230/sec.$^{-1}$ and 1.15/sec.$^{-1}$.

TABLE I
DISTRIBUTION OF PATIENTS ACCORDING TO THE GENERAL DISEASE (N=39)

No. of patients	General disease
11	Heart disease with decreased cardiac output
9	Cerebral-cerebellar vascular insufficiency
9	Polycythemia
4	Monoclonal gammopathy
6	Multiple myeloma

Results

Auditory Findings

All patients in the present study showed some degree of bilateral, progressive sensorineural hearing loss. In 16 subjects the hearing was tested at the onset of the ear complaints, and in ten of these only high-tone loss was found. The finding, however, changed over time and during the present study the most common hearing loss was a flat curve. The mean loss for the entire group, for frequencies between 0.5–4.0 KHz, was 50 dB HL in the more damaged ear and 32 dB HL in the less damaged ear. The Speech Reception Threshold (SRT) matched the average loss for speech frequencies. Discrimination for PB words yielded scores slightly lower than expected from the pure tone level. Loudness recruitment testing gave generally negative results, with exceptions observed occasionally during acute exacerbations in the condition of the ear.

Results of Threshold Tone Decay were normal. Stapedial Reflex Decay was tested in 15 cases, and in none was pathological reflex decay detected. Auditory Brainstem Response (ABR) was checked in only nine patients. Despite the high hearing threshold, the response obtained at 85 dB HL showed normal latency and pattern. High pitch tinnitus and 'fullness' in the ear were generally mentioned by the patients in relation to acute exacerbations of hearing loss. Thirty-one out of the 39 patients complained of one or more periods of fluctuating hearing loss, or temporary spontaneous improvement in hearing. Improvement was considered as a change in the PT audiogram of 10 dB or more in at least two of the four speech frequencies. Such an improvement was associated with a better SRT and/or improved discrimination scores, and was generally observed in the less affected ear.

Vestibular Findings

Spontaneous nystagmus was observed in eight patients, and in another eight subjects over-shooting during calibration was noted. Optokinetic tests showed slight dysmetric and dysrhythmic tracing in eleven subjects. The pursuit test was saccadic in seven subjects. In nearly one third of the patients (12) positional nystagmus of a peripheral type was observed. The findings of the Hallpike Caloric Test were abnormal in eleven cases (slight preponderance). Per-rotatory nystagmus, at least in one direction, was elicited in nine subjects only at 6°/sec^2, but the post-rotatory response was different for the two directions of rotation in 15 cases. The results are summarized in Table II.

It can be seen that almost all of the patients showed pathological results in at least some of the vestibular tests, in spite of their rather modest complaints concerning vertigo. It is probable that in time a compensation of central origin occurred, thus minimizing the discomfort caused by the symptoms.

Blood Analysis

The examination of the whole blood for viscosity showed that at low shear rates—1.15/sec^{-1}—most of the patients showed pathological values of viscosity, and

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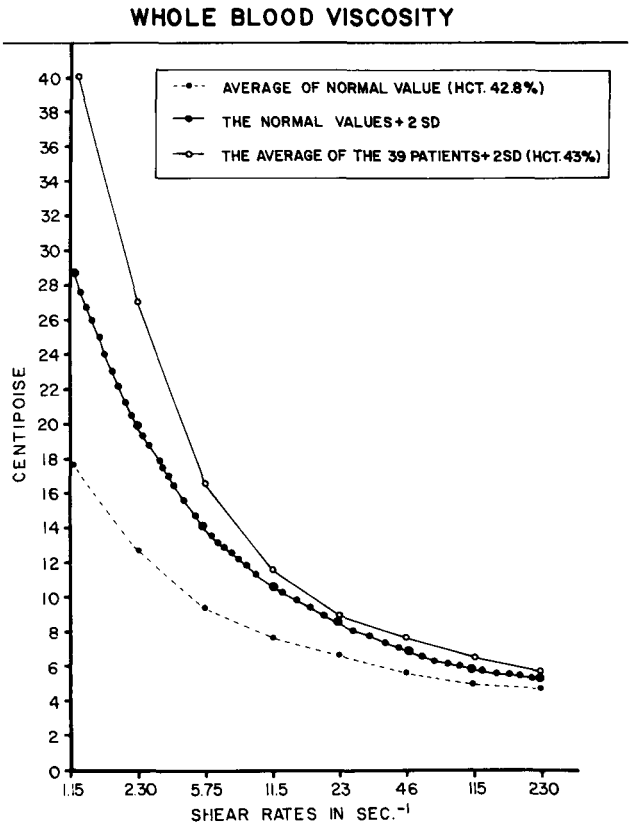


FIG. 1

The graph illustrates the average viscosity of the 39 patients at 8 shear-rates and the upper limit of normal viscosity—the 2 S.D. curve.

even at high shear rates—230/sec^{-1}—one third of the patients still showed higher normal values of whole blood viscosity. Figure 1 presents the mean value of viscosity at the eight shear rates for the whole group. The results can be compared with the normal values obtained in our laboratory, as presented in Table III.

Discussion

The present study intended to describe the clinical profile of cochlear and vestibular ischemia due to decreased blood supply to the labyrinth. Such a condition may be the result of a pathology affecting only the labyrinthine artery (Valvassori and Dobben, 1979); however, it is more frequently associated with impaired circulation to the whole brain (Glen *et al.*, 1984), or to the entire body (Davis and Nilo, 1965). At present state of the art methods for quantifying a decrease in the normal blood supply to the inner ear itself are not available. Consequently, a diagnosis of chronic labyrinthine ischemia is based on supporting evidence, with greater or lesser degrees of probability. It should, however, be kept in mind that even in the presence of clinical symptomatology and radiological evidence of decreased blood flow to the brain, a simultaneous labyrinthine damage need not necessarily be due to a poor labyrinthine blood supply.

Clinically, the difference between a localized and a more general pathology was in the extent of symptoms presented by the patient. Symptomatology was considered restricted when only the labyrinth was affected, and more complex when additional parts of the brain showed functional deficit. Labyrinthine ischemia might occur not only in cases with impaired posterior brain circulation (vertebrobasilar), but with anterior (carotid) artery involvement as well (Mills and Ryals, 1985). Correlation between the severity of hearing loss and the extent of blood circulatory disturbances could not be established.

From among the three main causes of such a condition—altered cardiac output, affected blood vessels and abnormal blood viscosity—rarely can a chronic deterioration of hearing or equilibrium be explained on the basis of only one of these pathologies. For instance, a high hematocrit (HCT)—hence elevated viscosity—in a young subject with normally elastic vessels and a good cardiac output (good *vis a tergo*) would not impair the blood supply to the labyrinth, but if it was associated with narrowed and inelastic vessels it certainly might. This would also be the case in an older but asymptomatic patient with an insufficient vertebro-basilar system com-

TABLE II

AUDITORY AND VESTIBULAR FINDINGS IN 39 PATIENTS WITH SUSPECTED CHRONIC LABYRINTHINE ISCHEMIA

Auditory findings	Percentage of Occurrence
Bilateral sensorineual loss of hearing—mean for entire group (0.5–4.0 kHz) 50 dB worse ear, 32 dB better ear	100%
SRT matched average PT loss for speech frequencies	100%
Discrimination score slightly lower than expected from PT	76%
Occasional tinnitus of high and changing pitch	16%
Loudness recruitment present only in acute exacerbation (loudness balance os SISI)	13%
Threshold tone decay (Rosenberg modification)	None
Fluctuating threshold	79%
Decay in stapedial reflex (15 patients tested)	None
Normal pattern and latency in ABR (9 patients tested)	100%
Vestibular findings	
Overshooting during calibration	20%
Spontaneous nystagmus	20%
Optokinetic test slightly dysmetric and dysrhythmic	28%
Pursuit test saccadic	30%
Caloric test—slight preponderance	23%
Elevated threshold on per-rotatory slow acceleration test	28%
Assymetric post-rotatory nystagmus	38%

TABLE III

NORMAL VALUES OF WHOLE BLOOD VISCOSITY FOUND IN OUR LABORATORY

Shear rate sec^{-1}	Mean Centipoise	S.D.
230	4.5	0.3
115	5.0	0.4
46	5.7	0.5
23	6.6	0.8
11.5	7.7	1.4
5.75	9.4	2.3
2.30	12.8	3.6
1.15	17.5	5.4

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pensated for by blood streaming backward from the circle of Willis. A temporary increase in blood viscosity could break the poor compensation and decrease the blood supply to the labyrinth. Thomas *et al.* (1977), for example, found that lowering the HCT from 53.6 to 45.5 was associated with a 30 per cent decrease in viscosity and with a 73 per cent increase in cerebral blood flow (CBF). Willison *et al.* (1980), examining patients with high HCT for alertness, found significantly poorer performance than in a group of subjects matched for age and occupation, but with normal HCT. Rubinstein *et al.* (1977), comparing aged patients with and without cardio-vascular disease, found a significantly higher hearing threshold in the group with an impaired cardio-vascular system.

In the current study, 39 patients with blood-flow disturbances due to various pathological conditions, and presenting affected equilibrium and bilateral sensorineural hearing loss, were re-evaluated for a possible relationship between abnormal blood circulation and damage to the ear. During the relatively long period of observation (averaging approximately five years) slow progression of hearing deterioration with fluctuations in threshold were the most constant features of chronic labyrinthine ischemia. Temporary improvement sometimes occurred in both ears and sometimes only in the less affected ear. The magnitude of hearing improvement ranged between 10 and 20 dB, and lasted for several weeks or months. The patients were aware of the change in their hearing, and attributed it to their improved general condition.

Another interesting feature of this hearing deterioration is that speech reception threshold matched the mean loss for pure tones in the speech range, but the discrimination scores were generally lower than expected from the Pure Tone audiogram. The hearing and vestibular examinations reveal the fact that some of the tests indicate a peripheral lesion, while others indicate a more central one. For instance, absence of Recruitment and Stapedial Reflex elicited only at high suprathreshold sensation levels suggest a more central lesion that the peripheral transducer, but the normal pattern of the ABR response with latency prolonged only in accord with the elevation in threshold do not support this hypothesis. The same phenomenon is noted in the vestibular tests. Overshooting during calibration or saccadic tracking test indicates a central lesion but the positional nystagmus was obviously of peripheral origin.

This combination of peripheral and central lesions may be explained in two ways. Firstly, when the impaired blood flow to the labyrinth is due to a general pathology, the existence of ischemic lesions in other parts of the brain is highly probable. Secondly, the cochlear spiral ganglion, Scarpa's ganglion, and the beginning of the VIII nerve have the same blood supply as the peripheral transducer, but damage to these structures will probably show up as a central lesion.

The discrepancy observed between the deficit in vestibular functioning, as demonstrated by the results of the various examinations, and the modest complaints of the patients can be explained by central nervous system compensation. According to Igarashi (1984) '. . . this compensation implies the reorganization of the neural circuits which eliminates the functional deficit and reinstitutes bilateral balance.' Therefore, in spite of the abnormal responses obtained in some of the tests, the subjective complaints concerning vertigo remained relatively mild.

Conclusion

In order to foster early identification of chronic labyrinthine ischaemia, as represented by a slow and progressive hearing loss, several diagnostic procedural steps are recommended. Firstly an atypical but still local pathology limited to the labyrinth should be considered. If none is identified, then the presence of a regional or a general pathology which might explain an eventual impairment of the blood flow to the ear should be considered. Investigation of symptoms of impaired blood flow to regions other than the labyrinth, e.g. amaurosis fugax, intermittent claudication, etc. by cardio-vascular insufficiency should be made.

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